

A case series on allergic fungal rhino sinusitis – variable presentations



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Submission: 16-02-2022

Revision: 04-03-2023

Publication: 01-04-2023

ABSTRACT

Allergic fungal rhino sinusitis (AFRS) is a non-invasive fungal sinusitis resulting from an allergic and immunologic response to the presence of extramucosal fungal hyphae in the sinuses. Defined largely by the presence of allergic fungal mucin, which is a thick, tenacious, and eosinophilic secretion with characteristic histologic findings. To present a case series on the variable presentations in patients diagnosed with AFRS. A retrospective study of 10 cases that presented to Rajiv Gandhi Government General Hospital ENT outpatient department with allergic fungal rhinosinusitis confirmed by diagnostic nasal endoscopy and CT scan who were managed successfully by endoscopic sinus surgery. In our study, out of 10 cases, nine patients had unilateral symptoms, only one patient has bilateral symptoms. Most common presentation was headache, facial pain, nasal obstruction, and nasal discharge. Some patients had variable presentations such as proptosis, oroantral fistula, blurring of vision, watering of eye, and unilateral frontal headache. On diagnostic nasal endoscopy, two patients had nasal polyposis. Complete disease clearance was achieved in nine cases by functional endoscopic sinus surgery. Patients were started on topical and oral corticosteroids, pre/postoperatively. On follow-up, only one patient had recurrence. Preventing and treating the condition, as well as its spread and complications, are greatly aided by early detection and management of AFRS. The key to enhancing the result is surgical intervention (endoscopic sinus surgery) and medical therapy (corticosteroids). Antifungals have limited role in treatment of AFRS. Longstanding unilateral Sinusitis should always raise suspicious of AFRS.

Key words: Allergic mucin; Allergic sinusitis; Fungal sinusitis; Endoscopic sinus surgery; Corticosteroids

INTRODUCTION

Allergic fungal rhino sinusitis (AFRS) is a subset of polypoid chronic rhinosinusitis, which is characterized by the presence of eosinophilic mucin with fungal hyphae within the sinuses and Type I hypersensitivity to fungi.¹ According to various research, 5–10% of individuals with chronic rhinosinusitis have allergic fungal sinusitis,^{2,3} although in other studies, the incidence is substantially greater.⁴

The fact that many publications in the literature are describing AFRS with various clinical presentations, some even with multiple clinical presentations, is interesting including the concomitant occurrence of more than one kind of fungal rhinosinusitis.^{5,6}

The discovery of fungus in eosinophilic mucin without Type I hypersensitivity in the majority of chronic rhino sinusitis cases has made it difficult to explain allergic fungal rhino sinusitis. To highlight the function of eosinophils, Ponikau et al., retained a new name for this illness and eosinophilic fungal rhinosinusitis.⁷ A non-invasive fungal sinusitis caused by an allergic and immunologic reaction to the presence of extra mucosal fungal hyphae in the sinuses is currently known as allergic fungal rhinosinusitis.⁸ Hence, the main objective of this study was to study the various clinical presentations of allergic fungal rhino sinusitis, characterize the same, and correlate the histopathological profile.⁸

MATERIALS AND METHODS

In this retrospective study, we collected all the case details of 10 patients, who were admitted in ENT ward, Rajiv

Access this article online

Website:

<http://nepjol.info/index.php/AJMS>

DOI: 10.3126/ajms.v14i4.52573

E-ISSN: 2091-0576

P-ISSN: 2467-9100

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Gandhi Government General Hospital during the period of August 2022 to Jan 2023. All the patients presented with clinical symptoms suggestive of AFRS. Most of the patients presented with complaints of headache, facial pain, nasal obstruction, and nasal discharge. Some patients had variable presentations such as proptosis, oroantral fistula, blurring of vision, and watering of eye. On diagnostic nasal endoscopy, most patients had mucosal thickening, thick purulent secretions, two patients had nasal polyposis, and one patient had oroantral fistula. All patients did computerized tomography of the paranasal sinus (CT PNS), which showed heterogeneous intensities within paranasal sinuses, which is unilateral in nine patients and bilateral in one patient. One patient has isolated frontal sinus involvement. All patients were started on oral corticosteroids, underwent functional endoscopic sinus surgery, and continued oral corticosteroids postoperatively for 3 weeks. We followed all patients postoperatively at 2 weeks, 3 months, and 6 months. Out of 10 patients, one patient had recurrence and underwent revision surgery and given antifungal drug Itraconazole for 3 weeks.

CASES WITH VARIABLE PRESENTATIONS

Case 1

A 17-year-old male with blurring vision of the right eye for 1 week and right-sided headache for 2 days. No history of anosmia and diplopia. Vision: RE-6/9, LE-6/6, EOM: B/L Full. Diagnostic Nasal Endoscopy showed thick mucopurulent secretions and mucosal edema.



