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CASE REPORT

Allergic fungal sinusitis eroding the pterygoid plates: a rare case series☆

Sinusite fúngica alérgica como causa de erosão das lâminas pterigóideas: série de casos raros

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Introduction

Allergic fungal sinusitis (AFS) is a non-invasive form of allergic sinusitis, resulting from the IgE-mediated hypersensitivity reaction to fungal antigen in atopic individuals. It was first described in 1983 by Katzenstein.[1] It usually follows a slow, non-aggressive course, and a wide variety of fungal agents have been implicated, with the vast majority belonging to the Dematiaceae family, such as *Bipolaris spicifera* or *Curvularia lunata*, or to the Aspergillus family, such as *A. fumigatus*, *A. flavus*, or *A. niger*.[2,3] AFS can present with a continuum of symptoms, ranging from simple nasal obstruction to diplopia, facial deformity, osteolytic destruction, and involvement of the skull base.[4] CT scan of these patients is always abnormal, and evidencing bone erosion, chronic rhino sinusitis, and central areas of hyper-attenuation (double density). Incidence of bone erosion varied significantly in different series.[5]

In this article, three cases with erosion of pterygoid plates are presented. Although bone erosions are common and are observed in 20% of the patient with AFS,[5] erosion of the pterygoid plates is a very rare phenomenon, and has not yet been reported in the literature (according to PubMed search). These patient presents with distinct symptoms. Unilateral disease, in addition to bony erosions and typical presentation of symptoms are rare in patients with AFS.

The pathophysiology, clinical features, and detailed management of these three patients are discussed.

Case report

Two of the patients were female, and one was a male in the age group of 18–30 years. They presented to this outpatient clinic with the history of unilateral nasal obstruction, rhinorrhoea, repeated sneezing episodes, and headache, with median duration of symptoms from eight to nine months. All the patients were residents of Makkah, Saudi Arabia.

On inquiry, these patients also provided history of dull aching, unilateral, ill-defined, intermittent hemifacial pain,

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and the pain was exacerbated with chewing and opening of mouth, with no relieving factors. Upon endoscopic examination, multiple nasal polyps were seen, with unilateral involvement. No local temporomandibular joint tenderness was present.

All these patients were investigated with computed tomography (CT) scan of the paranasal sinuses in the axial and coronal planes, serum IgE levels, differential leukocyte count, absolute eosinophilic count, and fasting sugar levels. The surgically removed (endoscopic) nasal polyps and intersinus debris were sent for histopathological examination and fungal culture, which yielded *Aspergillus terreus* in one case and *A. fumigatus* in the other two cases.

CT of the paranasal sinuses revealed presence of chronic rhinosinusitis, involving multiple unilateral sinuses, with areas of hyper-attenuation in the center.

Erosion of the pterygoid plates was present in the side of hemifacial pain, and no extension of disease was present in infra-temporal fossa.

All the patients were treated with endoscopic clearance of the sinuses, with post-operative steroids and nasal irrigation. Erosion of the pterygoid plates to a variable extent was present in all the three cases.

Post-surgically, all the patients were relieved of unilateral hemifacial pain.

There were no recurrences during the follow-up period of nine months for one patient, and of six months for the other two patients.

Discussion

Fungal sinusitis was once considered a rare disorder, but has been reported with increasing world-wide frequency in the last two decades. Histopathologically, two distinct forms of fungal sinusitis are recognized: invasive and non-invasive. Invasive fungal sinusitis includes acute fulminant, chronic invasive, and granulomatous invasive forms of sinusitis; the non-invasive forms include AFS and fungal mycetoma. AFS is clinically, pathologically, and prognostically a distinct form of rhinosinusitis, and it occurs in atopic immunocompetent individuals. AFS is most common among adolescents and young adults; the mean age of presentation is 21 years. The male-to-female (M/F) ratio is equal.

Patients with AFS normally present with signs and symptoms of allergic rhinitis, or chronic sinusitis that includes nasal congestion, purulent rhinorrhea, postnasal drainage, or headache. The presentation of AFS is often subtle. Patients typically complain of gradual nasal airway obstruction lasting from months to years, and symptoms are usually unilateral.

