An Isolated Orbital Fungal Granuloma: A Rare Case Report

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Abstract

Isolated orbital fungal lesions is an uncommon entity as the already reported cases are secondary to the involvement of paranasal sinuses especially in immunocompetent healthy individuals. Even though it is rare and the paranasal sinuses are the primary site of inoculation of the infective organism, it is a serious infection that may first present to an ophthalmologist. Here, we report a case of an isolated orbital fungal granuloma of a 42-year-old immunocompetent female who exhibited protrusion and watering of the left eye with no vision loss. The patient was diagnosed with an ovoid well-defined solid lesion in the medial extraocular compartment of left eye in magnetic resonance imaging orbit and a hyperdense lesion with erosion of medial orbital wall in computerized tomography nose and paranasal sinus. The patient underwent endoscopic exploration with biopsy which revealed the lesion to be necrotizing fungal granuloma. The patient was put on oral antifungals over a course of 3 months. There was a complete resolution of symptoms at the end of 1-year follow-up.

Keywords: Aspergillosis, fungal, orbital, proptosis

INTRODUCTION

Aspergillosis belongs to the category of systemic mycoses and primarily affects the nose and paranasal sinuses in head and neck. Though it is rare in immunocompetent individuals, invasive aspergillosis in the immunocompromised patients shows increased incidence and has only increased over the past 2 decades. Orbital involvement not only worsens the prognosis but are also associated with high mortality because of further intracranial spread to the ready available pathways, such as superior orbital fissure or optic canal which directly open into the middle cranial fossa. It thus becomes vital to diagnose fungal disease at the earliest and start appropriate therapy. Surgeons, especially ENT and ophthalmologists, should recognize the clinical spectrum of disease and the wide range of presentation, as early diagnosis and rapid institution of classic approach to therapy including local treatment, debridement and systemic amphotericin B with oral antifungals are crucial in the management of invasive aspergillosis. Here, we present such a case that presented with an isolated orbital invasive aspergillosis.

CASE PRESENTATION

A 42-year-old female with poor socioeconomic status consulted the outpatient department of otorhinolaryngology of a tertiary care center with chief complaints of protrusion of the left eye associated with swelling in the left eye and watering for 6 months. Eyeball protrusion was present and was neither associated with double vision nor with visual deterioration. There were no associated nasal complaints such as obstruction, epistaxis, nasal discharge, and anosmia. She had no history of other known medical illnesses for which she was on treatment. No history of any previous surgeries. Anterior rhinoscopy examination revealed a midline nasal septum with left inferior turbinate hypertrophy and pale nasal mucosa. On Ophthalmology examination, the orbital margins were free with no palpable mass. Fundus examination was also normal. Extraocular movements were affected and visual acuity was normal. There was axial proptosis with restricted extraocular movements in the lateral direction. Diagnostic nasal endoscopy revealed normal findings with a boggy middle turbinate on the left side. Ear and throat examinations were found to be normal. All other systemic examinations were normal. Plain computerized tomography (CT) of nose and paranasal sinus showed hyperdense lesion with mild thinning and erosion of medial orbital wall and was seen in continuity with ethmoid sinus hyperdensity. Magnetic resonance imaging (MRI) (Figure 1) of orbit revealed a well-defined ovoid solid lesion in the medial extraconal compartment in left orbit. Lesion exhibits T1 isointense signal and T2 hypointense signal with mild enhancement with lesion displacing the medial rectus muscle laterally and causing proptosis.
The patient was taken up for Endoscopic orbital decompression with biopsy of orbital mass (Figure 2) under general anesthesia. Intraoperatively, unhealthy mucosa was in the left ethmoid region with a hard mass seen adherent to the periosteum of left orbit abutting the medial rectus muscle and extending both superiorly and inferiorly. Uncinectomy along with anterior and posterior ethmoidectomy were performed. Sphenoidotomy was done and maxillary ostium was widened. The biopsy specimen (Figure 3) was sent for histopathological examination. Postoperative period was uneventful. Biopsy report revealed fibrocartilagenous tissue partly lined by ciliated columnar epithelium stroma showing fibrosis along with foreign body giant cells, epithelioid granuloma and necrosis more in favor of tuberculosis and fungal etiology (Figure 4). On a special acid-fast bacilli stain, tuberculosis was ruled out and fungal elements were confirmed using periodic acid-Schiff stain. Further evaluation with skin prick test for fungal antigens showed hypersensitivity to Aspergillus species. Patient was discharged with oral itraconazole 100 mg twice daily and continued over a period of 2 months. At 1-year follow-up, the patient did not show any symptoms of recurrence.

