

## Allergic fungal sinusitis

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### Abstract

Allergic Fungal Sinusitis (AFS) is believed to be an allergic reaction of the sinus mucosa to environmental fungi that is finely dispersed into the air. We present a 23 year old man who presented to us with a unilateral nasal mass with foul smelling discharge and headache. Functional Endoscopic Sinus Surgery (FESS) was performed. Microbiology and histopathological examination of the mass confirmed it to be a case of AFS.

**Key words:** Allergic fungal sinusitis, Nasal polyps, FESS

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In the early 1980s, Katzenstein described Allergic Fungal Sinusitis (AFS), a sinus process clinically and histopathologically similar to allergic bronchopulmonary aspergillosis<sup>1</sup>. AFS is a form of chronic sinusitis characterized by nasal obstruction, sinus pain, rhinorrhoea, and frequent orbital symptoms. Although clinicians have begun to recognize this disorder in patients with refractory chronic sinusitis, most authorities feel that the disorder remains underappreciated<sup>2</sup>. We report a case of AFS that was treated with FESS and corticosteroid therapy.

### Case report

A 23 year old male reported to us with a history of left sided nasal obstruction, mucopurulent rhinorrhoea and headache of two years duration. ENT examination revealed a whitish mass arising from the middle meatus extending to the floor of the nasal cavity. On probing the mass was firm to hard in consistency and bled on touch and was inconsistent with the feel of a polyp.

Computed Tomography (CT) scan was done and it revealed a mass filling the right maxillary sinus and extending into the nasal cavity through a bony defect in the lateral wall of the nose. A contrast study was done and it revealed a mass with increased attenuation and the presence of charcot laden crystals filling the left maxillary antrum and extending into the left nasal cavity (Fig 1). A complete blood picture was done and it revealed an absolute eosinophil count of 1600 cells per cu.mm. ESR was normal.

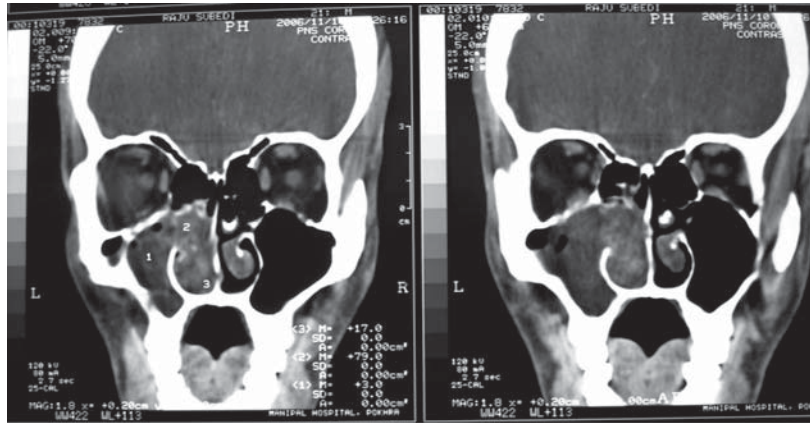
With a provisional diagnosis of AFS, FESS was performed. The mass in the nasal cavity was removed piece meal as it was very friable. The antral extension

of the mass was easily delivered due to the presence of a very wide accessory ostium and demineralization of the uncinat process (Fig 1). The mucosa of the maxillary sinus appeared normal. Anterior ethmoidectomy was performed. As there was no radiological or intraoperative evidence suggestive of extension into the posterior sinuses, they were not opened. The thick allergic mucin which was aspirated during surgery was sent to the microbiologist for fungal culture and the specimen was sent to the pathologist for histopathological examination. The nasal pack was removed after 48 hours and the patient was discharged on the 3rd post operative day. Post operatively he was advised oral antibiotics, saline nasal douching and nasal decongestants. After a week oral prednisolone (60mg divided in three doses) was started which was tapered over a period of 4 weeks. The patient was reviewed at weekly intervals where he underwent nasal endoscopy and cleaning to remove the crust and debris. Fungal culture grew *Aspergillus flavus* from the allergic mucin, and the pathologist reported it as a fungal polyp as abundant eosinophils, Charcot laden crystals (dead eosinophils) and fungal hyphae were seen on microscopic examination. At eight weeks of follow up he was asymptomatic. There was no evidence of recurrence or residual disease clinically, endoscopically and radiologically as confirmed by CT (Fig 3).