Pain is an uncommon symptom, but in the present study, all the patients had a history of typical pain. It was a dull-aching, non-localised, hemifacial pain, which was exacerbated by chewing and excessive opening of the mouth; all the movements of mastication caused increase in this pain. This associated symptom has never been described with sinonasal diseases in the literature.

The pathophysiology of AFS has not yet been fully understood; however, it is postulated to be similar to that of allergic bronchopulmonary fungal disease (a term replacing bronchopulmonary aspergillosis). Manning et al. have suggested that several interrelated factors and events lead to the development and perpetuation of AFS.[6] They have provided evidence of role of an IgE mediated reaction. First, an atopic host is exposed to fungi via normal nasal respiration, which provides the initial antigenic stimulus. An initial inflammatory response ensues as the result of both type I (IgE-mediated) and type III (immune complex-mediated) reactions, causing subsequent tissue edema. The resulting obstruction of sinus ostia, which may be accentuated by anatomic factors such as septal deviation or turbinate hypertrophy, results in stasis within the sinuses. This creates an ideal environment for further proliferation of the fungus, thus increasing the antigenic exposure to which the host is allergic. This process leads to further inflammation and obstruction, thus leading to a self-perpetuating cycle; it also leads to collection of allergic mucin, i.e. the secretions which fills the sinuses, which contains fungal hyphae.

The production of this allergic mucin and its eventual clinical, histologic, and radiographic characteristics are unique to AFS and serve as a hallmark of the disease. Grossly, allergic fungal mucin is thick, tenacious, and highly viscous. Often described as having a peanut butter appearance, accumulation of allergic fungal mucin eventually leads to the increasingly well-recognized radiographic findings characteristic of AFS, i.e. heterogeneous areas of signal intensity within the paranasal sinuses on CT scans, although these findings are not specific for AFS.

The areas of high attenuation are seen due to the collection of heavy metals (iron, manganese), along with calcium crystals in the inspissated mucin.[6]

Expansion, remodeling, or thinning of the involved sinus walls is common in AFS, and is thought to be caused by the expansile nature of the accumulating mucin, which also leads to the bony erosions seen on CT scan. The bone resorption is presumably caused by cytokines present in the allergic mucin.

Different criteria have been proposed for the diagnosis of AFS, out of which Bent and Kuhn criteria are widely accepted.[5] In 1994, Bent and Kuhn published specific diagnostic criteria based a on case series of 15 patients, which included five criteria: (1) type I IgE-mediated hypersensitivity; (2) nasal polyposis; (3) characteristic CT findings; (4) eosinophilic mucin; and (5) positive fungal smear or culture.

In 1997, De Shazo proposed similar diagnostic criteria for patients without atopy[6]; Bent and Kuhn's criteria were used for the diagnosis of AFS in all the present patients.

All the present patients had radiological evidence of unilateral pterygoid plate erosions. It is a rare radiological finding; as described above, these patients had typical pain at the side of the pterygoid plate involvement. The authors attribute it to presence of the chronic inflammation in the sinuses and at the site insertion of pterygoid muscles, over the lateral pterygoid plate. This pain can be explained by the erosion of the lateral pterygoid plate. The pterygoid process projects down from the body of the sphenoid bone, and inferiorly it divides into lateral and medial pterygoid plates. The erosion of the pterygoid process from the sphenoid sinus and further extension into the pterygoid plates leads to the chronic inflammatory changes in the site of attachment of pterygoid muscles; this inflammatory process also involves the branches of mandibular division of trigeminal nerve,

