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Allergic fungal rhinosinusitis infiltrating anterior skull base and clivus

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ABSTRACT

Bone erosion and skull base invasion are often suggestive of a malignant mass in paranasal and nasal cavities. Nevertheless, forms of chronic rhinosinusitis, such as allergic fungal rhinosinusitis (AFRS), could mimic malignant features. Here, we report AFRS patient with orbital, anterior cranial fossa, Turkish saddle and clivus erosion. A 48-year-old Caucasian female with history of drug-resistant headache, nasal obstruction and anosmia was referred to our institution. Imaging showed hyperdense featureless tissue with signs of medial orbital wall, cribiform lamina and clivus erosions and encasement of right internal carotid artery. Massive amounts of thick and grayish mucoid material were evacuated during surgery. In case of bony erosion, malignancy should always be excluded. Often the correct diagnosis will be obtained only by operative specimens. AFRS could usually be managed endoscopically. Appropriate medical management of the AFRS should be administered in order to prevent relapses.

1. Introduction

A bone-eroding mass in the nasal cavity and/or paranasal sinuses should be always investigated in order to exclude a malignancy. It is important to note however that a chronic, non-tissue invasive, inflammatory process of the nose and paranasal sinuses, the so called allergic fungal rhinosinusitis (AFRS), could mimic neoplastic features. It represents an immune hypersensitivity to extramucosal fungal antigens, which leads to the production and accumulation of allergic mucin. The presentation of AFRS is usually subtle, consisting of gradually increasing nasal obstruction, nasal crusting, viscous rhinorrea, and hyposmia. Unlike bacterial rhinosinusitis, facial or dental pain and headaches are less common [1,2]. Conversely, multiple nasal polyps are virtually identified in all AFRS patients, with unilateral distribution more common than bilateral [2–4]. The classic and still widely accepted diagnostic criteria for AFRS were described by Bent and Kuhn [3], who proposed the following: type 1 hypersensitivity; nasal polyposis; characteristic computed tomography (CT) scan findings; eosinophilic mucus without fungal invasion into sinus tissue; and a positive fungal stain of evacuated sinus contents. The disease's course is typically indolent and non-aggressive; however, as allergic mucin accumulates within the paranasal sinuses, it may form a mucocele and mimic an invasive process. In 6–56% of patients with AFRS, the expanding mass leads to bony erosion and involvement of adjacent structures [5]. However, investigators soon noted that in some cases, the allergic mucin evacuated from the sinuses did not have identifiable fungal elements; these patients were labeled as having an "AFRS-like syndrome" [6]. Additionally, Ferguson [4] proposed the term "eosinophilic mucin rhinosinusitis" (EMRS) to describe cases in which fungus was not identified histologically. Moreover, she demonstrated that although AFRS and EMRS had identical physical findings, there were clinical differences such as acetylsalicylic acid (ASA) sensitivity was more common in EMRS cases as well as the sinus disease was exclusively bilateral and asthma was present in nearly all EMRS patients.

The primary management for both clinical entities was represented by surgery. The goals of the surgery are:

1. to clear out the edematous mucosa,
2. to create patent sinus ostia large enough to facilitate adequate drainage and ventilation,
3. to marsupialize involved sinuses.

Although effective at removing large portions of the disease, surgery is insufficient in eradicating the relentless inflammation of AFRS. In the absence of medical adjuvant therapy, recurrence rates

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of up to 100% have been reported [2,7]. Here, we report a case of an AFRS with anterior skull base and clivus involvement treated by functional endoscopic sinus surgery (FESS).

2. Case report

A 48-year-old Caucasian female referred to our institution with a month history of drug-resistant headache, nasal obstruction and anosmia. Her past medical history was significant for ASA sensitivity and asthma. Sinonasal endoscopy revealed many large polyps, occluding the nasal cavities, no muco-purulent discharge nor post nasal dripping as well as no neck masses nor pathologic lymph nodes were found. The patient underwent a CT scan that showed hyperdense formless tissue involving all paranasal sinuses with signs of medial orbital wall erosion, reduction of ethmoidal cells thickness, massive encasement of right internal carotid artery in sphenoid sinus, erosion of the cribiform lamina and clivus (Fig. 1). A contrast-enhanced magnetic resonance imaging (MRI) was performed in order to clarify the extension of the mass and to visualize any hypervascularization. This heterogeneous mass did not appear to invade the dura, the pituitary gland, the carotid and basilar arteries; furthermore, a peripheral contrast-enhancement was noted (Fig. 2). The patient underwent an endoscopic drainage, with the neurosurgical team on standby in case an open neurosurgical approach was required. As first step biopsies of nasal polyps were obtained and intraoperatively examined. Inflammatory tissue was shown in all specimens, therefore we performed wide openings of all paranasal sinuses removing a dense and grayish mucin that revealed an intact paranasal mucosa. The intraoperative examination of the mucin demonstrated few eosinophilic cells without any fungal hyphae. This tenaceous material fulfilled each paranasal sinuses involving the medial orbital wall at both sides; after its complete removal in sphenoid sinus, a right internal carotid and optic nerve dehiscences were found. Further, frontal disease was cleared. The operation was free of complications and open approaches were not required. The subsequent pathology report (Fig. 3) indicated that no fungal elements were visualized within the tissue; the mucin was described as an amorphous mucoid material with an extensive collection of eosinophils and neutrophils, Charcot–Leyden crystals were not identified and fungal staining and cultures were negative. Serum levels of immunoglobulins showed low IgE (238 mg/dL) and a IgG deficiency (523 mg/dL) especially for IgG1 (317 mg/dL). The patient was discharged with topic corticosteroid therapy and saline irrigations. At the 3-month follow-up, the patient had healed completely, the paranasal sinus ostia were well opened with excellent functional results and no mucin was present. All of her rhinosinusitis-symptoms had resolved.