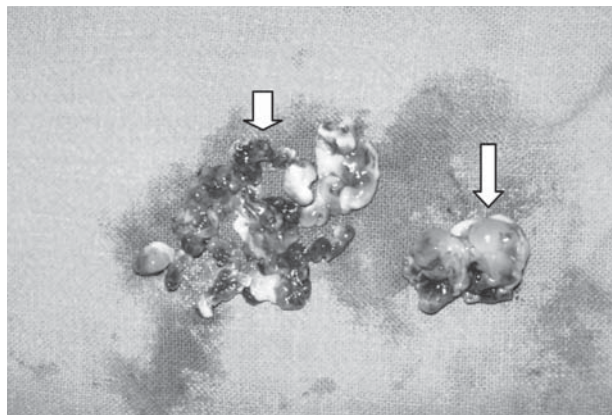
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### Correspondence

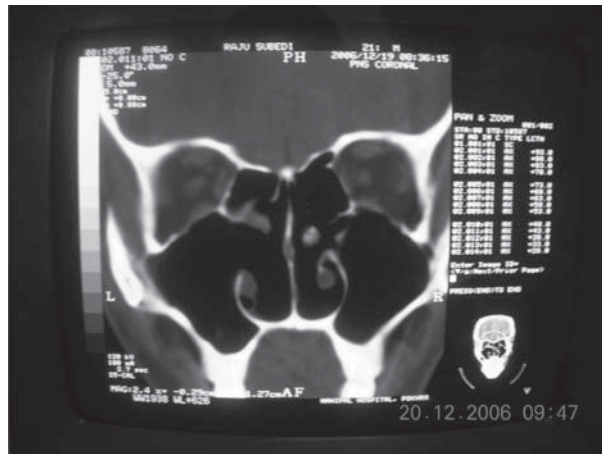
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**Fig 1:** Pre op CT showing mass in the nasal cavity and the maxilloethmoid complex with areas of hyperattenuation.



**Fig 2:** Photograph showing the removed specimen from the nasal cavity (small arrow) and the maxillary sinus (long arrow)



**Fig 3:** Post op CT showing complete removal of the disease from the nasal cavity and the maxillo ethmoid complex.

## Discussion

The prevalence of AFS in chronic sinusitis patients who require surgery is between 5% to 10%<sup>3,4</sup>. AFS is more widely recognized now because of changes in the fungal taxonomy and improved culture techniques<sup>5</sup>.

AFS is mainly noted in the younger age group, mean age of large series may vary from 23 to 42.4 years<sup>2,6,8</sup>. The disease is more common in warm, humid climates<sup>6</sup>. One third to half the patients has asthma<sup>2,6,7</sup>.

Patients with AFS are by definition atopic but they otherwise have a normal immune system and their ESR is normal unless they have a superadded bacterial infection<sup>7</sup>.

Patients with AFS may be similar to other rhinosinusitis patients. Patients present with nasal polyposis and may have previous Sino nasal disease. They may have a documented atopic disease. Seventy five percent of the patients who have AFS give the history of expelling darkly colored rubbery nasal casts<sup>7</sup>.

Bent and Kuhn developed the following diagnostic criteria for AFS which includes (i) Evidence of type I hypersensitivity (ii) Nasal polyposis (iii) characteristic CT findings (iv) Positive fungal stain or culture and, (v) Allergic mucin with fungal elements and no tissue invasion<sup>8</sup>. The incidence of polyposis in AFS is almost 100%<sup>7</sup>. Nasal polyposis is a non specific indicator of chronic nasal inflammation and patients undergoing FESS for polypoidal rhinosinusitis are expected to be afflicted with AFS in 5% to 10% of the cases.

CT scan findings in AFS are characteristic. Central areas of hyperattenuation within the sinus cavity are seen. These central areas represent the proteinaceous allergic mucin. The central attenuation seen on CT may show various patterns including a starry sky, ground glass or serpigenous pattern. Bony loss is common as the expanding inflammation pushes and thins the surrounding bone. Almost half of the AFS patients have unilateral disease, although involvement of the nose and the contiguous sinus is common<sup>9</sup>.