CT PNS shows soft-tissue thickening with hyperdensities noted in the right maxillary sinus, right ethmoid sinus, bilateral frontal sinus, sphenoid sinus, focal bony erosions noted in the right posterolateral and medial wall of maxillary sinus, and right lamina papyracea.

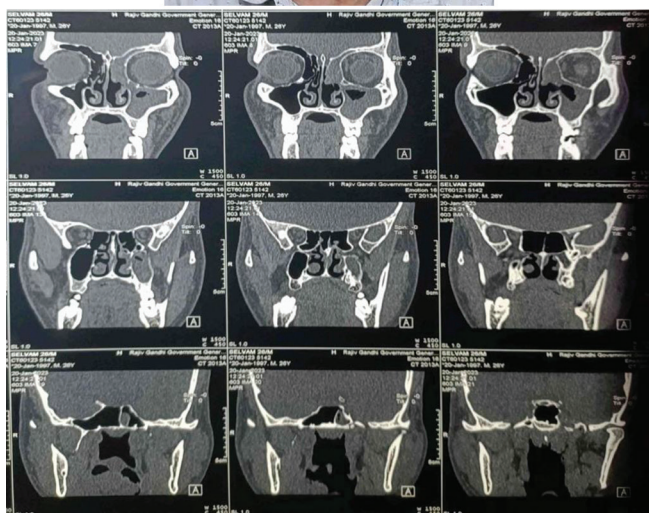
Case 2

A 35-year-old female with presenting complaints of the left oroantral fistula for 1 month. History of anosmia present, no history of diplopia/blurring of vision. Diagnostic nasal endoscopy shows mucosal edema, polypoidal mass in the left middle meatus, and mucopurulent secretions in the left nasal cavity. CT PNS shows extensive mucosal thickening of the left maxillary sinus, left ethmoid sinus, left frontal sinus, and left sphenoid sinus. Enlarged left maxillary sinus filled with soft-tissue density lesion (3.3*3.4 cm) with peripheral rim calcification and mottled internal air foci with bowing of medial wall of the left maxillary sinus.



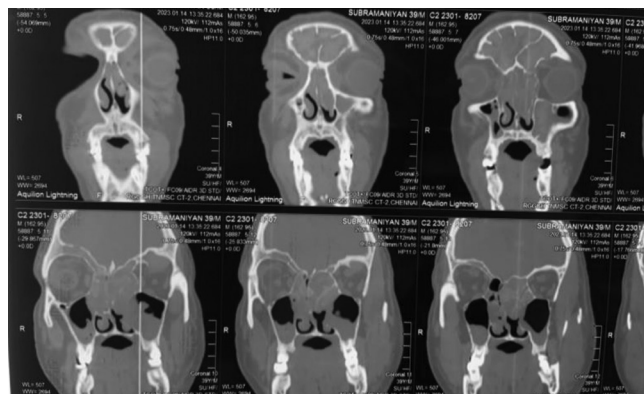
Case 3

A 26-year-old female with presenting complaints of the left eye pain and watering from the left eye for 2 weeks. Vision was normal. Vision: RE-6/6, LE-6/6, EOM: B/L Full. Diagnostic nasal endoscopy shows mucosal edema in the left nasal cavity. CT PNS shows soft-tissue opacification of the left maxillary sinus, left ethmoid sinus, and left frontal sinus. Intraoperatively, patient had erosion of the left lamina papyracea and medial wall of orbit.



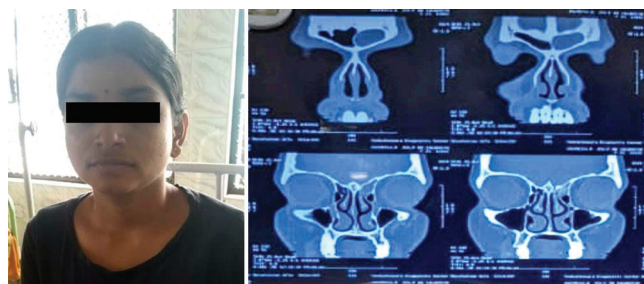
Case 4

A 39-year-old male with presenting complaints of proptosis for 6 months and anosmia for 4 months. Diagnostic nasal endoscopy shows mucosal edema, polypoidal mass in bilateral middle meatus, and mucopurulent secretions in bilateral nasal cavity. CT PNS shows opacification of bilateral maxillary, bilateral ethmoid, bilateral frontal, and bilateral sphenoid sinus.



Case 5

A 25-year-old female with presenting complaints of the left-sided frontal headache. Vision was normal. Vision: RE-6/6, LE-6/6, EOM: B/L Full. Diagnostic nasal endoscopy shows mucosal edema in the left nasal cavity. CT PNS shows isolated frontal sinus involvement.