**DISCUSSION**

Fungi are ubiquitous eukaryotic organisms found in dead, decaying vegetative matter, soil, and air. Among the various fungal infections, the most common orbital infections encountered are mucormycosis and aspergillosis. Mucormycosis is caused by fungus of the order Mucorales, of which Rhizopus species is the most common. Fungus of the order Eurotiales and genus Aspergillus causes Aspergillosis. Initial site of involvement in both forms is usually the paranasal sinuses especially sphenoidal and ethmoidal with secondary involvement of the orbit.
Aspergillosis is a common primary or secondary fungal infection. As it lacks keratolytic enzymes, it cannot actively penetrate undamaged and intact mucus membrane or skin. The pathologic response varies with the severity of the infection, local and systemic immunologic and physiologic state of the host. In healthy patients, the disease is usually chronic and insidious in onset, whereas immunocompromised patients can have a fulminant presentation occurring in the form of cutaneous, sino-orbital, pulmonary, central nervous system, or disseminated infection. It can be associated with foreign body entry as it occurs in eye, bone, burn wounds, prosthetic valves, or can result in local tissue damage.

The primary risk factors for invasive aspergillosis especially in immunocompromised patients are neutrophil defects and corticosteroid use. Various other predisposing factors include human immunodeficiency virus infection, diabetes mellitus, use of prosthetic devices, trauma, or hematological malignancies. There were no such risk factors in our case.

Orbital involvement in the form of vision loss occurs as there is contiguous spread of the disease from paranasal sinuses, by expansion or bone erosion. This is due to the pressure effect of the polyps or fungal tissue invasion. The extension of the infection occurs either by direct spread or through osseous structures like the lamina papyracea or through hemogenous spread by valveless venous plexus to the orbit, brain, or skin.

Aspergillus fumigatus and Aspergillus flavus are the most common fungal contaminants of the sinuses. Most common species affecting immunocompromised patients is A. fumigatus while A. flavus is common in immunocompetent. Although the organism has a characteristic microscopic appearance, culture is the gold standard for identification. This fungus is hematoxophilic with 45° branching septate hyphae which are 2-4 mm wide, best seen on Gomori methenamine silver and periodic acid-Schiff stains. On periodic acid-Schiff stain, intracytoplasmic budding fungal stains. On periodic acid-Schiff stain, intracytoplasmic budding fungal stains. In contrast to neoplasms and bacterial infections

Computerized tomography scans show heterogeneous soft tissue masses with calcification because of the presence of iron, calcium or manganese in the fungal concretions and also bony erosion. In MRI, there is contrast-enhancing masses that are hypointense both on T1- and T2-weighted images in contrast to neoplasms and bacterial infections that show hyperintensity on T2-weighted images. Fine-needle aspiration biopsy is helpful in diagnosing orbital aspergillosis, if the case is undermined to suffer surgery. In Sabouraud’s dextrose agar, aspergillus shows fumigatus colonies as gray-green, flavus as yellowgreen, and niger as black.

At present there are no universal guidelines for the treatment of invasive aspergillosis. The treatment of invasive aspergillosis is considered to be surgical and medical. Conventional therapy suggests the use of systemic amphotericin B followed by oral azoles which includes mainly itraconazole and voriconazole. Some authors recommend exenteration in all patients with retrobulbar or apical involvement while others suggest less aggressive debridement. In our patient, surgery was the first step followed by oral azoles which showed marked improvement. Debridement surely helps in reducing the fungal load and improving delivery of and response to antifungal remedy. We conclude that prolonged conservative medical therapy alone can achieve high cure rates without recurrences.

CONCLUSION

Fungal infections of the orbit, even though rare, are a cause of significant morbidity and mortality, especially in immunocompromised individuals, thereby challenging the diagnosis and early institution of therapy. Newer techniques in discovery and safer modalities of antifungal treatment have brought in a paradigm shift in the mode of management from aggressive surgery to sight and globe-conserving measures. Once the diagnosis is established, endoscopic debridement of disease followed by appropriate oral antifungal therapy appears to be modality of treatment.

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