CHORISTOMA OF THE TONSIL

P. C. Modayil[1], A. Joseph[2], I. Thomas3, G. Sulochana[4], J. Xavier[1]

ABSTRACT: *A 13 year old female patient presented to ENT OPD with chronic recurrent tonsillitis and underwent tonsillectomy. The histopathological examination revealed heterotopic cartilage in both the tonsils. The postoperative period was uneventful. The histopathologic finding of choristoma of the tonsil is rare.*

Key Words: *giant cell reparative granuloma*

Choristoma or heterotopias are aggregates of microscopically normal cells or tissues in aberrant locations (Rubin and Farber, 1988). Hamartomas are tumour like, but nonneoplastic overgrowth of tissue that is disordered in structure. e.g., haemangiomas.

The word choristoma (choris = separated, oma = tumour) implies a neoplasm where as heterotopia refers to displaced tissue without necessarily being a swelling or neoplasm. Since the present case had tonsillar enlargement, we have used the term choristoma rather than heterotopia. Only, two cases have been reported so far (Bhargava and Raman, 1996).

CASE REPORT

A 13 year old girl presented to the ENT OPD with complaints of recurrent sore throat, fever and painful swallowing for the past 2 years. On general examination, the patient had no pallor or lymphadenopathy. On ENT examination, tonsils showed grade two hypertrophy. The ear and nose examination did not reveal any significant pathology. She underwent bilateral tonsillectomy and the specimen was sent for histopathological examination.

The histopathological examination surprisingly revealed tonsillar lymphoid tissue with islands of mature cartilage [Figure 1]. The cartilage was partially calcified and ossified [Figure 2].

DISCUSSION

Choristomatous salivary tissue was first described by Taylor and Martin (1961) in the middle ear. Since then, many other cases of choristomatous salivary tissue have been described in the middle ear, Eustachian tube and in the external ear (Friedman, 1989). Other sites of heterotopic bone formation have been reported in the eye (Akimou, 1989) and renal pelvis. (Melekos et al., 1988)

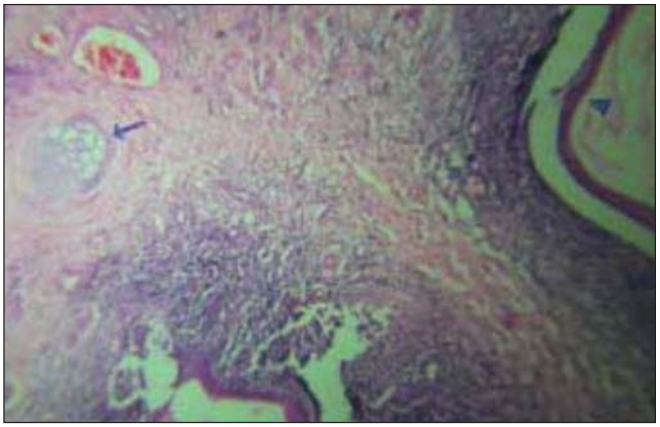

Figure 1: Histopathological photomicrograph
H & E section, 10X x 10. The single arrow shows island of mature cartilage and the lymphoid tissue can be seen on the bottom. The Arrow head shows tonsillar crypt with keratin pearls.

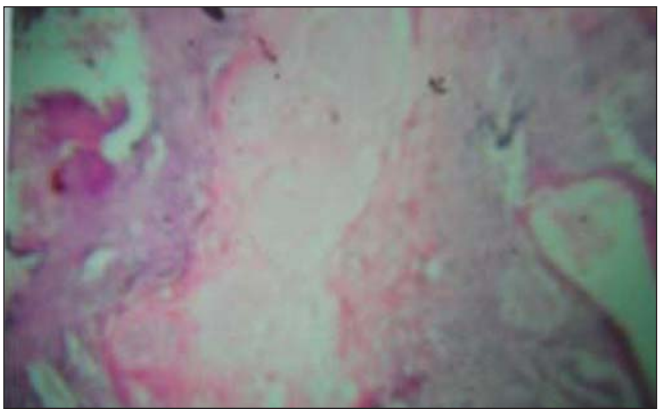

Figure 2: Calafication and ossification of the mass.
H & E section,10Xx 10. The island of cartilaginous tissue shows partial ossification. (eosin stained).

The mechanism of the pathogenesis of heterotopia was suggested by Lindholm et al., (1973). The chronic inflammation might have led to the release of osteogenic

[1]Registrar, [2]Assistant Professor, [3]Lecturer, Department of ENT, Pushpagiri Institute of Medical Sciences, Thiruvalla, [4]Professor, Department of Pathology, PIMS, Thiruvalla, Kerala, India

substances producing heterotopic cartilage proliferation and bone formation. Heterotopic rests need to be differentiated from dystrophic bone formation and metastatic calcification.

CONCLUSION

The histopathologic finding of heterotopia or choristoma is rare. The choristomatous rests are usually of academic interest only. Clinically, choristomatous rests can be confused with true neoplasms, when they are sufficiently large. Rarely, they are the site of origin of true neoplasm.

This congenital anomaly is better described as a heterotopic rest of cells. The term choristoma, connoting a neoplasm, imparts to the heterotopic rest a gravity far beyond its trivial significance. Although, regrettably the terminology of neoplasms is not simple, it is important because it is the language by which the nature and significance of tumours are categorized. (Robbins et al., 1989)

REFERENCES

1. Akimou OV. Heterotopic formation of bone tissue in the eye. Vestnik Oftalmologii (moskua) 1989;105:15-6.
2. Bhargava D, Raman MS. Heterotopia of the tonsil. J Laryngol Otol 1996;110:611-2.
3. Friedman I. Nose Throat and Ears. *In* Systemic Pathology, 3rd edn, vol.1, (Symmers, WC. ed.) Churchill Livingstone: Edinburgh; 1989. p. 321.
4. Lindholm ST, Hackman R, LindHolm RV. Histodynamics of experimental heterotopic osteogenesis by transitional epithelium. Acta Chirurgica Scandinavica 1973;139:617-23.
5. Melekos MD, Pantaakos A, Grekou AN, Ghikas C, Gianno U. Heterotopic bone formation in the renal pelvis. Br J Urol 1988;61:458-60.
6. Robbins RS, Cotran V, Kumar SL. *In* Robbins pathological basis of disease.7th edn (Reprint 2004). 1989. p. 168.
7. Rubin E, Farber JL. Pathology. JB. Lippincott Company: Philadelphia; 1988. p. 248.
8. Taylor GD, Martin HF. Salivary gland tissue in the middle ear. Arch Otolaryngol 1961;73:651-3.

Address for Correspondance
Dr. P. C. Modayil
Modayil House, Mallappally West p.o.,
Pathanamthitta dist
Kerala – 689585, India,
E-mail: princemodayil2000@yahoo.co.in

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ORIGINAL ARTICLE

Spaced Intranasal Corticosteroid Therapy: A Better Treatment Option in Allergic Rhinitis?

Ike Thomas[1] · Tina Thomas[1] · Geethu Mathew[2] · Mahika Anil Kumar[3] · Kalyan Varghese George[3]

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Abstract Intranasal corticosteroids are first-line therapy in the treatment of allergic rhinitis (AR) and are conventionally prescribed once daily as continuous therapy. The decreased consumption of drugs is proposed to have decreased side effects. The present study aimed at comparing the effect of INCS as a spaced therapy with the conventional continuous therapy. Case records of patients with Allergic Rhinitis, who were started on INCS were studied and improvement in symptom score was compared between continuous and spaced therapy groups. In total 182 patients with AR were studied, with 91 patients in each group. Among the total group, 57% were males, 54% were < 40 years of age, 54% had > 10 years of allergy history and 94% had no family history. There was significant improvement in mean Visual Analogue Score (VAS) for all patients in both groups ($p = 0.001$). However, the comparison of differences in VAS before and after therapy did not show significant difference for the two groups ($p = 0.791$). Our study suggests that the efficacy of INCS in controlling AR symptoms is observed to be similar with spaced therapy, as in continuous therapy. Spaced therapy may therefore be recommended for better patient compliance, lesser cost and avoidance of the side effects resulting in overall improvement of quality of life for allergic patients.

✉ Tina Thomas
tinaofficialent@gmail.com

[1] Department of Otorhinolaryngology, Believers Church Medical College Hospital, Tiruvalla, Kerala 689 103, India

[2] Department of Community Medicine, Believers Church Medical College Hospital, Tiruvalla, Kerala, India

[3] Believers Church Medical College Hospital, Tiruvalla, Kerala, India

Keywords Intranasal corticosteroids · Allergic rhinitis · Continuous therapy · Compliance

Introduction

Allergic Rhinitis (AR) is an IgE-mediated inflammatory disease of the nasal mucosa, triggered by exposure to airborne allergens. The primary symptoms of rhinorrhea, nasal block, sneezing and ocular symptoms cause a significant impact on the quality of life of the individual. Although the standard treatment regime starts with avoidance of allergens, pharmacologic agents are used when symptoms persist. Oral antihistamines, intranasal corticosteroids (INCS), combination intranasal therapy of antihistamines and corticosteroids and allergen specific immunotherapy are effective in controlling the symptoms [1]. INCS are the first-line therapy for the treatment of AR [2]. INCS inhibits the early and late-phase allergies in AR by preventing the recruitment of immune cells, and the release of inflammatory mediators from cells involved in the pathophysiology of AR [1].

INCS are prescribed once daily conventionally. The local action of the corticosteroids at the nasal mucosa and its systemic absorption, as the cause of its side effects, have been reported in literature [2]. The decreased consumption of drugs is thought to have decreased side effects. The efficacy of the "as-needed" use of INCS in improving the symptoms of patients is demonstrated in previous studies [3–5]. Clinicians have different modes of therapy to achieve control of clinical symptoms. The present study proposed aims to compare the effect of INCS as a spaced therapy with the conventional daily continuous therapy.

Materials and Methods

This retrospective study was conducted in the Department of Otorhinolaryngology, Believers Church Medical College Hospital, Tiruvalla, Kerala, India. The ethics committee approval has been obtained from the institutional ethical committee. Data on patients who visited our outpatient department with Allergic Rhinitis and who were started on INCS (Fluticasone propionate each puff delivering 50 mcg) were retrospectively obtained from the medical records department of the institution for the duration of the three years (January 2016–January 2019). Variables selected for the study were demographic (age, gender) as well as clinical data (duration and treatment history of allergy along with family history and mode of therapy). In addition to these variables obtained from the case records, Visual Analog Scores (VAS) at the beginning and after three months of therapy and side effects, if any, were also extracted and entered into the data sheet. Participants with complete information in hospital records as per the data sheet requirements of AR were included in the study and those with therapies other than intranasal corticosteroids were excluded.

Mode of therapy for AR:

Clinicians administered INCS either as conventional dose or spaced therapy.

1. In conventional method, once daily dosage was administered.
2. In spaced therapy, patients were started on once daily dosage for a month followed by, once in alternate days for another month, then once in 3 days for a month, and once in 4 days thereafter for long time or as decided by the treating doctor with consultation. If the patient developed an upper respiratory infection during the therapy, daily use of sprays was advised till acute symptoms subsided and then gradually continued with spaced therapy as before.

Patients were divided into two viz., spaced and continuous. The sample size of each group was 91 patients, with a total of 182 patients for spaced and continuous modes of therapy together. The psychometric response scale, the visual analogue score(VAS) was used to monitor the response to the therapy [6, 7]. The VAS is a 10-point scale, where patients were asked to give a score from 0 to 10, based on their individual symptoms of nasal block, rhinorrhea, itching, sneezing with scores of 0 indicating absolute absence of symptoms and 10 indicating the worst symptoms and discomfort possible. The average of these scores were then used for the data entry.

Statistical Analysis

The data obtained were entered into Microsoft Excel sheets and transferred into statistical software (Statistical Package for Social Sciences, Version- 18, Chicago, IL). Descriptive measures on age, gender, years of allergic history, family history and mode of therapy were calculated. Differences in VAS scores (before and after the therapy) were compared for the two groups (Spaced and Continuous) using independent *t*-test. Further, paired *t*-test was used to compare pre- and post-differences for spaced and continuous therapy. A p value of less than 0.05 was considered statistically significant.

Table 1 VAS scores classified according to demographic and clinical variables

	N	Frequency (%)	Mean VAS score before treatment	Mean VAS score after treatment
Age group in years				
< 40	98	53.8	27.05	5.30
40–59	52	28.6	33.58	7.15
> = 60	32	17.6	30.72	6.31
Gender				
Male	103	56.6	29.30	6.13
Female	79	43.4	29.90	5.85
No. of years of allergy				
< = 5 years	32	17.6	27.41	5.38
6–10 years	51	28	28.98	6.16
> 10 years	99	54.4	30.56	6.13
Family history				
Yes	88	48.4	30.98	6.81
No	94	51.6	28.23	5.26

Table 2 Comparison of pre- and post VAS Scores and improvement in VAS scores according to mode of therapy

Mode of therapy According to VAS Scores					
	Frequency	Mean VAS score before treatment	Mean VAS score after treatment	t-value	*p*-value
Spaced	91	29.34	5.70	50.80	0.001
Continuous	91	29.78	6.31	56.96	0.001
**Difference in VAS scores*					
Spaced therapy	91	23.64	4.44	0.265	0.791*
Continuous therapy	91	23.47	3.93		

*indicates the statistical difference of VAS scores between the two groups, expressed as' *p* value'

Results

Totally information on 182 patients was retrieved from the database. 91 patients were on conventional therapy and remaining were on spaced therapy. Among the whole group of patients 103 (56.6%) were males and 79 (43.4%) were females, with a median age of 37 years (range 7–80 years).

88 patients (48.4%) had a family history of allergic rhinitis. 180 (98.9%) patients had taken other modalities of treatment prior to starting intranasal Fluticasone therapy. It was found that 105 (57.7%) patients had sought multiple treatment methods over the years. The previous treatment methods were the use of different tablets at the onset of symptoms by 73 (40.1%) patients, and 2 (1.1%) patients opted for ayurvedic treatment (Table 1).

There was significant improvement in mean VAS Scores for all patients in both continuous and spaced therapy groups ($p = 0.001$). When the difference in VAS scores before and after therapy for the two groups (spaced and continuous therapy) was compared, there was no significant difference in the groups ($p = 0.791$, Table 2).

Discussion

The efficacy of intranasal corticosteroids in controlling symptoms of allergic rhinitis is well established [8]. Intranasal steroids work locally in the nose, are more effective than systemic steroids with its systemic absorption, and thus leading to lesser side effects. INCS are usually recommended at once daily dosage. The onset of action for INCS starts at time points ranging from 3 to 5 h to 60 h after the first dosage [8]. The sensory attributes of aftertaste, nose runout, throat rundown, and smell may be important factors in patient preference and adherence to therapy. The most common side effects of INCSs are a result of local irritation and include dryness, burning, stinging, blood-tinged secretions, epistaxis and rarely septal perforations [8]. Studies have evaluated atrophy of mucosa and squamous metaplasia on continuous use but without conclusive evidence. Effects of INCS on the hypothalamic pituitary axis have also been studied in the past. There have been reports studying effects of INCS on ocular pressure, glaucoma, lens opacity and posterior subcapsular cataract [8]. The benefits however outweigh the risk when used to treat AR. The compliance of the patient to treatment depends on the effects they feel, in terms of improvement in symptoms and other physical changes they experience, after the medications. The awareness of taking steroids and the fear of dependence on the medication often scares the patient and they decide to quit the medicine on their own.

Our patients were mostly males with a negative family history, who had more than ten years of allergy and most of them have already tried some other modality of therapy to achieve symptom control. There was clinical improvement with both spaced and continuous therapy with INCS, without a statistically significant difference. This can be attributed to the efficacy of the drug, which is achieved equally in spaced and continuous therapies.