RESULTS

We studied 10 patients of allergic fungal rhinosinusitis (Table 1), out of 10, five patients were male and five patients were females. The mean age of presentation was 13–60. Maxillary sinus was involved in nine cases, Ethmoid sinus was involved in eight cases, Sphenoid sinus was involved in three cases and frontal sinus was involved in four cases. In our study, eight cases disease present unilateral, in two cases disease present bilaterally. Variable presentations are three presented with anosmia, two presented with eye pain, watering of eyes, two presented with facial swelling, one presented with unilateral frontal headache, one presented with proptosis, one presented with oroantral fistula, and one presented with blurring of vision (Figure 3). Blood profile showed two patients with peripheral eosinophilia. Common findings in diagnostic nasal endoscopy are mucosal edema, thick purulent nasal discharge, two patients had nasal polyposis. In CT PNS common findings are heterogeneous intensities within paranasal sinuses, which is unilateral in eight patients and bilateral in two patient had erosion in lamina papyracea, two had sinonasal polyposis, one had erosion

Table 1: AFRS-case details								
S. No.	Age (years)	Gender	Symptoms	Sinus involvement	Laterality	Procedure	Recurrence	
1.	13	F	Nasal obstruction nasal discharge	Maxillary ethmoid	Bilateral	FESS	Nil	
2.	17	M	Nasal obstruction blurring of vision headache	Maxillary ethmoid frontal	Unilateral	FESS	Nil	
3.	35	F	Nasal obstruction oroantral fistula	Maxillary ethmoid frontal	Unilateral	FESS	Nil	
4.	50	F	Nasal obstruction facial swelling	Maxillary ethmoid	Unilateral	FESS	Nil	
5.	60	M	facial pain	Maxillary ethmoid	Unilateral	FESS	Nil	
6.	27	M	Watering from eye eye pain	Maxillary ethmoid frontal	Unilateral	FESS	Nil	
7.	39	M	Proptosis	Maxillary ethmoid sphenoid frontal	Bilateral	FESS	Nil	
8.	50	F	Eye pain watering from eye	Maxillary ethmoid sphenoid	Unilateral	FESS	Present	
9.	25	F	Frontal headache	Frontal	Unilateral	FESS	Nil	
10.	42	M	Facial swelling	Maxillary sphenoid	Unilateral	FESS	Nil	

