

Simultaneous Primary Bilateral Orbital Invasive Aspergillosis: A Rare Presentation

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ABSTRACT

The occurrence of invasive orbital aspergillosis in an immunocompetent individual is rare. To our knowledge, primarily bilateral invasive orbital aspergillosis has not been reported in the English literature. We are reporting this case of primary bilateral invasive orbital aspergillosis, which posed a great dilemma regarding its ideal management, to initiate a discussion and get a feedback regarding the ideal management protocol in such a situation.

Keywords: Orbital, Bilateral, Invasive aspergillosis, Management.

INTRODUCTION

The occurrence of invasive orbital aspergillosis in an immunocompetent individual is rare. To our knowledge, primarily bilateral invasive orbital aspergillosis has not been reported in the English literature. We hereby reported a case of bilateral invasive orbital aspergillosis, which posed a great dilemma regarding its ideal management.

CASE REPORT

A 36-year-old female presented to the Outpatient Department of ENT of Postgraduate Institute of Medical Education and Research, Chandigarh, India, with the complaints of bilateral painless, slowly progressive protrusion of eyeball of 8 months duration, affecting left eye more than right, associated with diplopia and decrease in visual acuity. She denied any history of nasal obstruction, allergy, immunodeficiency or steroid use. Examination revealed bilateral forward proptosis with restriction of eyeball movements affecting all the muscles with predominant involvement of medial rectus bilaterally. Visual acuity was 6/18 on the right side and 6/24 on left side. Nasal examination was normal and fundus examination was also normal bilaterally. Investigations revealed patient to be immunocompetent and radiology in the form of CECT showed bilateral homogenous soft tissue density in bilateral orbits (Figs 1 and 2). The patient was subjected to a nasal endoscopy and biopsy of the lesion under general anesthesia. The histopathology revealed features of invasive aspergillosis. She received a total dose of 2.5 gm of

intravenous amphotericin B in the dosage of 1.5 mg/kg/day and after completion of 2.5 gm of amphotericin B was started on oral antifungal agents in the form of itraconazole in the dose of 10 mg/kg/day in two divided doses. There was no worsening of the symptoms and the proptosis decreased during this period. This was planned to be continued for 6 months but after 2 months of oral itraconazole, patient was lost to follow-up. Later on, we came to know that the patient's condition had worsened and she had stopped treatment and expired. The details related to the mortality are not known.

DISCUSSION

Invasive orbital aspergillosis in immunocompetent healthy individuals is an uncommon entity. Most of the reported cases with sino-orbital aspergillosis are of noninvasive allergic variant. The commonest presenting symptom is painless progressive proptosis and is associated with high degree of morbidity and mortality. There is no consensus on the management protocol.¹⁻⁵

The prognosis of invasive sino-orbital aspergillosis is significantly worse than the noninvasive forms of sinus aspergillosis,^{6,7} likely because of penetration of bone and blood vessel walls, which often cannot be eradicated by surgery given the anatomic location where drug penetration may also be worse. So far, the literature mentions a survival rate of around 30%.⁸

There is no uniformly accepted treatment protocol for such clinical entity. Management usually begins with surgical debridement followed by a systemic antifungal drug



Fig. 1: CT scan (coronal cuts) showing the soft tissue density in both the orbits with sinuses free of disease



Fig. 2: CT scan showing the heterogeneous density in bilateral orbits with normal maxillary, ethmoid sinuses and frontal recess

in the form of intravenous amphotericin B. There are very few reports in the English literature of the management of this entity by itraconazole alone. Massry⁹ and Streppel¹⁰ have successfully used itraconazole in patients with sino-orbital aspergillosis. Combinations of itraconazole and amphotericin have been used but there is *in vitro* evidence of antagonism.^{5,10} A few recommend giving the maximum daily dose of the chosen medication and, after the disease is controlled, prolonged administration of oral itraconazole to ensure eradication.¹¹⁻¹³ It should continue well past any remaining signs of disease. The results achieved after such a protocol resulted in 30% survival rate. Data from various sources suggest that response rates of the different drugs are only 40 to 60%. There is an evidence of resistance to itraconazole as well.¹⁴

The management protocol is usually based on the extent of the disease. In limited disease, surgical debridement followed by oral itraconazole resulted in excellent outcome and amphotericin B and its associated morbidity could be avoided. In more extensive disease, radical surgical debridement, amphotericin B followed by itraconazole resulted in better final outcome. Newer antifungals, like voriconazole, has shown a great promise in sino-orbital invasive aspergillosis but long-term results are awaited.^{15,16}

We were in a real dilemma regarding the ideal management protocol in this case because of the near normal vision as no standard ideal management protocol exists in the literature for such lesions. We ultimately lost our patient which would have been most probably be due to angioinvasion by the fungus, though an autopsy would have been more conclusive. This case is being reported first due to its rarity as it probably is the first case of primary bilateral invasive orbital aspergillosis to our knowledge in English literature, and second to initiate a discussion and get a feedback regarding the management protocol in such a situation.

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