The results of our study clearly demonstrate that VAS score differences (before and after therapy) were not significantly different for the spaced therapy from continuous mode of therapy. From our results it may be concluded that the spaced therapy is as effective as the conventional mode of therapy in the treatment of AR. With the achievement of similar control of clinical symptoms, spaced therapy may therefore be recommended for better patient compliance and avoidance of the side effects.

In every allergic reaction, there is an early response with the allergen exposure, within minutes where there is mast cell degranulation and histamine release causing sneezing, rhinorrhea and congestion [9] (Fig. 1). There is also a late response, hours later where there is a cellular influx mainly eosinophils and an increase in nasal reactivity to further antigen exposure, called priming, clinically causing congestion and is less dramatic. Intranasal corticosteroids are reported to have profound inhibitory effects to this late response [9]. Kazuba et al.studied the 'as needed' use of

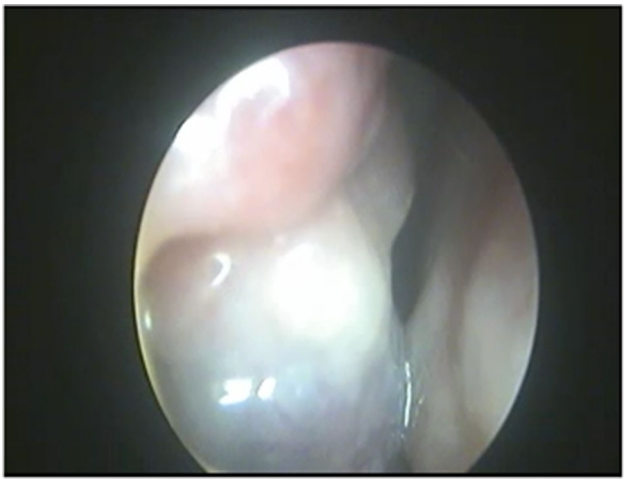

Fig. 1 Endoscopic image of Allergic turbinate hypertrophy

intranasal fluticasone, based on the belief that the allergic individuals who use medications as needed would treat themselves after sensing an early reaction [10]. An intranasal corticosteroid, taken after sensing the symptoms of an immediate response, is thought to block eosinophil infiltration and priming, as reported by Anderson and colleagues in their study [11]. They hypothesized that the 'as needed' use of intranasal corticosteroids would reduce allergic inflammation and provide symptom relief. Juniper et al.compared regular and 'as -needed' usage of aqueous Beclomethasone and found regular usage to be superior. In another study where the quality of life was assessed, the same authors did not find clinically significant degree of improvement for regular use compared to 'as -needed' use [3, 4]. Jen et al.studied the efficacy of 'as-needed' intranasal corticosteroid usage, and compared with 'as-needed' placebo. They studied the symptom diary, eosinophil count and eosinophilic cationic protein level in nasal lavage with both groups and found the quality of life and other parameters were significantly better in the 'as needed' use of INCS [5]. In our study, we postulate the symptom control with spaced usage of steroid. The cellular infiltration of the nasal mucosa and the reactivity of nasal tissue on repeated allergen exposure are thought to be better under check with the spaced usage of corticosteroids.

Continuous use of intranasal corticosteroids is reported to offer many benefits, such as blockage of the early response and reductions of mast cell migration to the epithelium, IgE synthesis, and the number of dendritic cells in the nasal mucosa. Also, the regular use of corticosteroids could be responsible for increased efficacy by providing a quantitatively superior inhibition of eosinophil influx and the priming response related to the higher cumulative dose [10]. By offering spaced therapy as in our study group, we believe the same effects of INCS on the cellular level would be achieved while the unpleasant side effects of the drug could be avoided, thus improving the patient compliance to the medications.

There is an increasing prevalence of allergic diseases in Indian subcontinent and there is interplay of genetic and environmental factors, resulting in this condition. The air pollution, variations with respect to weather, pollens and fungal spores, insects such as cockroaches, parasitic infestations and other living conditions, across India and sparse meteorological data about environmental allergens makes characterisation of the disease difficult. Lack of standardised allergen tests and unreliable epidemiological data limits our understanding of the disease. Clinical management of the patients can be further compromised by knowledge gaps among practitioners, religious beliefs and myths among patients or parents, social stigma of a chronic ailment, and fear of inhalers being addicted to medications [12]. Published evidence from western literature is not directly applicable to Indian population. The improvement in quality of life by intranasal steroids, is established in Indian populations [13]. The spaced steroid therapy helps to give good symptom control with better patient compliance, lower cost and lesser apprehension about side effects of the drug, and thereby preventing the physiologic changes from the continuous therapy.

Our study has several limitations. This is single center study and the sample size is rather small and not randomized and study design is that of a retrospective design. The actual changes at cellular level on usage of INCS were not studied in the laboratory set up. The interplay of multiple factors in our geographical location limits our understanding of time of the allergic trigger. However our experience with the spaced mode of INCS was acceptable to patients and demonstrated similar efficacy to the conventional therapy. A larger group of patients with multiple clinicians in a multicenter set up could validate these results in future.

Conclusion

The efficacy of intranasal fluticasone therapy in controlling symptoms of allergic rhinitis is observed to be similar in spaced therapy, as in continuous therapy. With the achievement of similar control of clinical symptoms, spaced therapy may therefore be recommended for better patient compliance and avoidance of the side effects. It also helps in achieving lesser cost as well as overall improvement in quality of life of allergic patients.

Author contributions All authors contributed to the study conception and design. Conceptualization: IT, TT; Methodology: IT, MA,

KG; Formal analysis and investigation: GM, TT, MA, KVG;Writing—original draft preparation: TT; Writing—review and editing: IT, TT; Resources—IT, TT, MA, KVG; Supervision-IT.

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Declarations

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Hossenbaccus L, Linton S, Garvey S, Ellis AK (2020) Towards definitive management of allergic rhinitis: best use of new and established therapies. Allergy Asthma Clin Immunol 27(16):39. https://doi.org/10.1186/s13223-020-00436-y
2. Wise SK et al (2018) International consensus statement on allergy and rhinology: allergic rhinitis. Int Forum Allergy Rhinol 8:108–352
3. Juniper EF, Guyatt GH, O'Byrne PM, Viveiros M (1990) Aqueous beclomethasone dipropionate nasal spray: regular versus "as required" use in the treatment of seasonal allergic rhinitis. J Allergy Clin Immunol 86:380–386
4. Juniper EF, Guyatt GH, Archer B, Ferrie PJ (1993) Aqueous beclomethasone dipropionate in the treatment of ragweed pollen–induced rhinitis: further exploration of "as needed" use. J Allergy Clin Immunol 92:66–72
5. Jen A, Baroody F, de Tineo M, Haney L, Blair C, Naclerio R (2000) As-needed use of fluticasone propionate nasal spray reduces symptoms of seasonal allergic rhinitis. J Allergy Clin Immunol 105:732–738
6. Klimek L, Bergmann KC, Biedermann T, Bousquet J, Hellings P, Jung K, Merk H, Olze H, Schlenter W, Stock P, Ring J, Wagenmann M, Wehrmann W, Mösges R, Pfaar O (2017) Visual analogue scales (VAS): measuring instruments for the documentation of symptoms and therapy monitoring in cases of allergic rhinitis in everyday health care: position paper of the German Society of Allergology (AeDA) and the German Society of Allergy and Clinical Immunology (DGAKI), ENT Section, in collaboration with the working group on Clinical Immunology, allergology and environmental medicine of the german society of otorhinolaryngology, head and neck surgery. Allergo J Int 26(1):16–24. https://doi.org/10.1007/s40629-016-0006-7
7. Sybilski AJ (2018) Visual analogue scale. A simple tool for daily treatment monitoring in allergic rhinitis. Pediatria I Medycyna Rodzinna-Paediatrics Family Med 14(3):277–81
8. Ganesh V, Banigo A, McMurran A, Shakeel M, Ram B (2017) Does intranasal steroid spray technique affect side effects and compliance? Results of a patient survey. J Laryngol Otol 131(11):991–996. https://doi.org/10.1017/S0022215117002080
9. Naclerio RM, Solomon W (1997) Rhinitis and inhalant allergens. JAMA 1997(278):1842–1848
10. Kaszuba SM, Baroody FM, deTineo M, Haney L, Blair C, Naclerio RM (2001) Superiority of an intranasal corticosteroid compared with an oral antihistamine in the as-needed treatment of seasonal allergic rhinitis. Arch Intern Med 161(21):2581–2587. https://doi.org/10.1001/archinte.161.21.2581 (**PMID: 11718589**)
11. Anderson M, Anderson P, Pipkorn U (1988) Topical glucocorticosteroids and allergen induced increase in nasal reactivity: relationship between treatment time and inhibitory effect. J Allergy Clin Immunol 82:1019–1026
12. Krishna MT, Mahesh PA, Vedanthan PK, Mehta V, Moitra S, Christopher DJ (2020) The burden of allergic diseases in the Indian subcontinent: barriers and challenges. Lancet Glob Health 8(4):e478–e479. https://doi.org/10.1016/S2214-109X(20)30061-9
13. Kumar R, Kumar D, Parakh A (2012) Fluticasone furoate: a new intranasal corticosteroid. J Postgrad Med 58:79–83

Management of Fungal Rhinosinusitis – A Retrospective Study from a Tertiary Hospital in Kerala

Ike Thomas[1], Tina Thomas[2], Reena Anie Jose[3], Elizabeth Joseph[4]

[1, 2, 3, 4] Department of ENT, Believers Church Medical College Hospital, Tiruvalla, Kerala, India.

ABSTRACT

BACKGROUND

Fungal rhinosinusitis (FRS) is an increasingly common pathology in chronic rhinosinusitis and is often diagnosed late. The present study intended at analysing and understanding the clinical, pathological, microbiological, radiological characteristics of the disease in a tertiary hospital in Kerala.

METHODS

Case records of patients with pathologic or microbiologic diagnosis of FRS were retrospectively studied from January 2015 to January 2021, with reference to their demography, clinical presentation, comorbidities, and imaging features. The treatment aspects were also studied.

RESULTS

A total of 36 patients with pathological or microbiologic evidence of FRS were studied. There were 58 % cases of allergic FRS, 33 % cases of fungal ball and 8.3 % cases of invasive FRS. 58 % of patients were females, 77 % patients had nasal block as their presenting symptom, and 66 % of patients had duration of symptoms between 1-6 months. In radiological imaging, the maxillary sinus was most commonly involved. Treatment was always surgical removal. Allergic FRS (AFRS) needed prolonged topical steroids and invasive FRS needed systemic antifungals.

CONCLUSIONS

Our study suggests the importance of early diagnosis of FRS in all chronic rhinosinusitis patients by a high index of clinical suspicion. Tissue samples from the nose and sinuses should be studied for pathology and microbiology in all suspected cases to reach a diagnosis. Radiological imaging can aid in concluding diagnosis. Surgical options, supported by medical management play a vital role in the effective management of the disease.

KEY WORDS

Fungal rhinosinusitis, Allergic fungal rhinosinusitis, Fungal ball, Invasive fungal rhinosinusitis

Corresponding Author:
Dr. Tina Thomas, M. S.,
Consultant ENT Surgeon &
Assistant Professor,
Department of Otorhinolaryngology,
Believers Church Medical College Hospital,
Tiruvalla - 689 103, Kerala, India
E-mail: tinaofficialent@gmail.com

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BACKGROUND

Fungi are increasingly being recognised as a cause of chronic rhinosinusitis (CRS). Being ubiquitous in nature, fungal spores are continuously being inhaled and stored in the respiratory mucosa. Healthy individuals have mostly saprophytic responses but fungi cause disease in certain conditions related to hosting defence.[1] Classification of fungal rhinosinusitis (FRS) has been controversial. The consensus is that they are of two types, invasive and non-invasive. The invasive FRS is thought to be an infective process, whereas it is not clear whether the inflammation in the non-invasive FRS is caused by fungal colonisation or acute infection.[2] Allergic fungal rhinosinusitis (AFRS) was defined as a subset of FRS characterised by Type I allergic reaction to fungi, whereas some studies reported sensitisation to fungi in virtually all cases of CRS when IgE mediated allergy was not observed, thus suggesting 'eosinophilic FRS' as a term for AFRS.[3] FRS is now understood not to be a virtually continuous spectrum of pathology but the disease states are distinct and involve discrete pathologic diagnoses that rarely show a transition from one condition to another.[4] Irrespective of subtype, the diagnosis of FRS is by proving the presence of fungus in the tissue by pathology or microbiology. The diagnosis of FRS is made with clinical suspicion, radiological imaging and proven by microbiological / histopathologic presence of fungus. Treatment is by surgical removal of fungus and inflammatory tissue with systemic antifungal when indicated.

The pathophysiology of the host and environmental factors in the disease progression is not fully understood though the diagnostic methods have improved.[5] The increasing use of broad-spectrum antibiotics, immunosuppressive therapy, immunodeficiency states, cancer chemotherapy and increased use of intensive care interventions are thought to be causative factors. When the presence of fungus within the nose and paranasal sinus starts making pathological changes, the symptoms of nasal obstruction and nasal discharge set in. Patients tend to ignore the symptoms or may find temporary relief with local medical treatment. Polypoidal changes within the nose are treated symptomatically by otolaryngologists with antibiotics, steroids and nasal sprays. The pressure symptoms of FRS on the eye causing eye pain, watering and swelling around the eyes are treated by ophthalmologists with broad-spectrum antibiotics, steroids and antibiotic eye drops. The headache and facial pain get treated by physicians and neurologists with antibiotics and migraine prophylactics. When the symptoms persist and cause a significant effect on their daily life, they seek detailed evaluation. Many of them present to the Department of Otorhinolaryngology with severe morbidities of eye pain, swelling and watering of eyes, severe facial pain and headache, often at the end of several months of the onset of less intense symptoms. Early identification of diagnosis with clinical suspicion and the use of diagnostic methods to initiate prompt treatment would help to bring down the burden of disease. With the increasing number of cases of fungal sinusitis in our CRS patients, this study aimed at understanding FRS in our geographical location.

Objectives

1. To study the demographic, clinical and radiological features across the subsets of FRS.
2. To study the pathological and microbiological features of the disease subtypes.
3. To study the treatment and recurrence pattern of the disease.

METHODS

This was a retrospective review of 36 cases of FRS that were diagnosed at the Department of Otorhinolaryngology, Believers Church Medical College Hospital, Tiruvalla, Kerala, India. This was a descriptive study in design. Ethics committee approval was obtained from the institutional research committee. Data on patients who underwent nasal surgery were collected from January 2015 to January 2021, from the hospital records. Patients diagnosed with fungal sinusitis from the surgical and pathologic findings were considered. Inclusion criteria were cases with histopathology or microbiology proved FRS. The study excluded patients with other types of chronic sinusitis and sinonasal polyposis. The following data were collected:

1. Demographic data - age and gender
2. Presence of comorbidities (Immunocompromised conditions)
3. Clinical picture - presenting symptoms, duration since onset
4. Imaging findings- side involved (right, left, or bilateral), sinus involved (sphenoid, maxillary, frontal, and/or ethmoid), presence of mucosal thickening, complete opacification, increased intrasinus attenuation, sinus expansion, remodelling / wall thinning and the involvement of adjacent soft tissues in each involved sinus
5. Laboratory data - blood eosinophil rate
6. Pathology findings - the presence of eosinophilic mucin, eosinophils, fungal hyphae, and Charcot-Leyden crystals
7. Microbiology findings, KOH mount and fungal culture
8. Management, including therapeutic strategy and postoperative medication (antihistaminic agents, antifungal drops, corticosteroids, and antibiotics).