AFRS: Allergic fungal rhino sinusitis

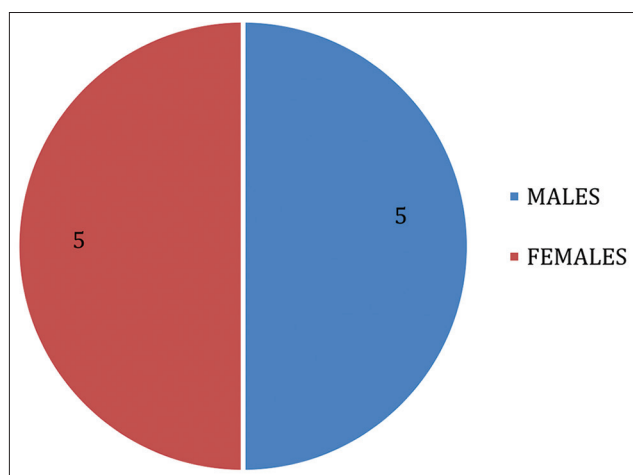


Figure 1: Sex distribution

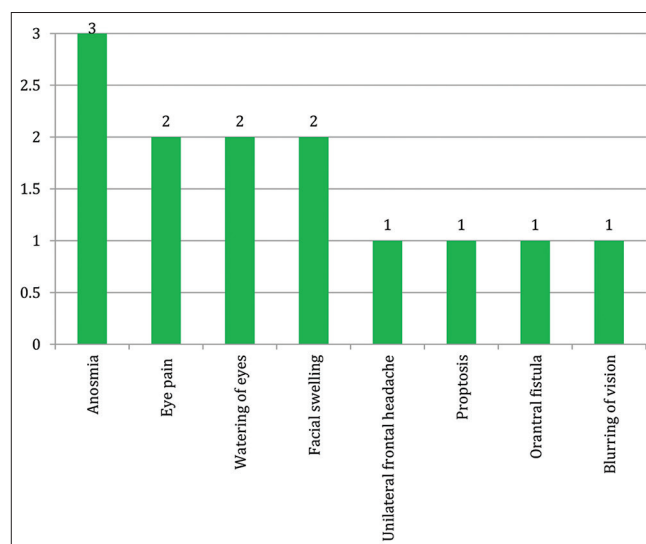


Figure 3: Variable presentations

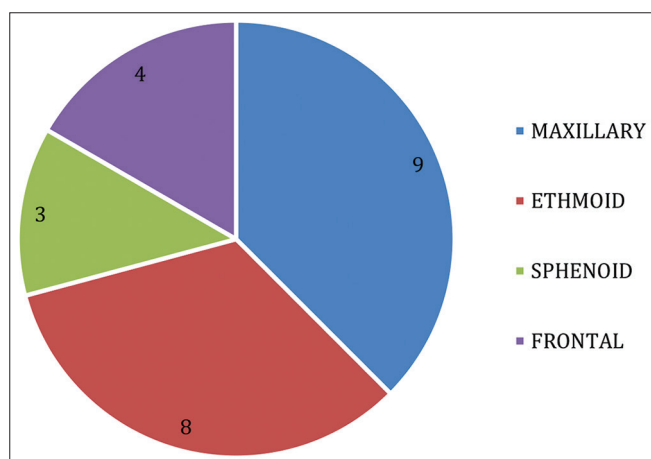


Figure 2: Sinus distribution

2 weeks, 3 months, and 6 months of study, and nine patients showed complete clearance without recurrence, one patient had recurrence and underwent revision surgery and started on anti-fungal Itracanzole for 3 weeks.

The following chart represents sex distribution in patients with AFRS (Figure 1).

The following graph represents sinus distribution in patients with AFRS (Figure 2).

DISCUSSION

Although there are no pathognomonic symptoms, unique patients often present with nasal polyposis and thick yellow-green nasal or sinus mucus.¹⁰ The nasal polyposis may form an expansive mass that causes bone necrosis of the thin walls of the sinuses, it may cause proptosis.¹¹ It may also cause erosion

of medial wall of orbitorbit, and one patient had unilateral frontal sinus involvement. All 10 patients were started on oral corticosteroids, underwent functional endoscopic sinus surgery and oral corticosteroids were continued postoperatively for 3 weeks, were followed up regularly at

of lamina papyracea and medial wall of orbit. Polypoid material can also push the nasal septum into the contralateral airway.¹² CT scans often reveal characteristic serpiginous sinus opacification of more than one sinus, mucosal thickening, and erosion of bone, but this does not represent tissue invasion.¹³ In addition, allergic fungal sinusitis may be suspected when a patient with nasal polyposis, having no other known disease, responds only to oral corticosteroids.¹⁴

Different clinical presentations of AFRS were described in the literature as seen in our patients. This includes intraorbital and intracranial extension, as well as association with other types of fungal sinusitis. Some cases even presenting clinically with epiphora and eye discharge as a result of nasolacrimal gland obstruction.¹⁵ In general, though, patients often complain of symptoms such as nasal obstruction, congestion, purulent or clear rhinorrhea, anosmia, and headache.^{16,17} In our study, only two had orbital complications and no patient had intracranial complications. As for imaging, characteristic computed tomography findings include typical heterogeneity of the signal within involved sinuses, complete opacification of at least one paranasal sinus, and the expansion and attenuation/erosion of the bone with displacement of adjacent anatomic compartments.^{18,19} In our study, patients had similar CT findings except two patients had sinonasal polyposis and two had erosion of lamina papyracea and medial wall of orbit, and one patient had isolated frontal sinus involvement.

In several studies, minor but relevant criteria include a history of asthma, predominance of unilateral disease, radiographic evidence of bone erosion, and peripheral eosinophilia.²⁰ In our study, majority had unilateral disease and two had bilateral disease and one patient had peripheral eosinophilia. In this article, we will review the literature regarding AFRS cases with similar clinical presentation as our cases to assess for the different presentation of the disease. This includes intraorbital and intracranial extension, association with granuloma, skull base as well as orbital erosion has been well documented in the literature with coexistence of both in many cases.^{21,22} The expanding mass leads to bony erosion and involvement of adjacent structures in 6–56% of AFRS patients.²³ Bozeman, et al., reviewed of 34 patients with complications of AFS, this showed that 38% of cases had complications related to orbital wall involvement making it the most common. Erosion of the sinus wall was the second most common complication, occurring in 24%. Ophthalmic complications of sinusitis includes orbital medial wall erosion, proptosis and visual loss in extreme cases.²⁴ In our study, two patients had bony erosions of lamina papyracea and medial wall of orbit. One patient had proptosis, and none of the cases had visual loss although one patient had blurring of vision as presentation.

CONCLUSION

Preventing and treating the condition, as well as its spread and complications, are greatly aided by early detection and management of AFRS. The key to enhancing the result is surgical intervention (endoscopic sinus surgery) and medical therapy (corticosteroids). Longstanding unilateral sinusitis should always raise suspicious of AFRS.

ACKNOWLEDGMENT

We wish to thank the Head and all faculties of Department of ENT for the encouragement and support to carry out the research work.

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Source of Support: Nil, **Conflicts of Interest:** None declared.