The mucous of AFS is thick and tenacious; its colour ranges from tan to green, brown or black<sup>10</sup>. Fungal species of the dematiaceous species are most commonly the cause<sup>10</sup>. Examination of the allergic mucin with haematoxylin and eosin stain reveals eosinophils, charcot laden crystals and possibly fungal hyphae with a background of eosinophilic or basophilic mucinous material. The charcot laden crystals, consists of lysophospholipase depicted specially well with brown brenn stain<sup>10</sup>. GMS stain typically is used to visualize fungal elements within the allergic mucin.

In order to minimize recurrence of the disease treatment of AFS is directed towards removal of the inciting antigenic material. Three goals should be achieved:

- a. Complete extirpation of the allergic mucin and fungal debris, greatly reducing or eliminating the antigenic inciting factor within the atopic individual.
- b. To impart permanent drainage ventilation of the affected sinus while preserving the integrity of the underlying mucosa. This has been improved greatly by the recent advent of tissue sparing instruments<sup>11</sup>.
- c. Adequate ventilation and drainage also provide for the final goal for the surgery i.e. post operative access to the previously diseased areas. Even under ideal circumstances, small residua of fungus may remain in situ, inciting recurrence if not controlled post operatively.

These three surgical goals can be accomplished through a number of approaches and techniques, the device of which is influenced by the experience and training of the surgeon. Endoscopic powered instruments have demonstrated its effectiveness through the ability of this technique to remove soft tissue and thin bone while maintaining good visibility. Great care should be taken while using these powered instruments, due to the well recognized bone dissolution associated with AFS, it increases the risk of inadvertent orbital or intracranial penetration<sup>11</sup>. In the event of extensive remodeling or bone erosion, image guided systems may be of benefit<sup>16</sup>.

Pre operative steroids may confuse the diagnosis of AFS causing resolution of the typical allergic mucin required for histopathological diagnosis of disease.

Post operative care begins immediately following surgery in the form of saline nasal douching. Weekly visits are required to allow regular inspection of the operated areas as well as debridement of crusts and retained fungal debris if necessary. Systemic steroids are continued post operatively at an individualized dose based on the overall plan of treatment<sup>12</sup>. According to Marple et al the patient is weaned off slowly from pre operative prednisolone doses of 0.5 to 1 mg/kg/day over a period of one month<sup>13</sup>.

The use of topical and systemic antifungal therapy for AFS has been studied by Kuhn and colleagues with mixed to poor results<sup>8</sup>. Few studies showing the effectiveness of antifungals have been published with disappointing results<sup>17</sup>. In general even patients whose symptoms, endoscopy and CT scan findings cleared after systemic antifungal therapy experienced a recurrence immediately after antifungal therapy was discontinued.

Mabry and colleagues have made considerable effort investigating immunotherapy for AFS<sup>13,14,15</sup>. Allergic individuals including those with AFS or allergic rhinitis are injected subcutaneously with small graded doses of allergen against which they are reactive. Of the 11 relevant fungal antigens used for testing and immunotherapy all patients with AFS showed sensitivity to multiple fungal antigens. They stated that immunotherapy produced decreased amount of crusting and polyposis as well as a reduction in the need for topical and systemic corticosteroids in these patients. Immunotherapy research may be a promising direction in which to develop a supplemental treatment option for surgery and steroid therapy of this difficult disease. A lack of availability of the specific fungal antigens would be a major obstacle to progress from an immunotherapy point of view. Additionally, precise fungal identification may be necessary to accurately use this treatment method.

### Conclusion

Diagnosis of AFS required a high index of suspicion. AFS is a newly recognized noninvasive disease that accounts for approximately 5 to 10% of all chronic sinusitis requiring surgical intervention. AFS should be suspected in any atopic patient with refractory nasal polyposis and characteristic radiographic signs. Thick, tenacious, allergic mucin encountered at surgery can be confirmed histologically and hyphae can be demonstrated on special fungal stains or confirmed by a positive fungal culture. Current therapy includes conservative but complete removal of all allergic mucin, which usually can be accomplished endoscopically. Although the use of steroids is controversial, adjunctive systemic steroids are used short term and topical nasal steroids long term. Recurrence of AFS with associated symptomatology is common, necessitating close clinical, endoscopic, and radiographic follow-up.

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