Statistical Analyses

Statistical analyses of the data were performed. The prevalence of AFS was calculated as the percentage of patients diagnosed and treated as AFS among all cases of chronic rhinosinusitis was surgically treated in the same period. Descriptive statistical tools (Frequency and percentage) were used to describe the demographic and clinical characteristics, as well as the imaging and pathological characteristics.

RESULTS

Altogether there were 172 cases of functional endoscopic sinus surgeries in our hospital from January 2015 to January

2021. We had 36 cases of fungal rhinosinusitis diagnosed by histopathology or microbiology, which corresponded to 20.9 % of our total cohort of chronic sinusitis cases. Of the total patients, 3 (8.3 %) were invasive FRS, where tissue invasion was seen in addition to fungal elements on histopathology. 21 patients (58.3 %) belonged to AFRS, based on the presence of allergic mucin and fungal elements on histopathology with supporting radiologic findings. 12 (33.3 %) cases were a fungal ball, based on fungal elements without allergic mucin, with characteristic radiologic findings.

There were 21 female patients (58.3 %) and 15 (41.7 %) male patients. Our youngest patient was 11 years old and the eldest patient was 74 years, mean age of the patients was 47.9 years. The 21 - 40 age group and the above 60 age groups were mostly affected [Table 1].

Clinical Findings

Most of our FRS patients presented with a nasal block (75 %); the other symptoms were facial pain with headache, nasal discharge, blood-stained discharge, eye pain with swelling, swelling of face localised to sinus locations. Out of 27 patients with nasal block, most of them had AFRS (70.3 %). Localised facial pain and headache were seen in 75 % of patients with the fungal ball. The least common presentation was blood-stained nasal discharge, out of which AFRS was the most common entity. Duration of symptoms ranged from 5 days to many years, mean duration was 3.22 months. History of previous nasal surgery was present in 16.6 % of cases, 3 in the invasive group and 3 in the AFRS group. Among the comorbidities, diabetes was the most common entity, and it was common in the fungal ball group. Peripheral eosinophilia was observed in 4, 100 % cases in the AFRS group and absent in other types.

Radiologic Findings

Deviated nasal septum (DNS) was observed in 78 % of cases. Bilateral involvement was seen in 25 % of AFRS cases. All cases of invasive FRS were unilateral and involved maxillary, ethmoid and frontal sinuses on the affected side. The most commonly involved sinus was the maxillary sinus in all types of fungal sinusitis. Maxillary (95.2 %) and ethmoid (90.4 %) sinuses were affected in AFRS; maxillary sinuses were affected in 91.6 % of cases of the fungal ball. Intrasinus attenuation and mucosal opacification were the most consistent radiological findings on CT in 83.3 % and 86.11 % cases respectively. Remodelling and thickening of the walls of the sinus were seen in 66 .6 % and 71.4 % cases of AFRS. Soft tissue involvement was seen in 66.6 % of cases of invasive FRS and 28.5 % cases of AFRS [Table 1].

Histopathology and Microbiology Findings

Histopathology studies of the 29 cases were available, which showed respiratory epithelium with mixed inflammatory cell infiltrate in all the cases. Allergic mucin was seen in 14 cases of AFRS and 1 case of the fungal ball (Figure 1). Fungal hyphae (thin septate filaments) were demonstrated in 15 cases of AFRS and 7 cases of the fungal ball. In cases of invasive fungal sinusitis, intense inflammatory reaction in mucosa with extensive necrosis was observed. Fungal elements (broad aseptate hyphae) and angioinvasion were evident as well, suggestive of mucormycosis. On microbiology analysis, fungal elements were demonstrated in 23 cases (63.8 %)(Figure 2). Out of 10 samples sent for fungal culture, 4 cases proved fungal growth, all of which showed *Aspergillus flavus* (40 %) (Figure 3,4).

	Invasive FRS (N =3) (8.3 %)	AFRS (N = 21) (58.3 %)	Fungal ball (N = 12) (33.3 %)	Total (N = 36)
Age				
<20 years	0	1	0	1(2.7%)
21-40	0	12(57.1%)	1	13(36.11%)
41-60	3	4	2	9(25%)
>60	0	4	9(75%)	13(36.11%)
Sex				
Male	1	9	5	15(41.66%)
Female	2	12	7	21(58.33%)
Presenting Symptom				
Nasal block	2	19(90.4%)	6	27 (75%)
Facial pain / headache	2	7	9(75%)	18(50%)
Nasal discharge	2	5	4	11(30.5%)
Blood stained discharge	0	2	0	2(5.5%)
Eye symptoms	1	4	1	6(16.6%)
Localised swelling	1	4	2	
Duration of Symptoms				
< 1 month	1	4	4	9(25%)
1-6 months	2	14(66.6%)	8	24(66.6%)
6 months-1 year	0	3	0	3(8.3%)
Co-morbidities				
Diabetes	3	3	5	11(30.5%)
Hypertension	1	4	6	11(30.5%)
Allergy/Asthma			2	2(5.5.%)
Eosinophilia	0	4	0	4(11.1.%)
Radiological features				
DNS	2	18	8	28(77.78%)
Bilateral involvement	0	9	0	9(25%)
Maxillary	3	20(95.2%)	11(91.6%)	34(94.4%)
Ethmoid	3	19(90.4%)	4	26(72.2%)
Sphenoid	2	15(71.4%)	1	18(50%)
Frontal	3	13	0	16(44.4%)
Intrasinus attenuation	3(100%)	17(80.9%)	10	30(83.3%)
Mucosal opacification	3(100%)	17(80.9%)	11	31(86.11%)
Remodelling	3(100%)	14(66.6%)	1	18(50%)
Wall thickening	3(100%)	15(71.4%)	1	19(52.7%)
Soft tissue involvement	2(66.6%)	6(28.5%)	0	8(22.2%)

Table 1. Clinical and Radiologic Features

Management and Post-Operative Follow-Up

All patients received a perioperative short course of antibiotics, analgesics and were started on saline nasal douches on the second postoperative day. Surgery was done for disease clearance and ventilation of the sinuses. Postoperative follow-ups were done in a 10-day interval, where nasal endoscopic cleaning was performed. Steroid nasal douches were started for AFRS patients on their first follow up visit. 14 cases of AFRS were relieved of their symptoms after surgery, 5 were lost after 3-4 follow up visits and 2 had a recurrence of nasal polyps. 10 cases of fungal balls were completely relieved, 2 were lost after 3-4 follow-up visits. 1 case of invasive fungal sinusitis underwent revision surgery and 2 patients received antifungal drugs (IV amphotericin, oral posaconazole) and were on follow up for a

year. 1 patient with invasive FRS died 12 days after the surgery due to other comorbidities.

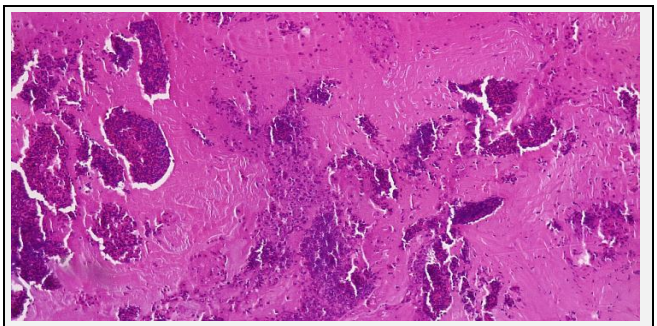

Figure 1. Allergic Mucin Seen in AFRS on Histopathology

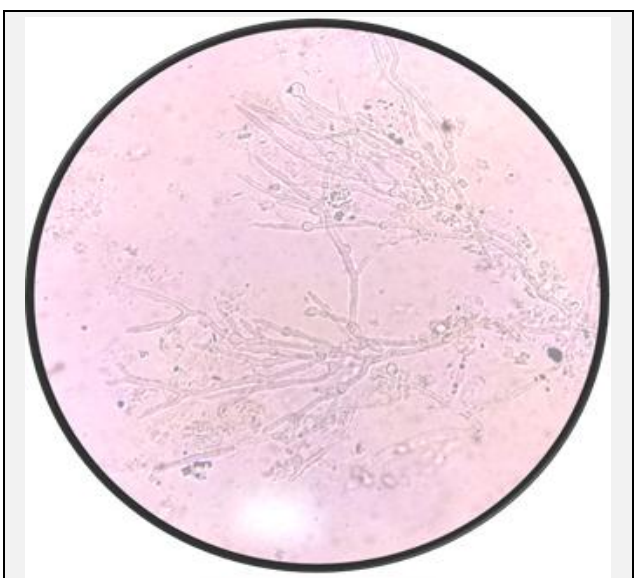

Figure 2. Fungal Filaments in KOH Wet Mount

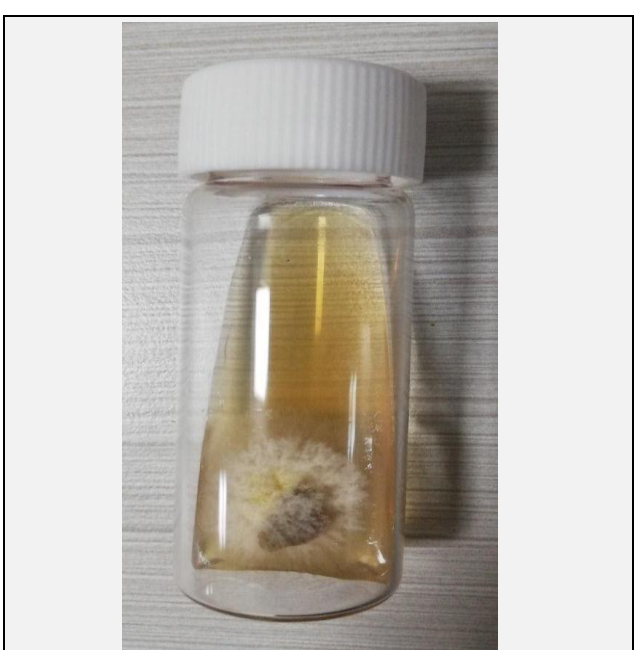

Figure 3. Growth of Aspergillus flavus in Culture in Sabouraud Dextrose Agar

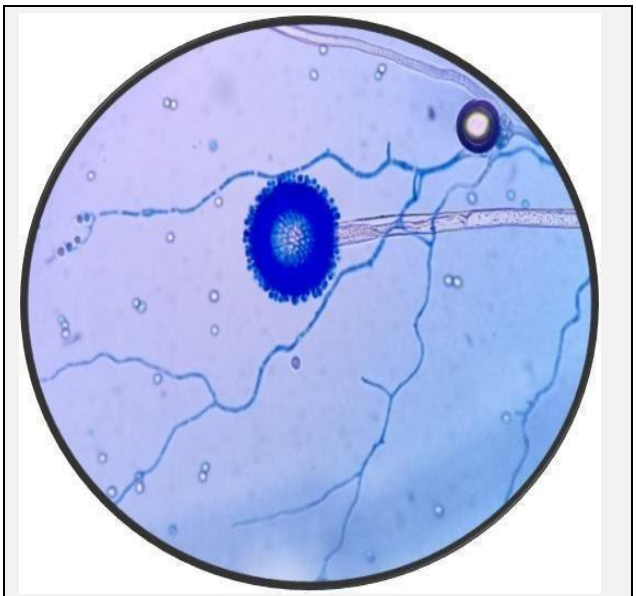

Figure 4: Lactophenol Cotton Blue (LPCB) Mount of Aspergillus Flavus

DISCUSSION

In our patient group of surgical CRS patients, 21 % (36/172) were observed to have FRS. The incidence of FRS reported in our patient group was very well comparable to that reported in the literature. In a recent study from Kerala, there were 26.9 % of FRS cases.[6] In another study from Karnataka, 26 % prevalence of fungal sinusitis by fungal culture among CRS patients were observed.[7] Das et al. at Chandigarh, reported fungal rhinosinusitis with an incidence of 42.7 % of all the 665 cases of chronic rhinosinusitis over 5 years.[8] In a further retrospective study on surgical samples of chronic sinusitis in Singapore*, 8.3 % of chronic sinusitis were fungal in origin.[5] The prevalence of the disease in our study in Kerala, having hot and humid tropical climate and the rich vegetation is self-explanatory.

Most of our patients had allergic FRS (58.3 %), followed by the fungal ball (33.3 %) and invasive FRS (8.3 %) was least common, which compares to other studies reported in the literature. In a study from South India, where FRS was diagnosed by microbiology analysis of 211 samples, 63 % were AFRS, 34 % were invasive and 3 % were fungal granulomas.[9] In the present study, *Aspergillus flavus* was the commonest isolate which was comparable with other studies across the country (6, 7, 9, 10). Other than aspergillus spp., dematiaceous fungi were the commonest reported isolates in AFRS.[9,10]

The classification of FRS has always been a cause for confusion. Bent and Kuhn criteria for AFRS and De Shazo criteria for invasive FRS and fungal balls are widely used to arrive at a diagnosis.[11,12] Though pathological evidence of fungus is diagnostic, clinical and radiologic features are considered as valid criteria for diagnosis. In our study, we considered histopathologic or microbiological proof of fungus at the beginning of the study. Adapting the laid down criteria, samples with tissue invasion were categorised as invasive FRS, samples with the presence of allergic mucin and fungal hyphae and supporting radiological findings were categorised as AFRS, dense fungal hyphae without allergic mucin with its

radiologic findings were included as fungal ball. There were no cases of chronic invasive or granulomatous changes in our pathology studies. AFRS is the most common type of FRS in our study, as in previous studies from India.[13,14]

Most of our patients were females. The age group of 21-40 and above 60 groups were most affected. In a study from Andhra Pradesh, 24.5 % of fungal sinusitis cases belonged to the 20 - 29 years age group,[15] while in a study from Kerala, 36 - 55 years age group were mostly affected.[16] 21 - 40 years were most affected in another study reported from Karnataka.[7] 57 % of our AFRS patients were in the 21- 40 years age group while 75 % of fungal ball patients were in the above 60 age group. AFRS is a disease of adolescents and young adults,[17] where males predominate in children and females among adults. In our study, young female adults were more affected. Most of the patients had nasal block as their presenting symptom. The nasal block was the predominant clinical symptom in similar studies.[7,15] Most of our patients had the persistent nasal block, which was not responding to conventional medical management of CRS. This triggers the physician to evaluate for a fungal element in the pathology of symptoms. Peripheral eosinophilia was noted in 11 % and all of them belonged to the AFRS group. AFRS is understood to be a type I allergic reaction to the presence of fungus in the sinus lumen, where the tissue oedema due to inflammation results in polypoidal changes blocking sinus ostia and CRS.

The most commonly involved sinus on CT imaging in our studies was the maxillary sinus in all types of fungal sinusitis. Intrasinus attenuation and mucosal opacification were the most consistent radiological findings on CT in 83.3 % and 86.11 % cases respectively. Remodelling and thickening of the walls of the sinus were seen in 66 .6 % and 71.4 % cases of AFRS. Soft tissue involvement was seen in 66.6 % cases of invasive FRS and 28.5 % cases of AFRS. Maxillary and ethmoids were mostly affected for AFRS in our study. In AFRS, there was often pansinusitis or bilateral involvement of multiple sinuses, with ethmoid involvement being the most common.[17] There was hyperattenuating and opacification of sinus lumen due to allergic mucin while mucosal linings appeared hypointense, with expansion and remodelling of bony sinus walls.[18] 91.6 % cases of fungal ball involved maxillary sinus. Classical findings in CT demonstrated the involvement of only one paranasal sinus with a hyperintense (‘‘metal-dense’’) spot at the centre of the fungus ball, often with sclerosis of the adjacent bone, taking contour of the sinus lumen.[18] Our CT findings were consistent with other reports existing in the literature.