3. Discussion

In AFRS-suffering patients, multiple nasal polyps are identified with unilateral distribution more common than bilateral [2–4]. Further, in 6–56% of AFRS patients, the expanding mass leads to bony erosion and involvement of adjacent structures [5]. Although the erosion may be extensive, it is important to note that the disease process itself does not invade tissues. Some postulate that the expanding mucin exerts pressure on neighboring mucosal blood vessels, thus compromising blood supply to the underlying bone. Ischemia ensues, weakening bony structures, which become increasingly susceptible to mechanical stress and necrosis [8]. The location of the disease, as well as the path of least resistance, will determine where expansion occurs. Owing to its innate weakness, the lamina papyracea is the most common location for bony destruction and the orbit is the most common location for extrasinus disease spread. When allergic mucin extends into the orbit, the patient often presents with ocular signs ranging from proptosis to visual field loss [5]. In advanced presentations, lesions may erode the skull base, leading to intracranial presence of mucin.

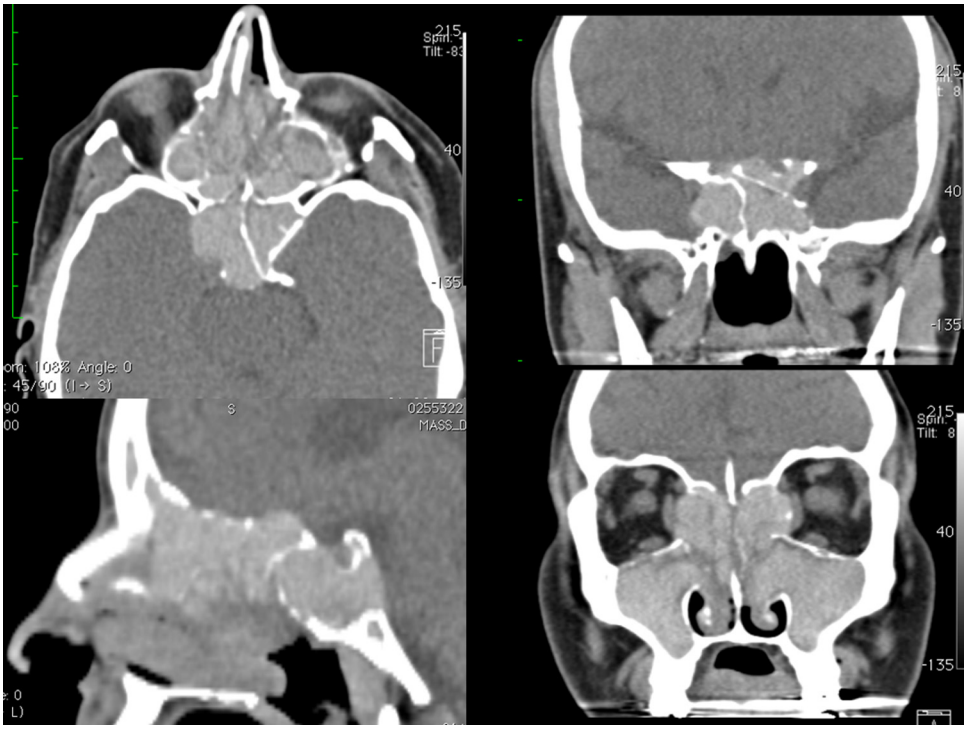

Fig. 1. CT scan shows hyperdense formless tissue involving all paranasal sinuses, medial orbital wall erosion, reduction of ethmoidal cells thickness, massive encasement of right internal carotid artery in sphenoid sinus, erosion of the cribiform lamina and clivus.