Case Report

Sino-orbital Zygomycosis: an atypical presentation of a rare disease in an immunocompetent child

Pragalatha Kumar A.^{1*}, Vinyasa Kolli¹, Prem Kumar P.², Rajashekar Murthy G. R.¹

¹Department of Pediatrics, Indira Gandhi Institute of Child Health, Bangalore, Karnataka, India

²Department of ENT, Indira Gandhi Institute of Child Health, Bangalore, Karnataka, India

Received: 27 November 2017

Accepted: 26 December 2017

*Correspondence:

Dr. Pragalatha Kumar A.,

E-mail: drprahalad@gmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Zygomycosis is a rare life threatening fungal infection in an immunocompromised child. The clinical manifestations of zygomycosis mainly are rhino cerebral, pulmonary, cutaneous, gastrointestinal, and cardiac disease. The diagnosis of zygomycosis is confirmed by direct examinations of clinical specimens and histopathological examination of tissue. The characteristic feature of Zygomycetes in tissue is, the formation of wide, ribbon-like, hyaline, aseptate or sparsely septated hyphae with wide-angle (approximately 90°) branching. The current report describes a case of sino-orbital zygomycosis in a 16-month-old immunocompetent child with atypical presentation. Histopathological examination (HPE) of the tissue confirmed the diagnosis of Zygomycetes.

Keywords: Atypical presentation, Immunocompetent child, Sino orbital zygomycosis

INTRODUCTION

Zygomycosis refers to a group of uncommon, but frequently fatal mycosis caused by fungi of the class zygomycetes¹. Usually zygomycosis is seen in an immunocompromised host with risk factors like lymphoma, leukemia, neutropenia, on corticosteroids and other long term immunosuppressive therapies. The clinical manifestations of zygomycosis mainly are rhino cerebral, pulmonary, cutaneous, gastrointestinal, and cardiac disease. In this case report an atypical presentation of a rare sino orbital zygomycosis, which is uncommon in an immunocompetent child is discussed.

CASE REPORT

A 16-month-old female child, apparently doing well presented with insidious onset of swelling below and around right lower lid (Figure 1) for 1 month which was

painless gradually increasing in size and no history of fever, redness or watering of eyes or nasal discharge. On admission, the child was conscious oriented and with normal vital signs. On examination a soft ill-defined swelling noticed below right lower eye lid measuring 2 × 2 cm with no signs of inflammation, eye movements and vision were normal. Fundus examination and nasal cavity examination was normal. No other swellings noticed. Rest of the systemic examinations were normal. Routine investigations including peripheral smear examination was normal, X-ray paranasal sinus and orbit showed haziness in right maxillary sinus. Orbital and paranasal sinus CT scan (Figure 2) showed an enhancing soft tissue thickening involving right maxillary and ethmoidal sinus, infero-medial extraconal aspect of right orbit, preseptal region, areas of erosions involving walls of ethmoid air cells, medial wall of orbit, walls of right maxillary sinus. Child underwent excision biopsy and right eye orbitotomy. Histopathological examination of the tissue

revealed intact epithelium, beneath which there were mucinous glands, moderate lymphoplasmacytic infiltrate with few eosinophils and granulomas with giant cells and focal necrosis, also noted fungal hyphae with wide angle branching which is in favor of zygomycetes, the child was started on Amphotericin B for 2 weeks and followed by oral posaconazole for 4 weeks.

Figure 1: An ill-defined swelling below right lower eye lid, before treatment.

Figure 2: CT scan showing a mass filling Right maxillary and ethmoid sinus with extension to right orbital wall.

Figure 3: Improvement after treatment.

Investigations for Primary and secondary immunodeficiency were negative. The Child responded well to antifungal therapy (Figure 3).

DISCUSSION

Zygomycosis is an uncommon, but frequently fatal mycosis caused by fungi of the class zygomycetes¹ seen in an immunocompromised host and are rare in immunocompetent hosts.² Risk factors associated with invasive zygomycosis include immunodeficiency state, diabetes, malnutrition, prematurity or advanced age, and intravenous drug abuse.³

zygomycosis can be diagnosed on the basis of presenting signs and symptoms, imaging studies, direct examination and culture of clinical specimens and histopathological examination of the tissue. Zygomycetes are identified by the characteristic picture in the tissue specimen by the formation of wide, ribbon-like, hyaline, aseptate or sparsely septated hyphae with wide-angle (approximately 90°) branching. The lack of septation and the tendency of hyphae to branch at right angles usually serve to distinguish them from *Aspergillus* species, which are septate and smaller and branch at acute angles.³ In the present case, histopathology, was consistent with zygomycetes.

The early diagnosis and immediate initiation of treatment with an antifungal agent in combination with surgical intervention has proved the favorable outcome of zygomycosis.^{4,5} Conventionally the agent of choice was Amphotericin B, however, the availability of the less toxic lipid formulations backed by clinical data to support their use in zygomycosis, Posaconazole is now the alternate agent of choice based on the evidence and supported by individual case reports of successful treatment in patients with different underlying conditions.⁶⁻⁸ Surgical debridement should always be considered as an early option in management as it improves survival and prevents dissemination. The present case responded well to liposomal amphotericin B and posaconazole following surgical debridement.

CONCLUSION

Zygomycosis is a life threatening uncommon infection in an immunocompromised child, however a high index of suspicion is needed in an immunocompetent host. Early combined surgical and medical management will have favorable outcome.

ACKNOWLEDGEMENTS

Authors would like to thank the Director Dr. Asha Benakappa, faculty and staff of Indiragandhi Institute of child health, for their encouragement, support and guidance. The authors also express their gratitude to Dr. Smitha K. S., Dr. Fairoz P. M. of Prabha Eye Clinic and Research Centre for the surgical management and Dr.

Shweta, Dr. Usha Kini and Dr. Marjorie Correa of St. John's Medical college for their valuable histopathology report.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

REFERENCES

1. Rippon J. Zygomycosis. Mycopathologia. 1988;3:681-713.
2. Kontoyiannis DP. Lewis zygomycosis: update on pathogenesis, clinical manifestations, and management. Infect Dis Clin North Am. 2006;20(3):581-607.
Mantadakis E, Samonis G. Clinical presentation of zygomycosis. Clin Microbiol Infect. 2009;15(5):15-20.
3. Sarosi GA, Armstrong D, Davies SF. Laboratory diagnosis of mycotic and specific fungal infections. Am Rev Respir Dis. 1985;132:1373-80.
4. Petrikos GL. Lipid formulations of amphotericin B as first-line treatment of zygomycosis. Clin Microbiol Infect. 2009;15(5):87-92.
5. Rogers TR; Treatment of zygomycosis: current and new options. J Antimicrob Chemother. 2008;61(1):135-9.
6. Stark D, Milliken S, Marriott D. Rhizopus microsporus var. rhizopodiformis sinus-orbital zygomycosis in an immunosuppressed patient: successful treatment with posaconazole after a complicated clinical course. J Med Microbiol. 2007;56:699-701.
7. Page RL, Schwiesow J, Hiltz A. Posaconazole as salvage therapy in a patient with disseminated zygomycosis: case report and review of the literature. Pharmacotherapy. 2007;27:290-8.

Cite this article as: Kumar PA, Kolli V, Kumar PP, Murthy RGR. Sino-orbital Zygomycosis: an atypical presentation of a rare disease in an immunocompetent child. Int J Contemp Pediatr 2018;5:660-2.