Histopathology of fungal sinusitis in our study showed allergic mucin and fungal hyphae in 51.7 % and 75.8 % cases. Bharadwaj et al. in their study reported the presence of allergic mucin in 38 % of cases who underwent surgery.[19] Das et al. reported in their histopathology study 56.3 % AFRS, 3.9 % fungal ball, 16.9 % chronic granulomatous FRS, 1.4 % chronic invasive FRS, 17.2 % acute fulminant FRS and also 4.25 % mixed histopathological patterns.[8] Chronic granulomatous or chronic invasive FRS were not observed in our patient cohort.

FRS is a surgically treated disease and there was a relief of symptoms in 69.4 % of cases. Saline nasal douches were part of routine post-surgical care. Steroid douches were started in all AFRS cases as the disease is primarily an allergic reaction of the tissue to the presence of fungus. It is needed for prolonged durations in AFRS patients as fungi are always present in their environment and patients are sensitized already. Recurrence of symptoms is known and can be managed medically with steroid douches and endoscopic cleaning. Revision surgery may also be needed. The fungal ball gets treated completely with clearance of the dense fungus mass from the nose and sinus. Invasive FRS often needs repeated surgeries for debridement of necrotic tissue, along with systemic antifungals. In a study from Singapore, there was 13.6 % recurrence when followed up to 60 months.[5]

CONCLUSIONS

There is an increasingly high incidence of FRS among CRS patients. AFRS was common among the subtypes. Nasal block was the commonest presentation. This is always a surgically treated disease. The identification of the presence of fungus by histopathology or microbiology is always indicated and mandatory to categorise the disease. Samples should be sent from all suspected cases. Radiological imaging can aid in concluding diagnosis. AFRS needs surgical clearance and a prolonged post-op topical steroid, recurrence was seen. The fungal ball is completely treated by surgical removal of the fungus while invasive FRS needs surgical debridement and systemic antifungals.

Limitations of the Study

This article reports our experience of management of FRS and our study has several limitations. The data is from a single centre and the sample size is rather small and the study design is retrospective. We were not able to determine the presence of atopy, which is one of the crucial criteria for the diagnosis of AFRS. Laboratory investigations were not routinely ordered for all patients, there was a lack of data in all the parameters. Clinicians were probably not aware of the importance of sending tissue samples for pathology and microbiology analysis, not to miss a diagnosis. Despite all these limitations, we strongly believe that the analysis of the data helped us to understand the pathology involved and its incidence and diagnosis better and manage these groups of patients effectively.

Data sharing statement provided by the authors is available with the full text of this article at jemds.com.
Financial or other competing interests: None.
Disclosure forms provided by the authors are available with the full text of this article at jemds.com.

REFERENCES

[1] Patrascu E, Manea C, Sarafoleanu C. Difficulties in the diagnosis of fungal rhinosinusitis – literature review. Romanian Journal of Rhinology 2016;6(21):11-17.
[2] Meltzer EO, Hamilos DL, Hadley JA, et al. Rhinosinusitis: establishing definitions for clinical research and patient care. J Allergy Clin Immunol 2004;114(6 Suppl):155-212.

[3] Ponikau JU, Sherris DA, Kita H, et al. Intranasal antifungal treatment in 51 patients with chronic rhinosinusitis. J Allergy Clin Immunol 2002;110(6):862-6.
[4] Adelson RT, Marple BF. Fungal rhinosinusitis: state-of-the-art diagnosis and treatment. J Otolaryngol 2005;(34 Suppl 1):S18-23.
[5] Alshaikh NA, Alshiha KS, Yeak S, et al. Fungal rhinosinusitis: prevalence and spectrum in Singapore. Cureus 2020;12(4):e7587.
[6] Archana, Sindhu VN, Salima RW. Chronic rhinosinusitis-fungal etiology & clinical, histopathological, radiological profile in a tertiary care centre. Journal of Medical Science and Clinical Research 2018;6(9):863-8.
[7] Syiemlieh B, Mariraj J. Study on chronic rhinosinusitis: a clinico-mycological perspective in a tertiary care centre. International Journal of Current Microbiology and Applied Sciences 2020;9(7):3740-7.
[8] Das A, Bal A, Chakrabarti A, et al. Spectrum of fungal rhinosinusitis: Histopathologist's perspective. Histopathology 2009;54(7):854-9.
[9] Michael RC, Michael JS, Ashbee RH, et al. Mycological profile of fungal sinusitis: an audit of specimens over a 7-year period in a tertiary care hospital in Tamil Nadu. Indian Journal of Pathology and Microbiology 2008;51(4):493-6.
[10] Kaur R, Lavanya S, Khurana N, et al. Allergic fungal rhinosinusitis: a study in a tertiary care hospital in India. Journal of Allergy 2016;2016:7698173.
[11] Bent JP 3rd, Kuhn FA. Diagnosis of allergic fungal sinusitis. Otolaryngol Head Neck Surg 1994;111(5):580-8.
[12] deShazo RD, O'Brien M, Chapin K, et al. A new classification and diagnostic criteria for invasive fungal sinusitis. Arch Otolaryngol Head Neck Surg 1997;123(11):1181-8.
[13] Munjal M, Gupta V, Chinna D, et al. Demographic profile of fungal rhinosinusitis at a tertiary care centre. Int J Otorhinolaryngol Head Neck Surg 2021;7(1):61-5.
[14] Chakrabarti A, Rudramurthy SM, Panda N, et al. Epidemiology of chronic fungal rhinosinusitis in rural India. Mycoses 2015;58(5):294-302.
[15] Waghray J. Clinical study of fungal sinusitis. Int J Otorhinolaryngol Head Neck Surg 2018;4(5):1307-12.
[16] Santhi T, Rajan KV. Presence of fungal organisms in chronic rhinosinusitis with nasal polyposis: a clinico pathological study from Kerala. Int J Sci Stud 2015;3(3):99-104.
[17] Al-Dousary SH. Allergic fungal sinusitis: radiological and microbiological features of 59 cases. Ann Saudi Med 2008;28(1):17-21.
[18] Pillai S, Bhandarkar AM, Nair SS, et al. A radiological profile of fungal sinusitis. Asian Journal of Pharmaceutical and Clinical Research 2017;10(9):331-4.
[19] Bhardwaj BB, Gill JS, Tanya S. The role of CT scan and histopathology in diagnosis of allergic fungal sinusitis. Clinical Rhinology an International Journal 2014;7(3):117-20.

A Novel Technique of Using Sponge as Post-Operative Nasal Packing

Ike Thomas,[1] Jathin Sam Thekkethil,[1] Ramesh Chandra Kapoor,[2] Tina Thomas,[1] Pramod Thomas[1]

ABSTRACT

Introduction

In an effort to find a comparable but less expensive nasal pack with the qualities of Merocel®,this study was aimed at comparing the clinical efficacy and patient comfort level, while simultaneously using Merocel® and commercially available sponge as packing material in the same patient.

Materials and Methods

This study included those patients who underwent septoplasty, turbinoplasty or FESS and nasal packing was done randomly with Merocel® and commercially available sponge (polyurethane foam) on the same patient. Patients shared their experience on the symptom questionnaire on the first post-operative day and they underwent sequential diagnostic nasal endoscopy to assess the endoscopic status of the nasal cavity, which were documented meticulously.

Result

The post-operative bleeding control, pain during pack removal, general satisfaction, willingness to reuse and post-operative adhesion were same for both Merocel® and sponge.

Conclusion

The innovative technique of using a commonly available, commercially prepared sponge which is as good as Merocel® is well supported due to its efficacy in hemostasis, less mucosal trauma and less pain during pack removal. So it may be used in developing countries where cost is a factor for compliance of patients for undergoing surgeries without compromising on quality.

Keywords

Nasal Packing; Polyvinyl Alcohol; Polyurethanes

Nasal Packing is one of the most common procedures done by Otorhinolaryngologists worldwide. It is often done after septoplasty and it aims at preventing postoperative bleeding, septal hematoma or nasal synechiae, ensuring mucoperichondrium flap to be in position and cartilage stabilization in order to get the best surgical results.[1]

It is done after Functional Endoscopic Sinus Surgery (FESS) to prevent postoperative bleeding. The ideal packs are easy to insert and remove without causing pain and discomfort. A wide variety of nasal packing materials are available in the market. Use of nasal packs vary in different countries.[2] It may even vary in different places or institutions. Generally used nasal packing materials are antibiotic cream coated ribbon-gauze packs, custom made glove packs, Merocel® and the newer additions are Rapidrhino® and biodegradable nasal packs like Nasopore®. Most of the patients feel pain, pressure and discomfort while packing and on removing the pack on the first post-operative day.[3] Merocel® is a foam type non-absorbable nasal packing material which is a cross-linked polyvinyl alcohol, which is commonly used nowadays. It is equally effective for haemostasis and less traumatic to the operated nasal mucosa, but it is expensive when compared with ribbon-gauze and glove packs.

Nowadays, there is an increasing trend of avoiding nasal packs for better patient comfort. Our effort is to find a comparable nasal pack which has the qualities of Merocel® which is less expensive. Commercially produced Sponge (Polyurethane foam) is one such material, which is commonly available, less expensive and can be cut into specific sizes, autoclaved and used

1 - Believers Church Medical College Hospital, Thiruvalla, Kerala
2 - Badr Al Samaa Hospital, Ruwi

Corresponding author:
Dr Jathin Sam Thekkethil
email: jathinsam@gmail.com

as nasal pack.This study was aimed at comparing the clinical efficacy and patient comfort level, while simultaneously using Merocel® and commercially available sponge as packing material in either nasal cavity in the same patient.

Materials and Methods

This prospective randomized clinical study was conducted from July 2009 to June 2014. The study protocol was approved by the institutional ethical committee. The authors asserts that all procedures contributing to this work comply with the ethical standards of the relevant national and institutional guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008. The patients included in the study were counselled in detail regarding the use of two different nasal packs and their properties and a detailed informed consent was taken.

This study included patients age ranging from 17 years to 58 years, with chronic moderate to severe rhinosinusitis, septal deviations and turbinate hypertrophy who were planned for septoplasty surgery, turbinoplasty surgery or Functional Endoscopic Sinus Surgery (FESS) under general anaesthesia. The exclusion criteria for the study was revision FESS, patients with bleeding diathesis, patients on aspirin or anti-platelet drugs and hypertensive patients. Prior to the surgery, all patients underwent routine ENT examination and diagnostic nasal endoscopy. Computed tomography evaluation of paranasal sinuses were done for FESS and septoplasty patients. Patients were explained regarding the symptom questionnaire prior to their participation in the study. The patients included in the study were unaware of the type of nasal pack given in either nasal cavity.

The operating surgeon selected each side of the nose randomly for packing with Merocel® or Sponge without any bias. The size of the Merocel® was standard (custom made to 4cm along with a draw string used for removal) and the size of the commercially available sponge was cut and made into a size of 4cm long, 1 cm breadth and 1.5 cm height, which was then autoclaved and used for nasal packing(Fig. 1).

All the patients were provided with oral antibiotics from the day of surgery for a period of one week and intranasal saline douches after pack removal from the first postoperative day. Patients shared their experience on the symptom questionnaire on the first post-operative day, after removal of the packs on both sides. The questionnaire was based on the standard Visual Analog Scale (VAS), wherein score score '0' meant no symptom and score '10' meant unbearable symptoms. The VAS questionnaire evaluated the various aspects of patient comfort with respect to pain and pressure during removal of pack, sleep disturbance and general satisfaction where the patients willingness to use the material in future for any other nasal surgeries.

The patients underwent sequential diagnostic nasal endoscopy on the 10th day and 30th day to assess the endoscopic status of the nasal cavity with regard to mucosal injury and healing status, adhesion, synechiae, infection and granulation. A grading scale from 0 to 3 was created for assessment of the severity of each of the above signs and for the assessment of bleeding in the immediate post-op, which were documented meticulously. The various proportions were compared using chi-square test statistic. The statistical analysis of was done by using the statistical software R.

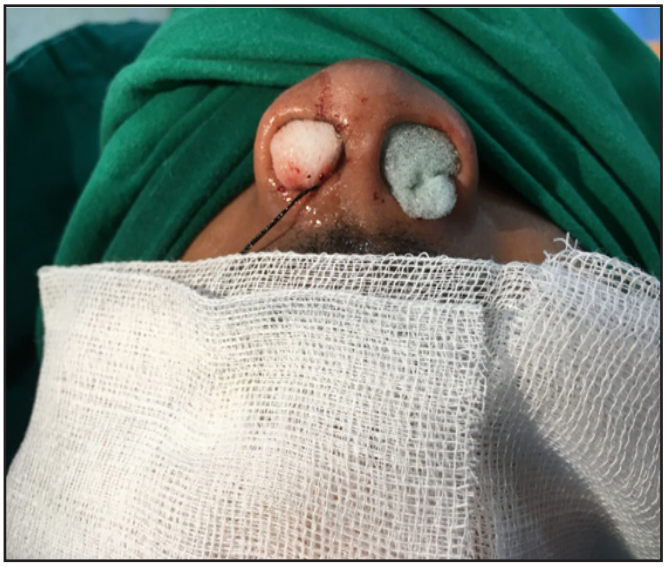

Fig. 1. Nasal packing with Merocel^{R} (with string attached to it) on one side and the other side packed with autoclaved sponge

Result

In our study, a total of 50 patients were considered, out of which 5(10%) patients were less than 20 years of age, 36(72%) patients aged between 21 years to 40 years and 9(18%) patients above 40 years. In this, 31 (62%) patients were males and 19 (38%) patients were female. Out of these 50 patients, 21 (42%) patients underwent septoplasty, 13 (26%) patients had FESS and 16 (32%) patients underwent turbinoplasty.

The first observation which was made during the study was regarding the hemostatic property or the bleeding control in the immediate postoperative period. The hemostatic property between the two materials in the immediate postoperative period was statistically similar. With regard to the hemostatic property, 41/50 patients (82%) had excellent bleeding control on the sponge pack side with no bleeding and 3/50 patients (6%) had minimal bleeding, where there was minimal filling up of blood within the nasal cavity which did not flow out of the nostril and did not require repacking (TableI). 4/50 patients (8%) on sponge pack side had moderate bleeding, 2/50 patients (4%) had severe bleeding and required repacking. The results on the sponge pack side were as comparable to the side with Merocel® pack,where 42/50 patients (84%) had excellent hemostsis with no bleeding and 4/50 patients (8%) had minimal bleeding, 3/50 patients (6%)with moderate bleeding and 1/50 patients (2%) had severe bleeding which required repacking. We did not face any secondary haemorrhage in any of the patients due to infections, on either side.

On analysing the data of the pain experienced by the patient during the time of pack removal, there was no statistical difference between the Merocel® side and the sponge side. On the Merocel® side, 27 (54%) had no pain to mild pain and 23 (46%) had moderate to severe pain (Table II). On the other hand, on the sponge side, 40 (80%) had no pain to mild pain and 10 (20%) had moderate to severe pain (Table II). Most notable finding is that 6 patients in the Merocel® side had severe pain while removing the nasal pack but none of the patients complained of severe pain on the sponge side, which may be due to the more rigid structure of the Merocel® in comparison with the sponge.

On evaluation of the general satisfaction of the patients regarding reusing the packing material in the future for any nasal surgeries and for recommending any specific packing material for friends, relatives or for others, there was no statistical difference between Merocel® and sponge group. On considering Merocel®, 34 (68%) patients were willing to reuse the material but 16 (32%) patients were not willing to reuse. On considering sponge side, 46 (92%) patients were willing to reuse sponge as packing material in future but 4 (8%) patients were not willing to reuse (Table III).

