INTRODUCTION

Skull base osteomyelitis is a locally destructive disease that is caused by bacterial or fungal infections typically originating from the external ear or sinonasal tract. (1, 2)

Rhinoscleroma is a chronic inflammatory condition of the nasal mucosa that is caused by a gram-negative, aerobic, encapsulated, pleomorphic bacillus, Klebsiella rhinoscleromatis. It is most common in the Mediterranean basin. (3)

Rhinoscleroma typically presents as an asymptomatic swelling that may progress to cause obstruction or sinusitis. (3)

The clinical course of rhinoscleroma can be slow and may extend over many years. (3)

Diagnosis is typically made by biopsy, which may show characteristic findings such as granulomatous inflammation with caseous necrosis. (3)

Approximately 70% of patients will respond to treatment with antibiotics. (3)

CASE REPORT

This 57 year old Haitian male with a history of poorly controlled diabetes mellitus presented with facial sinus pain and progressive right-sided visual loss for several weeks.

CT sinus imaging was consistent with extensive pansinusitis. In addition, there was also noted to be a small area of optic nerve dehiscence in the right sphenoid sinus.

Despite a prolonged treatment course of oral antibiotics the patient continued to have progressive visual loss, sinus disease, and development of new onset right sided periorbital swelling and headaches.

Patient underwent decompensated endoscopic sinus surgery. A right maxillary antrostomy revealed a purulent filled cavity. A right total ethmoidectomy and sphenoid sinusotomy were also performed. The mucosal linings of these sinuses were found to be extremely edematous and hyperemic however without signs of mucosal ischemia, necrosis, or mucopurulent drainage.

Intraoperative cultures and specimens obtained failed to reveal any causative organisms.

Patient continued to have progression of visual loss. Ophthalmologic examination of the affected eye revealed decreased visual acuity of 20/200. Horner’s syndrome, diplopia and restriction on lateral gaze. Hypoalgesia in the V1, V2, and V3 distribution were also noted.

Histopathology demonstrated the presence of thick fungal elements stained with hematoxylin and eosin has been shown to be an effective tool in rapid diagnosis of invasive fungal disease with sensitivities and NPV approaching 84% and 72% respectively when compared to the gold standard of permanent section. (16)

The invasive nature of mucor infection creates an acidic environment leading to direct tissue hypoxia. These fungi invade to arterial blood vessels, eventually leading to arterial occlusion and subsequent tissue necrosis. Bony involvement is quite rare and often occurs late in the disease process. (11)

The patient underwent combined endoscopic sinonasal and transcranial debridement in an attempt to resect all grossly diseased tissue were possible. Transnasal endoscopic debridement consisted of resecting remaining ethmoid and sphenoethmoidal mucosa along the right medial orbital wall and within the right maxillary and sphenoid sinuses. Multiple staged transcranial debridements were required to obtain optimal resection.

Adjuvant postoperative management consisted of an 8 week course of IV amphotericin B and strict blood glucose management.

DIAGNOSIS

The patient underwent dural and skull base biopsies through trans-frontal cranietomy approach with use of stereotactic image guidance. The raised frontotemporal bone flaps as well as the floor of the temporal fossa revealed an infectious process with frank purulence and invasive disease. Inspection of dura and epidural space also revealed gross infection with through and through thickness involvement. Histopathology demonstrated the presence of thick fungal elements consistent with a diagnosis of invasive skull base Mycetoma.

TREATMENT

The patient underwent combined endoscopic sinonasal and transcranial debridement in an attempt to resect all grossly diseased tissue were possible. Transnasal endoscopic debridement consisted of resecting remaining ethmoid and maxillary bony partitions, performing a right fronto sinusotomy, and stripping the mucosa along the right medial orbital wall and within the right maxillary and sphenoid sinuses. Multiple staged transcranial debridements were required to obtain optimal resection.

Adjuvant postoperative management consisted of an 8 week course of IV amphotericin B and strict blood glucose management.

REFERENCES


2. Harrill et al described ophthalmologic symptoms and subsequent MRI imaging led to a confirmed tissue diagnosis by way of transcranial dural biopsy. (3,4)

3. Poorly controlled diabetes has been strongly linked with cases of mucormycosis, and is present as a co-morbid condition in 60-80% of affected individuals. (5)

4. Harrill et al described ophthalmologic symptoms as an initial finding in up to 72% of patients with chronic mucormycosis, representing the most common presenting symptom. (8)

5. The invasive nature of mucor infection creates an acidic environment leading to direct tissue hypoxia. These fungi invade to arterial blood vessels, eventually leading to arterial occlusion and subsequent tissue necrosis. Bony involvement is quite rare and often occurs late in the disease process. (11)

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7. Prognosis is largely dependent on the efficacy of host immunity. In one study analyzing patients with paranasal sinus mucormycosis Bitzer et al demonstrated a 75% survival rate for immunocompetent host, 60% for those with diabetes, and only 20% for patients with other immunocompromised states. (6)

8. Effective treatment involves prompt diagnosis with immediate surgical aggressive debridement, intravenous liposomal amphotericin B, and correction of underlying compromised immunity where possible.