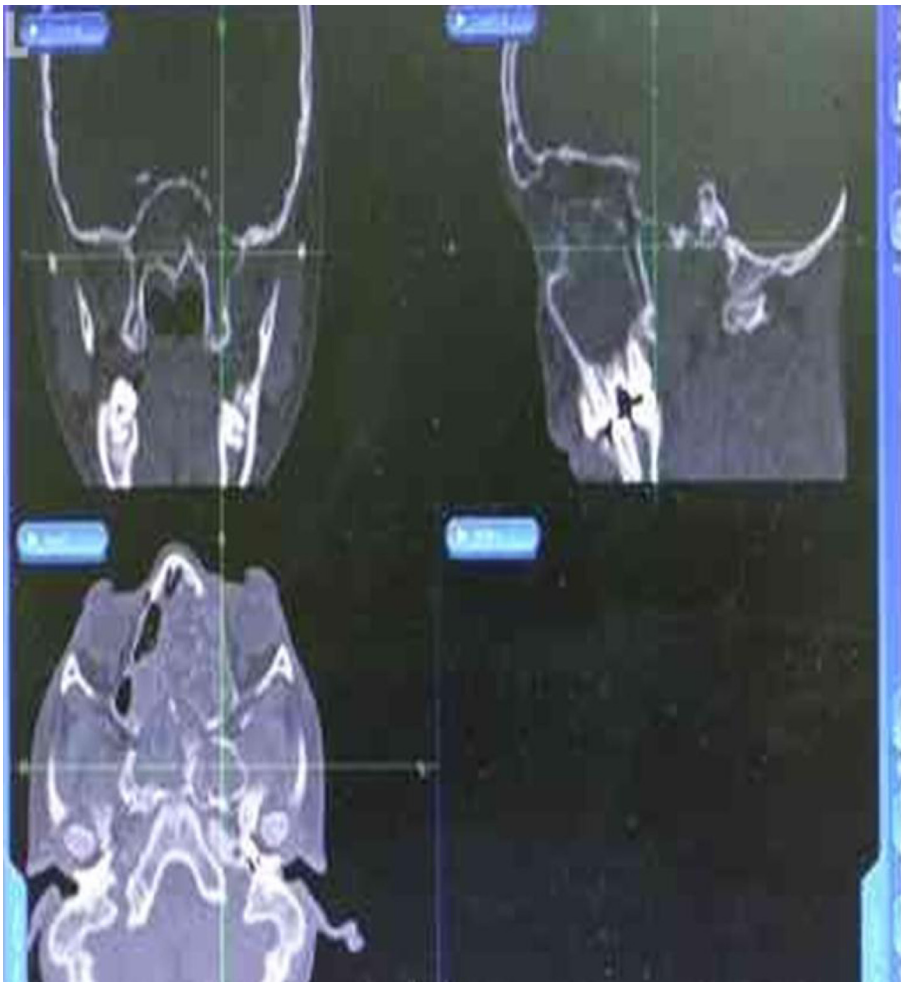

Figure 1 Navigation image showing site of erosion.

present between the media and lateral pterygoid muscles, especially the buccal division. These chronic inflammatory changes of muscles, along with the branches of the mandibular division of the trigeminal nerve can explain the typical pain experienced by these patients.

The authors have termed this as Marglani Syndrome, as the characteristics of this entity were described by the main author (Dr. Osama Marglani), which include unilateral hemifacial dull aching pain, which increases upon chewing and jaw opening in patients diagnosed with AFS with involvement of the pterygoid plates.

Management of these patients involves surgery followed by rigorous medical therapy. All the present patients were treated with functional endoscopic sinus surgery under image guidance. This involves removal of polyps and evacuation of sinuses, with widening of sinus ostia to facilitate sinus drainage, preserving the nasal sinus mucosa as much as possible. It is always difficult to visualize the areas of pterygoid erosion. Medical therapy is required to prevent recurrences. All the patients were treated with intranasal steroid spray. These patients also receive local budesonide irrigation, which delivers a larger dose of topical steroids compared to nasal sprays. A short course of oral steroids is prescribed in post-operative periods.

In postoperative follow-up, all the patients were relieved of unilateral pain. This retrospectively confirms the cause of pain as pterygoid plate erosion with inflammation of the pterygoid muscles.

Close follow-up of these patients is warranted, because recurrent disease may silently progress until the patient develops significant nasal obstruction (Fig. 1).

Conclusion

In an era when diagnosis of bony erosions in AFS patients is always confirmed by radiological investigations, it can still be clinically suspected based on signs and symptoms that are associated with the site of erosions. As this case series was described, the erosions of the pterygoid plates present with a unique set of symptoms, and warrant early surgical intervention, as they may progress to involve the infratemporal fossa.

Conflicts of interest

The authors declare no conflicts of interest.

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