In this evaluation, even though it is statistically not significant, one of the notable finding is that, out of the 16 patients who were not willing to reuse Merocel® as packing material in future, 15 (93.8%) are willing to

Table I: Grading of Postoperative bleeding control/Hemostasis

BLEEDING CONTROL	MEROCEL® (N=50)	SPONGE (N=50)
No bleeding	42	41
Minimal bleeding	4	3
Moderate bleeding	3	4
Severe bleeding	1	2

*p = 0.686

Table II: Grading of pain during removal of nasal pack

PAIN DURING REMOVAL	MEROCEL® (N=50)	SPONGE (N=50)
No pain	1	4
Mild pain	26	36
Moderate pain	17	10
Severe pain	6	0

*p = 0.089

Table III: General Satisfaction of patients

GENERAL	MEROCEL® (N=50)	SPONGE (N=50)
Willingness to reuse	34	46
Not willing to reuse	16	4

***p = 0.617**

reuse sponge as packing material (Table IV).

On analysing the post-operativeadhesions and synechiaeafter 10 days of pack removal, no statistically significant difference seen (*p=0.314).

On the Merocel® side, 35 (70%) had no adhesions and 15 (30%) had adhesions. On the sponge side, 39 (78%) had no adhesions and 11 (22%) had adhesions. All the adhesions in both groups were mild and easy to release and done as OPD procedure. No adhesions or synechiae seen on any side on the 30th day nasal endoscopy.

Discussion

Chronic rhinosinusitis (CRS), septal deviation, and inferior turbinate hypertrophy are among the most common diseases seen in the ENT department in current practice. These conditions are present in patients of all ages and both genders. Surgical procedures such as functional endoscopic sinus surgery (FESS), septoplasty and turbinoplasty, are often considered when medical treatments have failed. At the end of each of these procedures, nasal packs are placed into the nasal cavities to prevent bleeding of the wound.

Generally nasal packings include removable nasal packs like antibiotic cream coatedribbon-gauze packs, custom made glove pack,Merocel® etc. and recently introduced biodegradable nasal packing materials like Nasopore[R]. FESS and other nasal surgeries are constantly evolving and it makes otorhinolaryngologists to create modifications in the nasal packs.

The innovations in nasal packing were motivated by the innate defects of conventional packing materials in quality of life during early postoperative period and the pain tolerated during nasal pack removal.

The postoperative treatment regimen of FESS is as important as the surgery itself, since the ultimate goal is to re-establish normal mucociliary clearance in the sinuses. So the nasal packs were expected to improve mucosal healing and avoid adhesion of mucosa in the nasal cavity. But, the foremost use of nasal pack is to control bleeding after sinus or septal surgery. Hence, many packing materials were time tested and proven, and some still being in the evaluation phase.

The use of removable nasal packs like antibiotic cream coated ribbon-gauze packs, glove packs and

Table IV: Comparison of General satisfaction of patients on using MerocelR and sponge as packing material

		SPONGE		TOTAL
		WILLING TO REUSE	NOT WILLING TO REUSE	
Merocel®	Willing to Reuse	31 -91.2%	3 -8.80%	34 -100%
	Not Willing to Reuse	15 -93.8%	1 -6.3%	16 -100%
Total		46 -92%	4 -8%	50 -100%

***p = 0.617**

Merocel® are widely used worldwide. The advantages for Merocel® nasal pack includes easy manipulation and alignment within the nasal cavity, and provide better supporting ability.[4] But they have some disadvantages also. They are costlier than the ribbon- gauze pack or glove packs which are made in the hospital itself.[5]

Pain and pressure present when the pack is inside the nose and during removal are the common complaints of patients with Merocel® pack, which often decreases the quality of life of patients after nasal surgeries.[1] Some patients have mentioned that the removal of packing material was the most painful experience in the whole of their life.[6] Biodegradable nasal packs does not require the removal, as it gets absorbed inside the nose and thereby avoiding the pain during the pack removal. But, it increases the cost of nasal packs further. Hence, we developed the innovative idea of reducing the cost of nasal packs by using a material which is commonly available in the operation theatre and hospitals, and it is as good as other nasal packs.

Commercially produced Sponge (Polyurethane foam) is one such material. It is freely available in the operation theatre, which can be cut into specific sizes, autoclaved and used as packing material. Today's polyurethanes have been formulated to provide good biocompatibility, flexural endurance, high strength and high abrasion resistance. These attributes are important in supporting new applications of sponge by medical device manufacturers including artificial hearts, catheter tubing, feeding tubes, surgical drains, intra-aortic balloon pumps, dialysis devices, non-allergenic gloves, medical garments, hospital bedding, wound dressings and more.

Akita et al studied the benefits of polyurethane in split thickness skin graft donor wound healing, and they found out that the polyurethane dressing was superior to hydrogel in the wound healing time, amount of exudates, and frequency of dressing changes.[7] Handel N et al studied the long term safety and efficacy of polyurethane foam covered breast implants and the result showed that the incidence of capsular contracture was dramatically lower with polyurethane foam-covered implants compared to smooth or mechanically textured implants; this beneficial effect persisted at least 10 years after implantation.[8] These studies proved that the sponge can be used inside the body with long term durability without any complications. Long-term in vitro durability of polyurethane heart valves has been achieved and polyurethane valves manufactured from a commercially available textile polyurethane were capable of achieving more than 800 million cycles in laboratory fatigue testing (equivalent to more than 20 years of normal function).[9]

In our study, 46% patients had moderate to severe pain while removing the pack on the Merocel®side while only 20% patients had moderate to severe pain. Even though it is not statistically significant, the most notable finding is that, 6 patients in the Merocel® side had severe pain while removing the pack but none of the patients complained of severe pain on the sponge side, which may be due to the more rigid structure of the Merocel® in comparison with the sponge. Many studies compared the pain during removal of various nasal packs with Merocel®. Hesham et al reported that Rapidrhino® packs were less painful than Merocel® packs.[10] In our study, pain during pack removal ofMerocel® and sponge are statistically similar.

Likewise, sponge had comparable hemostatic property and post-operative adhesion as that of the Merocel® in our study with no statistical difference between the two. Ragunandhan et al reported that 86.6% Merocel® pack patients provided excellent hemostasis with no bleeding. In our study, 84% Merocel® pack and 82% sponge pack patient had excellent hemostasis with no bleeding.[3] Hence, the hemostatic property of sponge was at par with Merocel®. Yilmaz et al stated that adhesion developed in 7 (28%) patients in the Merocel® group in the 4-week follow-up.[11] In our study it was 30% and all the adhesion were mild and easy to release and did not proceed to become a synechiae.

While considering the general satisfaction of patients in reusing the packing material in future 68% patients were willing to reuse Merocel® and 92% patients were willing to reuse sponge. Out of total number of patients who are not willing to reuse Merocel® in future, 93.8% patients are willing to reuse sponge instead of Merocel®. Raghunandhan et al reported that the general satisfaction and willingness to reuse Nasopore^{R} was significantly high on comparison with Merocel®.[3] In our study, the general willingness to reuse sponge as

a packing material is at par with Merocel® and many patients prefer to use sponge instead of Merocel®, which is in accordance with the above quoted study. Hence, sponge is comparable to Merocel®.

Conclusion

Merocel® is a novel packing material which is used by most ENT surgeons after nasal surgeries due to its clinical efficiency in hemostasis and less trauma to nasal mucosa comparing to ribbon-gauze packing, but the disadvantages are the cost of Merocel® and the pain during removal of pack.

Even though 'no-packing technique' is there, but many ENT surgeons are not practicing it and still sticking on to the age old concept of nasal packing.

Biodegradable nasal packs are rejected due to its significant cost difference against other routine nasal packs. Hence, our innovative technique of using a commonly available, commercially prepared sponge (polyurethane foam) is well supported due to its efficacy in hemostasis, less mucosal trauma and less pain during pack removal.

The efficacy of sponge is as comparable to Merocel®, which is a time tested packing material which is already in use. Also the cost of sponge is less, as it is freely available in hospitals which can be autoclaved and used.

So sponge may be considered as an alternative packing material in developing countries where cost is a factor for compliance of patients for undergoing surgeries without compromising on quality.

References

1. Dubin MR, Pletcher SD. Postoperative packing after septoplasty: is it necessary? Otolaryngol Clin North Am. 2009; 42(2): 279-85
2. Rainer K. Weber. Nasal packing and stenting. GMS Curr Top Otorhinolaryngol Head Neck Surg. 2009; 8
3. Raghunathan S, Kameswaran M, Thomas JK. A prospective double blinded randomized controlled study comparing the efficacy of a novel biodegradable synthectic polyurethane foam (Nasopore) vs Standard polyvinyl acetate sponge (Merocel) as packing material after functional endoscopic sinus surgery: The first Indian experience. Clin Rhinol: Int J. 2014; 7(3): 105-11
4. Breshman M, Mehigan B, Curran A. An evaluation of merocel and series of 5000 nasal packs in patients following nasal surgery: A prospective randomized trial. Clin Otolaryngol. 2007; 32: 352-5
5. Dutta S, Mukherjee A, Saha J, Biswas G, Haldar D, Sen I, Sinha R. Modified Technique of anterior nasal packing: A comparative study report. Indian J Otolaryngol Head Neck Surg. 2012; 64 (4): 341-5
6. von Schoenberg M, Robinson P, Ryan R. Nasal packing after routine nasal surgery – Is it justified? J Laryngol Otol. 1993; 107(10): 902-5
7. Akita S, Akino K, Imaizumi T, Tanaka K, Anraku K, Yano H, Hirano A. A polyurethane dressing is beneficial for split thickness skin graft donor wound healing. Burns 2006; 32(4): 447-51
8. Handel N, Gutierrez J. Long term safety and efficacy of polyurethane foam-covered breast implants. Aesthet Surg J. 2006; 26(3): 265-74
9. Wheatley DJ, Raco L, Bernacca GM, Sim I, Belcher PR, Boyd JS. Polyurethane: material for the next generation of heart valve prostheses? European J Cardio thoracic Surg. 2000; 17: 440-8
10. Hesham A, Ghali A. Rapidrhino versus merocel nasal packs in septal surgery. J Laryngol Otol. 2011; 125(12): 1244-6
11. Yilmaz MS, Guven M, Elicora SS, Kaymaz R. An evaluation of biodegradable synthetic polyurethane foam in patients following septoplasty: A prospective randomized trial. Otolaryngol Head Neck Sur. 2013; 148(1): 140-4.

ENDOSCOPIC DACRYOCYSTORHINOSTOMY [DCR] WITH MEROCEL PACK IN THE LACRIMAL SAC- AN INNOVATIVE TECHNIQUE

Ike Thomas[1], Pramod Thomas[2], R. C. Kapoor[3]

[1]Assistant Professor, Department of ENT, Believer's Church Medical College, Thiruvalla, Kerala, India.
[2]Associate Professor, Department of Community Medicine, Believer's Church Medical College, Thiruvalla, Kerala, India.
[3]Consultant, Badr Al-Sama Hospital, Oman.

ABSTRACT

BACKGROUND

DCR is a surgical procedure performed to create a new tear drainage pathway between the eye and the nose when the tear drain becomes obstructed at the level of nasolacrimal duct, which is the commonest site of blockage in lacrimal apparatus. This technique of DCR with Merocel pack in the lacrimal sac completely avoids any complications like lacrimal pump failure, canthal erosion, canthal cheese wiring, although few patients exhibit complications like granuloma, synechiae/ adhesions and foreign body sensation inside the nose for few days. Patients are relieved of epiphora/ mucocele with less operative time and post-operative care.

The objective of the study is to describe the various complications, which can occur in endoscopic DCR procedure.

MATERIALS AND METHODS

It is a descriptive study. We enrolled 30 participants with nasolacrimal duct obstruction presenting with epiphora, mucocele and palpable lacrimal sac and were managed with endoscopic DCR with a 'Merocel pack in the sac technique.' The present study was conducted at the Ministry of Health Hospital, Rustaq, Sultanate of Oman, during 2007 to 2014. The sample size was taken for convenience during the study.

RESULTS

Results were compared with standard external DCR and endoscopic DCR with silicone stents. All patients were operated under general anaesthesia. Participants were followed for 6 months and outcomes (Absence of epiphora and the patency of sac) and complications (lacrimal pump failure, canthal erosion, canthal cheese wiring granuloma, synechiae/ adhesion and foreign body sensation) were measured. Lacrimal sac was opened and the Merocel pack was kept in the sac (1 cm x 0.5 cm) for 10 days and then removed. The present innovative technique gave good results (p-value < 0.0001), took less post-operative time and fewer post-operative complications.

CONCLUSION

Endoscopic DCR with 'Merocel pack in the sac technique' is an excellent novel technique for the treatment of epiphora and mucocele due to nasolacrimal duct obstruction. It should be the treatment of choice in view of maintenance of long-term results, patient compliance and cost effectiveness.

KEY WORDS

Dacryocystorhinostomy, Merocel Pack, Sac.

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BACKGROUND

Dacryocystorhinostomy [DCR] is the surgical procedure performed while patients present with epiphora, mucocele and palpable lacrimal sac. Tear drain becomes obstructed at the level of nasolacrimal duct (commonest site of blockage in lacrimal apparatus), DCR will create a new tear drainage pathway between the eye and the nose.

Nasolacrimal duct obstruction occurs either congenitally or from various acquired causes like facial trauma, chronic environmental allergies, toxicity from chemotherapeutic drugs, topical medications, long-standing sinus disease or following sinonasal surgery.[1,2] Dacryocystitis mostly results from descending inflammation from the eye or ascending inflammation from the nose.[2] Repeated occurrence of inflammatory reactions ends up with structural, epithelial and subepithelial changes leading to total fibrous closure of lumen.[2] DCR can be performed externally or endoscopically through nose. History of DCR stretches back nearly 2000 years. It was Celsus who described about DCR in first century.[3] Galen described about DCR in second century. Toti described about external DCR in 1904.[4] It was Caldwell who described transnasal DCR in 1983.[5] McDonogh and Meiring described the endoscopic DCR in 1989.[6] With the advent of endoscopes and audio-visual aids, endoscopic DCR has gained much popularity.

DCR can be done externally or endoscopically with or without using silicone stent. Endoscopic DCR has many advantages over external DCR like avoiding scar over the face, avoiding the need of division of medial canthal ligament and preservation of orbicularis oculi muscle pump action of the lacrimal sac.[1,2] Endoscopic DCR also enables correction

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Corresponding Author:
Dr. Ike Thomas,
Kizhake Veedu, Pamayely P. O.
Kottarakara-691532, Kollam,
Kerala, India.
E-mail: drikethomas@gmail.com
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of associated pathology like deviated nasal septum, chronic rhinosinusitis and polyps at the same time.[2] Pyoceles and acute infection of nasolacrimal duct with or without skin involvement can be drained safely endoscopically, thereby reducing risk of intracranial extension.[2] Closure of the rhinostomy opening was considered a major factor of failure of external DCR, while fibrosis of cutting edges of medial wall of lacrimal sac and reposition of nasal mucosa are common causes of failure of endoscopic DCR without stent.[1] With stents canthal erosion and canthal cheese wiring are seen.[1] Our technique of keeping Merocel pack in the sac for 10 days after opening the sac and drainage of pus is an innovative technique with patent drainage of tear into the nose. This technique of keeping Merocel pack in the lacrimal sac for 10 days heals the cut margins of the sac, hence prevents fibrosis and blockage of cut ends of the sac. Since there is no stents used, the ends of the stent will not irritate nasal mucosa which leads to synechiae formation, adhesion, foreign body sensation, granuloma formation and epistaxis, especially if the stent is kept inside the nose for long time. The minor complications seen in external DCR such as conjunctival haematoma, trauma to eyelids and periorbital oedema are not seen in our technique.

Objective of the Study

To describe the various complications, which can occur in endoscopic DCR procedure.

MATERIALS AND METHODS

It is a descriptive study. We enrolled 30 participants with nasolacrimal duct obstruction presenting with epiphora, mucocele and palpable lacrimal sac and were managed with endoscopic DCR with a 'Merocel pack in the lacrimal sac technique.' The present study was conducted at the Ministry of Health Hospital, Rustaq, Sultanate of Oman, during 2007 to 2014. The sample size was taken for convenience during the study. Participants approached the hospital with chronic dacryocystitis (Epiphora, mucocele, palpable lacrimal sac) were enrolled for endoscopic DCR after screening for the distal nasolacrimal duct obstruction. Those who enrolled were counselled regarding the use of Merocel packs and detailed informed consent was taken. Data on Age, Gender, Side operated and various complications present (lacrimal pump failure, canthal erosion, canthal cheese wiring, granuloma, synechiae/ adhesion and foreign body sensation) were noted at each visit. The patients were followed up on 10th day for Merocel pack removal, 3 weeks, 3 months, 6 months and 1 year post-operatively with nasal endoscopy. If there was any synechiae adhesion or granuloma formation seen in any of the visits, they were tackled in the outpatient department endoscopically.

Surgical Technique

All patients were operated under general anaesthesia, local decongestion of the nasal mucosa and middle turbinate was done with 4% Lignocaine with adrenaline solution soaked patties. The site for elevation of the flap was ascertained with the help of bayonet forceps, one prong was kept on the lateral nasal wall outside the distended sac area and the other prong on the mucosal surface of the lateral nasal wall. The area identified was infiltrated with 2% lignocaine with adrenaline. Following decongestion, the mucoperiosteum over the area anterior to the axilla of middle turbinate was either elevated as a flap or the mucosa overlying the bone was cauterised. The bone was removed using Kerrison rongeurs, drill or by osteotome, depending upon the thickness of the lacrimal bone in that particular patient. After removal of the bone, the lacrimal sac was identified by applying pressure over the sac from the outside. After identification of the sac, a vertical incision was made over the entire vertical extent of the sac and the pus was drained. The lacrimal sac was flushed and a portion of the medial wall of the sac was removed to prevent its subsequent closure. Any deviated nasal septum, polyps, paradoxical turbinate and concha bullosa which prevent the access of the surgery were tackled endoscopically at the same sitting.

A 1 cm x 0.5 cm size Merocel pack was used. This Merocel pack is tailor made from the commercially available Merocel nasal packs. A suture tie was placed on the lower end of the pack and this pack was placed inside the sac remnant. This suture tie was taken out through the ala of nose and taped with plaster. The Merocel pack was kept in situ for 10 days and on the 10th post-operative day the Merocel pack was removed from the sac by gently pulling the suture tie.

Statistical Analysis

P-value < 0.05 was used to establish the significance. SPSS software version 25 was used.

RESULTS

30 patients were enrolled into the study over a period of 90 months. There were 8 males [26%] and 22 females [73%] [Table 1]. Age of the participants ranged from 30 to 64 with a mean age of 49.04 [SD -9.99]. 12 patients [42%] were operated on left side and 13 patients [52%] were operated on right side. Over the period of six months follow- up, we observed 100% success (p-value < 0.0001). The complications like lacrimal pump failure, canthal erosion and canthal cheese wiring which are common in external DCR was not seen in our study [Table 1]. The complications seen in endoscopic DCR with silicone stents like granuloma formation, synechiae/ adhesion formation and foreign body sensation were less common, less than 16% [Table 1]. None or very few complications were observed in our series from 3 weeks follow-up onwards [Table 1]. When patients came for 3 weeks to 3 months follow-up, granuloma formation that were found was removed and was completely resolved by 3 months period. Synechiae found were released and by 3 months follow-up onwards synechiae had settled. Foreign body sensation settled immediately after the Merocel pack was removed [Table 1]. The minor complications seen in external DCR such as conjunctival haematoma, trauma to eyelids and periorbital oedema are not seen as the surgery is done endoscopically.

Endoscopic DCR without stents have high proportion of fibrosis of cut ends of the sac and hence cannot maintain the sac patency and tear drainage which leads to recurrence of patient's symptoms. Keeping the Merocel pack in the sac for 10 days aids in complete healing with no fibrosis of the cut ends of the sac, which helps to maintain the sac patency and tear drainage in long run.

	No Frequency (%)	Yes Frequency (%)
Lacrimal Pump Failure	30 (100%)	0 (0%)
Canthal Erosion, Canthal Cheese Wiring	30 (100%)	0 (0%)
Granuloma Formation		
10 days	28 (9%)	2 (8%)
3 weeks	28 (92%)	2 (8%)
6 weeks	28 (92%)	2 (8%)
3 months	30 (100%)	0 (0%)
6 months	30 (100%)	0 (0%)
1 year	30 (100%)	0 (0%)
Synechiae Formation		
10 days	26 (84%)	4(16%)
3 weeks	26 (84%)	4(16%)
6 weeks	26 (84%)	4(16%)
3 months	30 (100%)	0 (0%)
6 months	30 (100%)	0 (0%)
1 year	30 (100%)	0 (0%)
Foreign Body Sensation		
10 days	25 (80%)	5(20%)
3 weeks	29 (96%)	1(4%)
6 weeks	30 (100%)	0(0%)
3 months	30 (100%)	0 (0%)
6 months	30 (100%)	0 (0%)
1 year	30 (100%)	0 (0%)

Table 1. Frequency (%) level on success rate on Endoscopic DCR

Figure 1. Pre-op, 10th day post-op with Merocel pack in situ and 3 months Post-op

Figure 2. Pre-op, 10th day post-op with Merocel pack in situ and 3 months Post-op

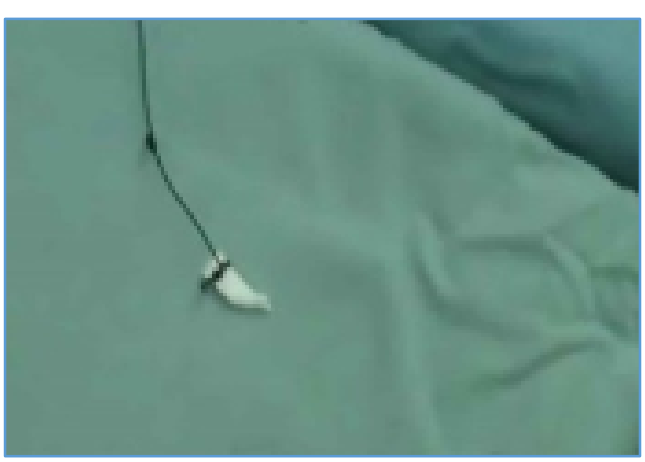

Figure 3. Merocel Pack with Suture Tie

DISCUSSION

Endoscopic DCR with 'Merocel pack in the sac technique' is an innovative, cost effective, less time consuming surgery with good patient compliance (p-value= 0.0001), less complications and good post-operative results. In our series there were no major complications compared to that seen in external DCR like lacrimal pump failure, canthal erosion and canthal cheese wiring. One study reported minor complications like orbital swelling, orbital haemorrhage and ecchymosis.[7,8,9,10,11,12,13,14] Our study did not find any of such complications of external DCR. Granuloma formation, synechiae/ adhesion formation and foreign body sensation seen were self-limiting and settled by 3 months' time. During serial visits if any were present, they were removed or released and later usually settles by itself. In any case, these complications were not seen in our cases after 3 months. The foreign body sensation usually settled with removal of Merocel pack. The Merocel pack in the sac for 10 days prevents synechiae between the opposing walls and hence sac patency is maintained, and patients get relieved of their complaints like epiphora and mucocele formation. Keeping Merocel pack in the lacrimal sac is less traumatic, easy technique and hence less time consuming which increases cost effectiveness and patient compliance.

CONCLUSION

Endoscopic DCR with 'Merocel pack in the sac technique' is an excellent novel technique for the treatment of epiphora and mucocele due to nasolacrimal duct obstruction. It should be the treatment of choice in view of maintenance of long-term results, patient compliance and cost effectiveness.

REFERENCES

[1] Jyothi AC, Kumar A, Chiluveru P. Endoscopic dacryocystorhinostomy: our experience. Int J Otorhinolaryngol Head Neck Surg 2016;2(4):254-7.

[2] Ray B, Datta D, Roy P. Long term success of Endo DCR with silicone tube catheter (STC) versus Endo DCR without STC: a comparative study. Bengal J Otolaryngol Head Neck Surg 2016;24(2):88-93.

[3] Chandler PA. Dacryocystorhinostomy. Trans Am Ophthalmol Soc 1936;34:240-63.

[4] Toti A. Nuovo metodo conservatore di cura radical delle suprazioni chroniche del sacco lacrimale. Clin Mod Firenze 1904;10:385-9.

[5] Metson R, Woog JJ, Puliafito CA. Endoscopic laser dacryocystorhinostomy. Laryngoscope 1994;104(3 Pt 1):269-74.

[6] McDonogh M, Meiring JH. Endoscopic transnasal dacryocystorhinostomy. J Laryngol Otol 1989;103(6):585-7.

[7] Maini S, Raghava N, Youngs R, et al. Endoscopic endonasal laser versus endonasal surgical dacrocystorhinostomy for epiphora due to nasolacrimal duct obstruction: prospective, randomized, controlled trail. J Laryngol Otol 2007;121(12):1170-6.

[8] Walker RA, Al-Ghoul A, Conlon MR. Comparison of non-laser non-endoscopic endonasal dacrocystorhinostomy with external dacrocystorhinostomy. Can J Ophthalmol 2011;46(2):191-5.

[9] Machin SJ, Laguarta CJM, De Gregorio AMA, et al. Lacrimal duct obstruction treated with nasolacrimal stent. Arch Soc Esp Oftalmol 2003;78(6):315-8.
[10] Bogdanici C, Lupascu C, Halunga M. Complications after catheterization of nasolacrimal duct. Oftalmologia 2002;55(4):39-42.
[11] Beloglazov VG, Atokova EL, Malaeva LV, et al. Intubation granulomas of the lacrimal ducts in patients with silicone implants. Vestn Oftalmol 1998;114(5):29-32.
[12] Allenn K, Berlin AJ. Dacrocystorhinostomy failure: association with nasolacrimal silicone intubation. Ophthalm Surg 1989;20(7):486-9.
[13] Walland MJ, Rose GE. The effect of silicone intubation and on failure and infection rates after dacrocyst orhinostomy. Ophthalmic Surg 1994;25(9):597-600.
[14] Unlu HH, Ozturk F, Mutlu C, et al. Endoscopic dacrocystorhinostomy without stents. Auris Nasus Larynx 2000;27(1):65-71.

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Original Research Article

Profile of allergic rhinitis patients in a tertiary care centre in Central Kerala

Ike Thomas[1], Geethu Mathew[2]*, Tina Thomas[1], Mahika Anilkumar[3], Kalyan George[3]

[1]Department of Otolaryngology, Believers Church Medical College Hospital, Thiruvalla, Kerala, India
[2]Department of Community Medicine, Believers Church Medical College Hospital, Thiruvalla, Kerala, India
[3]MBBS Student, Believers Church Medical College Hospital, Thiruvalla, Kerala, India

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***Correspondence:**
Dr. Geethu Mathew,
E-mail: matgeet@gmail.com

ABSTRACT

Background: Allergic rhinitis (AR) is a major public health problem which has a significant impact on the quality of life. Around 20-30% of the Indian population suffers from AR. The objective was to study the clinical profile and find out the factors associated with severity.
Methods: This retrospective record-based study was conducted among 182 patients with AR who visited Department of otorhinolaryngology in a tertiary care hospital in central Kerala. Variables collected included age, gender, family history of allergy, duration of disease and details of symptoms. The visual analogue scale (VAS) was used in identifying the severity of symptoms.
Results: The mean age of participants was 39±19 years. Nearly 50% of them had positive family history and 77.5% were found to have severe symptoms. The mean total VAS score was 25±5.1. Sneezing and nasal block were the common symptoms reported. Among the participants who had family history, 77 (87.5%) of them reported severe symptoms (χ^2=9.81, p=0.002). Almost all participants belonged to higher age group reported high severity (χ^2=48.130, p<0.0001). More than 80% of the participants who reported a longer duration of disease had severe symptoms (χ^2=7.28, p=0.007).
Conclusions: The study identified a significant proportion of sneezers and blockers among the study population. Older age group, family history and longer duration of disease were the factors associated with severity. Generating community awareness will help in early diagnosis, effective control with improvement in quality of life of these patients.

Keywords: Allergic rhinitis, Kerala, Profile

INTRODUCTION

AR, also known as hay fever, is a type of inflammation in the nose which occurs when the immune system overreacts to allergens.[1] It is an IgE-mediated inflammatory disease of the nasal mucosa, triggered by exposure to airborne allergens. It is also referred to as seasonal allergies and is triggered by grass, pollen, dust and dirt in the air and at times because of smoke and perfumes. AR is not severe or fatal until accompanied by asthma or anaphylaxis, which can be significant.[1,2] This is a major public health problem worldwide, with over 300 million people worldwide affected. Prevalence of AR has been increasing over the last few decades and around 20-30% of the Indian population suffers from AR. AR constitutes nearly 50% of all allergies in India.[3-5] AR

patients are classified as either sneezers runners and blockers due to their distinct clinical profile. In patients who are predominantly sneezer and runner main symptoms are sneezing, anterior rhinorrhoea, itchy nose and eyes, on the other hand blockers have nasal congestion as predominant symptoms in nasal blockage and thick mucous can lead to postnasal discharge breathlessness. These symptoms can be mild moderate or severe cause significant impact on the quality of life of the individual.[2] Conditions such as asthma, sinusitis, otitis media, nasal polyposis, lower respiratory tract infection and dental malocclusion are also associated with this condition.[3] Since viral respiratory infections occur frequently in young children and produce similar symptoms, it is very difficult to diagnose AR in the first 2 or 3 years of life. The prevalence of AR peaks in the second to fourth decades of life and then gradually diminishes.[6] AR contributes to missed or unproductive time at work and school, sleep problems and among affected children, decreased involvement in outdoor activities.[7] Even though the condition is quite prevalent in India, the literature on the clinical profile of AR from south Indian population is scarce. Hence this study was carried out to study the clinical profile of the patients and find out the association between selected variables with severity of AR.

METHODS

This retrospective record-based study was conducted among 182 patients with AR who visited department of otorhinolaryngology, in a tertiary care hospital in central Kerala for 3 years between January 2016 to January 2019. The study protocol was approved by institutional ethics committee prior to the commencement of data collection.

Considering a 68% prevalence of symptoms among patients, 10% precision at 95% CI, our sample size was calculated to be 182. Hence 182 patients with complete information on the hospital information system and those who cooperated for a phone call survey were included in the study using convenient sampling. Most of the information was taken from records and some missing information on selected records was collected through phone interviews. Study tool included a questionnaire which collected information on sociodemographic variables like age, gender, family history of allergy, duration of symptoms and details of symptoms. The psychometric response scale, VAS was used in identifying the severity of each symptom reported. The VAS is a horizontal long line with descriptors at its ends expressing two extremes of a feeling. AR patients were asked about their symptoms in detail and also were to mark a point in the VAS scale that best corresponds to the severity of their symptoms during the first visit. The respondent's cross is then assigned a score from 0 to 10. A score ranging from 1-5 is considered as mild and score more than 5 is considered as severe for that particular symptom. All symptom scores are added up to get a final VAS scoring which ranges from 0 to 50. Total score ≤25 is considered as mild and >25 is considered as severe.[8] Statistical analysis was performed using Epi info software.

Details of VAS scoring for individual AR symptoms

The details of VAS scoring for individual AR symptoms are congestion: 0 (free breathing), 10 (complete obstruction at day and night); rhinorrhoea/watering of eyes: 0 (dry nose/eyes all day), 10 (continuous secretion); headache: 0 (no headache, 10 (severe headache); itching: 0 (no itching), 10 (persistent itching disrupting everyday activities); sneezing: 0 (no sneezing), 10 (persistent sneezing fits for the whole day and night that disrupt normal functioning).

RESULTS

A total of 182 patients with allergic rhinitis were included in the study. The mean age of the participants was 39±19 years.

Table 1: Distribution of the study population based on demography and profile.

S. no.	Variables	Categories	Number (%)
1	Age groups (in years)	<18	24 (13.2)
		18-39	74 (40.7)
		40-59	52 (28.6)
		≥60	32 (17.6)
2	Gender	Male	103 (56.6)
		Female	79 (43.4)
3	Duration of allergy (in years)	≤5	32 (17.5)
		>5	150 (82.5)
4	Family history of AR	Yes	88 (48.4)
		No	94 (51.6)

Table 2: Distribution of study population based on the severity of symptoms.

S. no.	Variables	Categories	Number (%)
1	Sneezing	No symptom Mild symptom Severe symptom	1 (0.5) 0 181 (99.5)
2	Nasal block	No symptom Mild symptom Severe symptom	0 2 (1.1) 180 (98.9)
3	Headache	No symptom Mild symptom Severe symptom	45 (24.7) 104 (57.1) 33 (18.1)
4	Itching	No symptom Mild symptom Severe symptom	31 (17) 87 (47.8) 64 (35.2)
5	Watering of eyes	No symptom Mild symptom Severe symptom	3 (1.6) 39 (21.4) 140 (76.9)

Table 3: Association of severity of AR with selected variables.

S. no.	Variables	Categories	Severity of AR			Chi square, p value
			Mild (%)	Severe (%)	Total	
1	Age groups (in years)	<18	13 (54.2)	11 (45.8)	24	χ^2=48.130 p<0.0001
		18-39	28 (37.8)	46 (62.2)	74	
		≥40	0	84 (100)	84	
2	Gender	Male	26 (25.2)	77 (74.8)	103	χ^2=1.002 p=0.317
		Female	15 (19)	64 (81)	79	
3	Duration of disease (in years)	≤5	13 (40.6)	19 (59.4)	32	χ^2=7.28 p=0.007
		>5	28 (18.7)	122 (81.3)	150	
4	Family history	Yes	11 (12.5)	77 (87.5)	88	χ^2=9.81 p=0.002
		No	30 (31.9)	64 (68.1)	94	

Clinical profile of patients and factors associated with severity of symptoms

Among the study participants 141 (77.5%) were found to have severe symptoms according to total VAS scoring (total score >25). The mean total VAS score was calculated to be 25±5.1. The common symptoms reported were sneezing and nasal block among the participants. Almost all of them reported to have high severity of these symptoms. The mean VAS scores were 8.7±0.8 and 7.7±0.8 for sneezing and nasal block respectively. Nearly 75% of the participants had severe watering of eyes (mean score=6.1±2.2). Headache and itching were relatively uncommon among study population and majority of them reported only mild symptoms. Details of symptoms in Table 2. The association between severity of symptoms and collected variables were done. Among the participants who reported to have family history of AR, 77 (87.5%) of them reported severe symptoms. Similarly, almost all participants belonged to higher age group reported high severity. Duration of the disease was another factor that was found to be associated with severity. More than 80% of the participants who reported a longer duration of disease had severe symptoms. Even though females reported higher severity of symptoms compared to males, this association was not statistically significant, details in Table 3.

DISCUSSION

AR is a highly prevalent health condition which is often neglected in many developing countries including India.[9] It affects physical, mental and social aspects of life and can have significant impact on the quality of life of patients.[10] The present study explains the clinical profile of the patients with AR and factors associated with severity of the symptoms. Majority of the population were below 40 years with almost equal gender distribution. Results of this study is comparable to a similar study done in north India where majority of the patients belonged to younger age category. A similar study done in Kolkata showed that most of the patients with allergic rhinitis belonged to age group of 20-39 years. Also study done in Chhattisgarh revealed that majority of patients belonged to third decade of life.[3,11] It may be due to the decline in antibody levels and T cells associated with immunosuppression in old age.[3] Nasal

obstruction, sneezing, runny nose, nasal itching, postnasal drip are diagnostic symptoms of allergic rhinitis. The most common symptoms identified in this study were nasal block and sneezing. This study showed that the proportion of sneezers were comparable to the proportion of blockers. Results of the studies done in India and abroad showed variations in the proportion of sneezers/blockers depending on the study population and area.[3,9,11] Nearly three forth of the participants reported to have severe symptoms. Available literature also supports this finding. Probably due to the fact that patients most of the patients who come to hospital for consultation may have moderate to severe symptoms compared to others. The study also explored factors associated with severity of symptoms. We observed a slight female predominance among patients with severe symptoms in our study and is in is in consonance with other study findings. Another study done by Larson et al observed that women suffer from more severe rhinitis compared to men.[12] Gender differences may be due to differences in response to the same disease and severity level. It has been noticed that women are more emotionally distressed by the sheer presence of symptoms, while men do not react until their symptoms become severe and long-standing. Even though the disease condition is more common among younger age groups, severity of symptoms was found to be high among older people. Age related changes are known to worsen symptoms related to rhinitis. Longer duration of the disease was found to be significantly associated with severity of symptoms. Similar results were obtained in another study done in India where severity of the symptoms were directly proportional to the duration of the disease.[3] Nearly half of our study population had a positive family history of allergic rhinitis. We observed that the family history of allergic rhinitis was strongly associated with severity of the disease among patients. There is clear and strong evidence to support the influence of genetic predisposition in allergic rhinitis. The complex interaction of genetic and environmental factors play a pivotal role in the etiology of AR. Understanding these factors and sensitization will help in more effective measures of prevention and intervention.[13]

Limitation

As it was predominantly a record based study, comorbidities and other factors associated with severity of allergic rhinitis could not be included. Hospital based clinical profile and severity may not be a true representation of the situation in the community.

CONCLUSION

The study identified a significant proportion of sneezers and blockers among the study population. More than 75% of them had severe symptoms. Older age group, family history and longer duration of disease were the factors associated with high severity of symptoms. Generating community awareness regarding various aspects of the disease will help in early diagnosis, effective control and prevention of the disease with improvement in quality of life of these patients.

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REFERENCES EDITING REQ.

1. Sharma V, Kumar R. Clinical assessment of patients suffering from allergic rhinitis. Int J Med Heal Res. 2019;5(3);137-40.
2. Hossenbaccus L, Linton S, Garvey S, Ellis AK. Towards definitive management of allergic rhinitis: best use of new and established therapies. Allerg Asthma Clin Immunol. 2020;16:39.
3. Shukla SK. Clinical profile of allergic rhinitis patients in Bastar. Med Pulse Int J ENT. 2017;3(3):14-6.
4. Wikstén J, Toppila-Salmi S, Mäkelä M. Primary Prevention of Airway Allergy. Curr Treat Options Allergy. 2018;5(4):347-55.
5. Varshney J, Varshney H. Allergic rhinitis: An Overview. Ind J Otolaryngol Head Neck Surg. 2015;67(2):143.
6. Yonekura S, Okamoto Y, Horiguchi S, Sakurai D, Chazono H, Hanazawa T, et al. Effects of aging on the natural history of seasonal allergic rhinitis in middle-aged subjects in south Chiba, Japan. Int Arch Allergy Immunol. 2012;157(1):73-80.
7. Meltzer EO, Blaiss MS, Derebery MJ, Mahr TA, Gordon BR, Sheth KK, et al. Burden of allergic rhinitis: results from the pediatric allergies in America survey. Allergy Clin Immunol. 2009;124(3):43-70.
8. Klimek L, Bergman K, Biedermann T, Bousquet J, Hellings P, Jung K, et al Visual analogue scales (VAS): measuring instruments for the documentation of symptoms and therapy monitoring in cases of allergic rhinitis in everyday health care: position paper of the German society of allergology (AeDA) and the German society of allergy and clinical immunology (DGAKI), ENT section in collaboration with the working group on clinical immunology, allergology and environmental medicine of the German society of otorhinolaryngology, head and neck surgery (DGHNOKHC). Allergo J Int. 2017;26(1):16-24.
9. Ibekwe PU, Ibekwe TS. Skin prick test analysis in allergic rhinitis patients: a preliminary study in Abuja, Nigeria. J Allergy. 2016;2016:3219104.
10. Shah A, Pawankar R. Allergic rhinitis and comorbid asthma: perspective from India: ARIA Asia-Pacific Workshop report. Asian Pac J Allergy Immunol. 2009;27(1):71-7.
11. Deb A, Mukherjee S, Saha BK, Sarkar BS, Pal J, Pandey N, et al. Profile of patients with allergic rhinitis (ar): a clinic based cross-sectional study

from Kolkata, India. J Clin Diagn Res. 2014;8(1):67-70.
12. Larsson U, Taft C, Karlsson J, Sullivan M. Gender and age differences in the relative burden of rhinitis and asthma on health-related quality of life-a Swedish population study. Respir Med. 2007;101(6):1291-8.
13. Wang DY. Risk factors of allergic rhinitis: genetic or environmental? Ther Clin Risk Manag. 2005;1(2):115-23.

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Original Research Paper

Pathology

GORLIN - GOLTZ SYNDROME - A CASE REPORT AND REVIEW OF LITERATURE

Dr Asha Peedikayil Punnoose* Assistant Professor, Department Of Pathology, Believers Church Medical College, Thiruvalla,Kerala. *Corresponding Author

Dr Bichu Jacob Assistant Professor, Department Of OFMS, Believers Church Medical College, Thiruvalla,Kerala.

Dr Ike Thomas Associate Professor, Department Of ENT, Believers Church Medical College, Thiruvalla,Kerala.

ABSTRACT Gorlin-Goltz syndrome(GGS)(NBCCS) (Nevoid basal cell carcinoma syndrome) is characterised by multiple basal cell carcinomas appearing at a younger age, associated with odontogenic keratocysts of the jaw, skeletal abnormalities, ectopic calcification, and pits of the hands and feet with equal sex predilection. A case of 48 year old male patient presented with Gorlin- Goltz syndrome is presented in this study. Our report emphases the clinical and pathological features of this syndrome for the early diagnosis and genetic counselling for the patients.

KEYWORDS : Gorlin Goltz syndrome, Basal cell carcinoma, Odontogenic Keratocyst.

INTRODUCTION

The name Gorlin syndrome is derived from the name of American oral pathologist and human geneticist Robert J Gorlin (1923-2006) and his co-author Robert W. Goltz (1923-2014), an American dermatologist. It was first described in 1960 by Gorlin and Goltz [1]. It is an autosomal dominant disorder with complete penetrance and variable expressivity. Gorlin - Goltz syndrome has rarely been reported from India. We report here one such patient diagnosed at our hospital.

CASE REPORT

48 year male presented with skin lesions in right eyebrow, (Fig.1) posterior surface of pinna of right ear (Fig.2) and on scalp (Fig.3). Detailed clinical examination revealed swelling in right and left jaw and multiple cutaneous lesions over face, apart from ocular hpertelorism, heavy fused eyebrows (Fig.4) and multiple palmar pits (Fig.5). An orthopantomogram (Fig.6) revealed multiple radiolucent cysts in right mandible, left mandible and left maxilla. Excision of the swellings, enucleation and chemical cauterisation of the cysts were done.

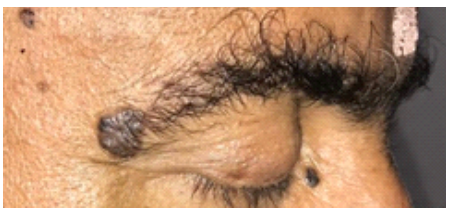

Fig.1 Swelling near right eyebrow

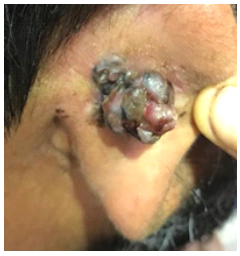

Fig. 2 Nodular swelling on posterior surface of pinna of right ear.

Fig.3 Swelling over scalp.

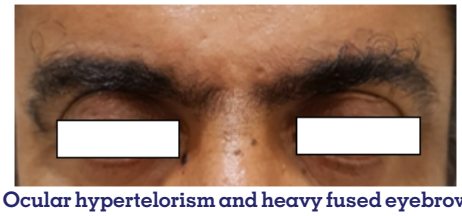

Fig 4. Ocular hypertelorism and heavy fused eyebrows.

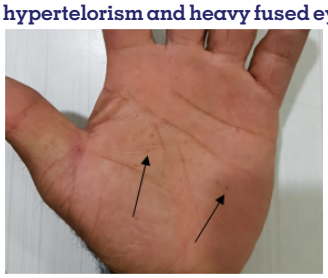

Fig.5 Multiple palmar pits.

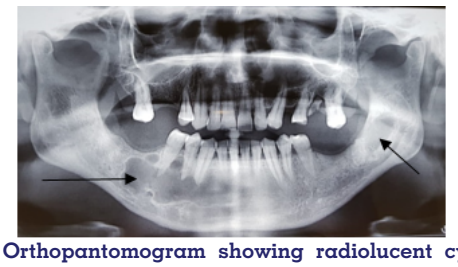

Fig.6 Orthopantomogram showing radiolucent cysts in right and left mandible.

Specimens were sent for histopathological examination. Sections from swelling in right eyebrow and ear show a malignant neoplasm arising from epidermis (Fig. 7) composed of lobules of basaloid cells with peripheral palisading (Fig.8) and retraction cleft between tumour lobules and stroma (Fig.9). Final diagnosis was basal cell carcinoma (nodulocystic type). Cysts from the jaw show odontogenic keratocyst (Fig.10).

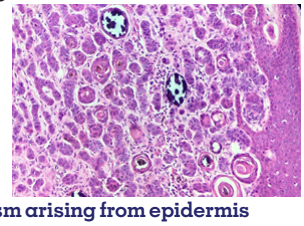

Fig.7 Neoplasm arising from epidermis

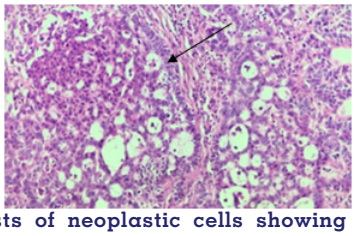

Fig. 8 Nests of neoplastic cells showing peripheral palisading.

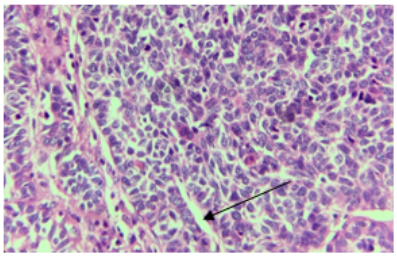

Fig.9 Artefactual cleft between stroma and neoplastic nests

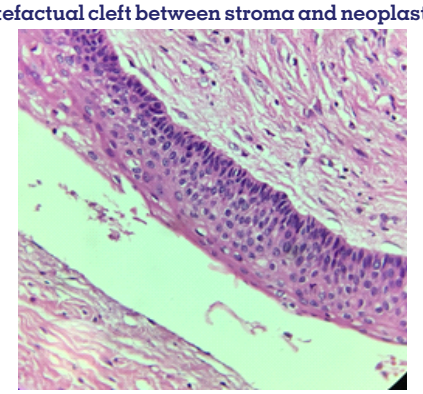

Fig.10 Odontogenic keratocyst.

DISCUSSION

NBCCS was first described by Jarish and White in 1894 and was later established as a unique syndrome by Gorlin-Goltz in 1960. Inheritance pattern is autosomal dominant with complete penetrance and variable expressivity. Significant number of cases show spontaneous germ cell mutations also. Genetic studies identified human homologue of Drosophila patched (PTCH) as the candidate gene mapped to chromosome 9q23.1-q31 [2].

Homozygous inactivation of the gene results in basal cell carcinomas whereas hemizygous germline mutations cause the numerous congenital abnormalities. This gene is part of the sonic hedgehog signaling pathway involved in control of cell proliferation and development of neural tube, pharyngeal pouches, somites, and limb buds in vertebrates. Gorlin - Goltz Syndrome is characterized mainly by the presence of multiple basal cell carcinomas (BCC), odontogenic keratocysts (OKCs) of the jaw, palmar pits and ectopic calcifications of the falx cerebri. More than 100 minor criteria have been described. The presence of two major and one minor criteria or one major and three minor criteria are necessary to establish a diagnosis.

Evans et al. [3] first established major and minor criteria for diagnosis of this rare entity, later modified by Kimonis et al. [4]. According to them diagnosis of GGS can be established when two major or one major and two minor features are present.

The major criteria are:

- Multiple Basal cell carcinoma or one occurring under the age of 20 years
- Histologically proven odontogenic keratocysts of the jaws.
- Palmar or plantar pits (three or more).
- Bilamellar calcification of the falx cerebri.
- Bifid, fused or markedly splayed ribs.
- First-degree relative with NBCCS.

The minor criteria are:

- Macrocephaly (adjusted for height).
- Congenital malformation: Cleft lip or palate, frontal bossing, coarse face, moderate or severe hypertelorism.
- Other skeletal abnormalities: Sprengel deformity, marked pectus deformity, marked syndactyly of the digits.
- Radiological abnormalities: Bridging of the sella turcica, vertebral anomalies such as hemivertebrae, fusion of the vertebral bodies, modelling defects of the hands and feet.
- Ovarian fibroma.
- Medulloblastoma.

In the present case, the diagnosis of the Gorlin-Goltz syndrome was made due to the presence of three major criteria - multiple basal cell carcinomas (BCCs), multiple odontogenic keratocysts of the jaw bone and palmar pits along with minor criteria of hypertelorism.

Basal cell carcinomas usually develops in adolescent age group, some reports suggest that it could also develop during early childhood [5,6]. They have a variable appearance characterised by small flesh-coloured or brown dome-shaped papules, soft nodules or flat plaques measuring up to 1 cm in diameter associated with numerous milia. The larger tumours are usually pigmented, often ulcerate, and typically behave in an aggressive fashion affecting both exposed and covered sites. The central part of the face is commonly the first area to be involved, followed by the chest, back, and scalp and then other exposed sites [7]. Rarely genitals can be involved [8].

Patients may harbour up to thousands of basal cell carcinomas, but about 10% of adult patients do not manifest skin tumours. Rarely, the tumours have a unilateral distribution. Multiple large epidermoid cysts are also often evident on the trunk and limbs.

All variants of basal cell carcinoma may be seen, but the solid and superficial types are most common, with 30 % patients having two or more histological types of BCC and morpheaform tumors being rare. There are no particular distinguishing features by which they might be distinguished from ordinary basal cell carcinoma [9].

Odontogenic keratocysts of the jaws occur in up to 65-100% of patients, usually in childhood or adolescence [10]. They are commonly multiple and show a predilection for the premolar area and are associated with tooth displacement, pain, and swelling of the jaws. It is common for recurrence following a surgical procedure.

The jaw cysts are lined by stratified squamous epithelium with a thick fibrous capsule and may be associated with the development of spindle cell squamous carcinoma, myxoma, ameloblastoma, and fibrosarcoma.

Skeletal abnormalities, which are present in up to 75% of cases, include generalized overgrowth, macrocephaly, bridging of the sella turcica, high-arched palate, vertebral abnormalities, splayed, fused, missing or bifid ribs, kyphoscoliosis, spina bifida occulta, hyperplasia of the mandibular coronoid processes, and bone cysts. There is a characteristic 'dished' facial appearance due to frontal and biparietal bossing, broadening of the nasal root, and ocular hypertelorism.

Ectopic lamellar calcification of the falx cerebri (85-90%) and also of the diaphragma sellae (60-80%), tentorium cerebelli (40%), and petroclinoid ligaments (20%) are frequent

manifestations. Craniocerebral manifestations may be evident in utero with epilepsy being an occasional complication.

The shallow pits of the palms and soles are pathognomonic. Pits can be seen in 85% of the patients above 20 years [11]. They are asymptomatic, 1-3 mm deep and 2-3 mm across, [12] and may be present in their hundreds. The palmar and plantar pits show a diminution or loss of the keratin and granular layers, associated with a thinned underlying malpighian layer. Electron microscopic examination reveals incompletely discharged Odland bodies. The pits are therefore believed to develop as a result of reduced 'intral-amellar cement' resulting in diminished adherence between the keratin lamellae. The rete ridge pattern is irregular in size and shape and some pits may show basaloid proliferation. No BCC has been found to arise from these pits [13].

Patients with the nevoid basal cell carcinoma syndrome may have many other abnormalities, including congenital blindness, hypogonadism, ovarian fibromas (75% of females), cardiac fibromas, and an increased incidence of central nervous system tumors, including medulloblastoma and meningiomas.

Rare associations include eosinophilic pustular folliculitis, nevus sebaceous, microphthalmia, ulcerative colitis, unilateral renal agenesis, multiple acrochorda, ameloblas toma, thyroid neoplasia, prenatal chylothorax, hepatic mesenchymal tumor, fetal rhabdomyoma, lymphomatoid papulosis, primary ovarian leiomyosarcoma, rhabdomy osarcoma, and Wilms' tumor as well as undifferentiated sinonasal carcinoma.

Differential diagnosis-

The nevoid basal cell carcinoma syndrome should be distinguished from the linear unilateral basal cell nevus (Carney's) syndrome, which comprises an extensive unilateral lesion consisting of basaloid follicular hamartomas in addition to comedones, epidermoid cysts, and areas of epidermal atrophy. Patients may also have scoliosis, but there are no other significant internal abnormalities. Other differential diagnoses include Bazex syndrome, trichoepithelioma papulosum multiplex and Torre's syndrome (Muir-Torre's syndrome).

Bazex syndrome is characterized by multiple BCCs, milia, reduced sweating (hypohidrosis), abnormal loss of hair (hypotrichosis), and follicular atrophoderma, a skin condition involving breakdown of the follicles of the skin and causing lesions, especially on the arms and legs. In most affected individuals, BCCs develop in the 20s or 30s. Additional symptoms can vary greatly from one person to another.

Muir-Torre syndrome is characterized by a predisposition to skin cancer and certain low grade visceral cancers. BCCs have been reported in individuals with Muir-Torre syndrome.
Clinical diagnosis relies on the specific criteria. Gene mutation analysis confirms the diagnosis. Genetic counselling is mandatory. Antenatal diagnosis is feasible by means of ultrasound scans and analysis of DNA extracted from fetal cells (obtained by amniocentesis or chorionic villus sampling).

Patients with Gorlin syndrome may be hypersensitive to and contraindicated from receiving radiation therapy.

Life expectancy in NBCCS is not significantly altered but morbidity from complications can be substantial. Regular follow-up by a multi-specialist team (dermatologist, neurologist and odontologist) should be offered.

CONCLUSIONS

Nevoid basal cell carcinoma is a multisystem disorder. Diagnosis requires a multisystem approach by dermatologist, odontologist and neurologist.

REFERENCES:

1. Gorlin RJ, Goltz RW: Multiple nevoid basal-cell epithelioma, jaw cyst and bifid rib: A syndrome. New Engl J Med. 1960 May, 5:908-912.
2. Gorlin RJ: Nevoid basal cell carcinoma (Gorlin) syndrome: Unanswered issues. J Lab Clin Med. 1999, 134:551-2.
3. Evans DG, Sims DG, Donnai D: Family implications of neonatal Gorlin's syndrome. Arch Dis Child. 1991, 66(10 Spec No):1162-3.
4. Kimonis VE, Goldstein AM, Pastakia B, et al.: Clinical manifestations in 105 persons with nevoid basal cell carcinoma syndrome. Am J Med Genet. 1997, 69:299-308.
5. Evans DG, Ladusans EJ, Rimmer S, Burnell LD, Thakker N, Farndon PA: Complications of the naevoid basal cell carcinoma syndrome: results of a population based study. J Med Genet. 1993, 30:460-464.
6. Shanley S, Ratcliffe J, Hockey A, et al.: Nevoid basal cell carcinoma syndrome: review of 118 affected individuals. Am J Med Genet. 1994, 50:282-290.
7. Casaroto AR, Loures DC, Moreschi E, Veltrini VC, Trento CL, Gottardo VD, Lara VS: Early diagnosis of Gorlin-Goltz syndrome: case report. Head Face Med. 2011, 7:2.
8. Giuliani M, Di Stefano L, Zoccali G, Angelone E, Leocata P, Mascaretti G: Gorlin syndrome associated with basal cell carcinoma of the vulva: A case report. Eur J Gynaecol Oncol. 2006, 27:519-522.
9. Gorlin RJ: Nevoid basal cell carcinoma (Gorlin) syndrome. Genet Med. 2004, 6:530-539.
10. Sun LS, Li XF, Li TJ: PTCH1 and SMO gene alterations in keratocystic odontogenic tumors.. J Dent Res. 2008, 87:575-579.
11. Gutierrez MM, Mora RG: Nevoid basal cell carcinoma syndrome. A review and case report of a patient with unilateral basal cell nevus syndrome. J Am Acad Dermatol. 1986, 15:1023-1030.
12. Friedlander AH, Herbosa EG, Peoples JR: Ocular hypertelorism, facial basal cell carcinomas, and multiple odontogenic keratocysts of the jaws. J Am Dent Assoc. 1988, 116:887-889.
13. Gorlin RJ: Nevoid basal-cell carcinoma syndrome. Medicine (Baltimore). 1987, 66:98-113.

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OROPHARYNGEAL CHORISTOMA – A CASE REPORT & REVIEW OF LITERATURE

Pathology

Dr. Ajitha.K* Professor Dept of pathology, Believers' Church Medical college, Thiruvalla, Kerala, INDIA. *Corresponding Author

Dr.Sunil Raj Associate Professor, Dept of Pediatrics, Believers' Church Medical college, Thiruvalla, Kerala, INDIA.

Dr Ike Thomas Assistant Professor, Dept of ENT, Believers' Church Medical college, Thiruvalla, Kerala, INDIA.

Dr Girija Mohan Professor, Dept of Pediatrics, Believers Church Medical college, Thiruvalla, Kerala, INDIA

ABSTRACT

We report an unusual case of a 9 year old boy who presented with a posterior pharyngeal wall lesion which on excision biopsy was diagnosed as oropharyngeal gastric choristoma. Gastric choristoma in the pharynx is extremely rare and only a few cases have been reported so far.

Summary: Oropharyngeal gastric choristoma presenting with features of obstruction in a child is unusual and necessitates surgical excision.

KEYWORDS

tumor like lesion, choristoma, oropharyngeal mass

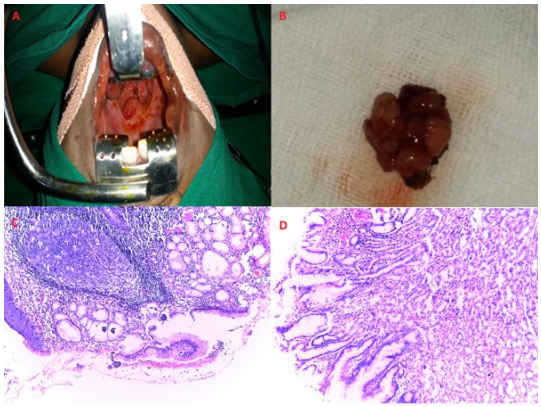
Introduction:

Choristoma is defined as the presence of histologically normal appearing tissue of a type which is not normally found in that anatomic site. They are tumorlike lesions which usually can occur in a wide range of anatomical locations. In the GIT,it has been found throughout the alimentary tract, ranging from scattered rests of cells to well formed mucosa with submucosal smooth muscle.[1] They are considered as developmental anomalies or embryological accidents and not neoplastic. They are mostly seen in the younger age group especially infants and neonates . In the older age group they are usually smaller in size , asymptomatic and hence incidental findings.They are also known as heterotopias and is composed of a single type of tissue but can show a mixed tissue pattern as well. Gastroinestinal or gastric type of choristoma is rare in the head and neck region.Most of these cases are reported in the oral cavity especially floor of the mouth. Pharyngeal gastric choristoma is extremly rare.

Case history:

9 year old child presented with history of growth retardation and snoring O/E Child was malnourished and short statured. Oropharyngeal mass lesion was seen with attachment to posterior pharyngeal wall(fig A)There was no attachment to the tongue. With a clinical diagnosis of aberrant adenoids, the child was referred to ENT department where an excisiom biopsy of the mass was done.

Grossly the excised specimen (figure B) was a nodular firm mass measuring 2x2x1.5cm. Cut surface showed a solid brownish homogeneous appearance. Under the microscope(figure C & D) it was a circumscribed lesion composed predominantly of full thickness normal gastric mucosal tissue with surface epithelium,lamina propria and muscle layer. It was was covered by oropharyngeal mucosa and a diagnosis of oropharyngeal choristoma was made.



A. Oropharyngeal mass lesion attached to posterior pharyngeal wall

B. Excised specimen

C. Oropharyngeal mucosal epithelium on the left side and gastric mucosal tissue on the right side

D. Gastric mucosal tissue

Discussion

Choristomas also called heterotopias present with normal tissues at abnormal locations.. Clinically and morphologically choristomas tend to resemble tumors. These are usually smaller asymptomatic lesions and very often incidental findings identified during endoscopic procedures.

There are reports of naso/oropharyngeal choristomas composed of brain, cartilage and skin tissues. Gastric heterotopias also can occur throughout the GIT. Upper esophagus[2] and tongue are the common sites. But pharynnx is an exceptional location for gastric choristoma and it was first reported by Stout and Lattes in 1957.[3]Clinically it can manifest as airway obstruction [4] and feeding difficulties especially in the neonate.[5] It can be swallowing/ breathing difficulties leading to developmental problems in older children as has happened in our case.

The most important differential diagnosis of choristoma is hamartoma which is considered as another tumorlike lesion. A hamatoma is a haphazardly arranged mass of tissues that are indigenous to the site where it is found. Unlike choristomas the margins of these lesions are ill defined and merges with surrounding tissues. Hamartomas and choristomas appear clinicaly as exophytic masses and the size vary from very tiny lesions to larger lesions measuring few centimetres. They have been described over a wide age range from birth to old age. Histologically these two lesions may show overlapping morphological features and sometimes may even be misdiagnosed as benign tumors.

Surgical excision is the treatment option as was done in our case and is curative. Even in asymptomatic patients excision biopsy is needed to establish the diagnosis

Conclusion :

Only very few cases of oropharyngeal choristomas are so far reported in literature and here we report one more case with review of relevant medical literature.

References

1. Hsu JM, Mortelliti AJ.Gastric choristoma of the hypopharynx presenting in an infant: a case report and review of the literature. Int J Pediatr Otorhinolaryngol. 2000 Nov 30;56(1):53-8.
2. Borhan-Manesh F, Farnum JB. Incidence of heterotopic gastric mucosa in the upper oesophagus. Gut.1991 Sep;32(9):968–972.
3. Hyun Chang, MD, Youngjin Ahn, MD, Yune Sung Lim, MD, and J. Hun Hah, MD Gastric Choristoma of the Oropharynx. Clin Exp Otorhinolaryngol. 2009 Jun; 2(2): 103–105.
4. Johnston C, Benjamin B, Harrison H, Kan A. Gastric heterotopia causing airway obstruction. Int J Pediatr Otorhinolaryngol. 1989 Sep;18(1):67–72
5. Marín Gabriel MA, Medina López C, Delgado Muñoz MD, Rodríguez Gil Y. Gastric heterotopia in the nasopharynx causing airway obstruction in the newborn.Int J Pediatr Otorhinolaryngol. 2004 Jul;68(7):961-4.