International Journal of Pediatric Otorhinolaryngology 79 (2015) 267–270

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Case Report

To exenterate or not? An unusual case of pediatric rhinocerebral mucormycosis☆

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ARTICLE INFO

Article history:
Received 21 July 2014
Received in revised form 21 November 2014
Accepted 21 November 2014
Available online 3 December 2014

Keywords:
Rhinosinusitis
Pediatrics
Infectious
Mucormycosis
Rhinocerebral
Fungal sinusitis

ABSTRACT

Rhinocerebral mucormycosis (RM) is a rare, potentially lethal fungal infection. Traditional teaching encourages aggressive surgical resection until viable bleeding tissue is encountered, often leading to orbital exenteration, skull base resection, and cerebral debridement, in addition to systemic antifungal therapy. We present a 2-year-old male with acute lymphocytic leukemia undergoing chemotherapy presenting with RM and unilateral orbital and intracranial involvement. After aggressive sinonasal debridement, systemic antifungal and hyperbaric oxygen therapies, he recovered without need for further aggressive tissue resection. We report the successful management of invasive orbital and intracranial RM without orbital exenteration or cerebral debridement.

1. Introduction

Rhinocerebral mucormycosis (RM) is a rare and potentially lethal invasive infection caused by saprophytic fungi of the class Zygomycetes, specifically of the Mucorales order. This disease most often develops in hosts made vulnerable by metabolic derangements such as diabetes, or, immunosuppression, as in hematological diseases [1,2]. Clinical manifestations parallel the rapid progression from sinonasal infection to involvement of the orbit, skull base and brain. Sinusitis might be the earliest expected clinical presentation; the more ominous signs of advanced disease largely reflect the fungi's propensity for angioinvasion resulting in thrombosis and necrosis of affected tissues and direct extension to neighboring structures [1–3].

The early diagnosis and treatment of RM is paramount to decreasing morbidity and mortality. The treatment of RM commonly involves aggressive surgical debridement and systemic antifungal therapy, along with correction of underlying metabolic abnormalities and reversion of immunocompromised states, when possible [3,4]. The degree to which radical surgical debridement of infected tissue plays a role in the successful management of disease is unknown. Traditional teaching encourages aggressive surgical resection until viable, bleeding tissue is encountered [5–7]. This has often lead to orbital exenteration, skull base resection, and cerebral debridement due to the characteristically rapid involvement of these structures. The potential for great and permanent physical and mental disability resulting from such aggressive surgical management presents a significant challenge to the control of RM particularly in the pediatric patient.

2. Case presentation

A 2-year-old male was admitted to an outside facility with 1-day history of right eye swelling and found to be febrile, pancytopenic and septic. This was day 21 of induction chemotherapy for B-Cell Precursor Acute Lymphocytic Leukemia. He subsequently developed a small black eschar on his right medial canthus. One week later the patient was transferred to our institution due to eschar progression and worsening post-septal cellulitis on repeat imaging.

On initial examination there was significant swelling and erythema of the right periorbita with a black eschar in the area of the right medial canthus (Fig. 1). There was minor periorbital swelling noted on the left and a midline ulcer was observed on the

☆ Presented at the Combined Otolaryngology Spring Meetings 2014 in Las Vegas, NV during the American Society of Pediatric Otolaryngology poster session, May 16 & 17, 2014.

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http://dx.doi.org/10.1016/j.ijporl.2014.11.028
0165-5876/

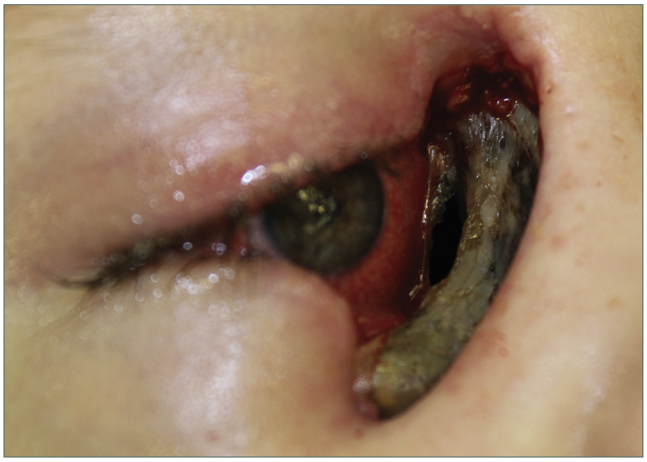

Fig. 1. Right medial canthal wound on initial presentation.

hard palate. The initial computed tomography scan revealed pansinusitis without bony destruction, with pre- and post-septal cellulitis of the right orbit (Fig. 2). Broad-spectrum antibiotics along with Amphotericin B, Posaconazole and Micafungin antifungal agents were started; induction chemotherapy was discontinued.

On hospital day (HD) 3 the patient underwent external incision and drainage of the right orbital post-septal debris, debridement of the right medial canthus ulceration, right middle meatal antrostomy with removal of necrotic tissue and right anterior and posterior ethmoidectomy with debridement of the nasal cavity. Due to expansion of the right medial canthal ulcer the patient returned to the operating room (OR) on HD 7. On endoscopy the right inferior turbinate, the remaining middle turbinate and the superior turbinate and fovea area were all found to be black and necrotic; These structures were debrided as much as possible. The septum was noted to be discolored but no tissue was removed at this time. Endoscopy showed the left nasal cavity to be filled with mucoid tissue and black, gangrenous appearing mucosa; No action was taken at this time. Debridement of the black, necrotic base of the medial canthal ulcer was carried down to bone, and the palatal ulcer was debrided until free bleeding occurred. Magnetic resonance imaging (MRI) on HD 8 demonstrated bilateral inferior-frontal cerebritis with persistence of orbital disease and suspected involvement of the fovea ethmoidalis especially on the right, the cribriform plate, the anterior clinoid and anterior body of the sphenoid (Fig. 3).

Upon return to the OR on HD 9, the right nasal cavity showed extensive signs of acute, invasive fungal sinusitis with extensive black crusting. The right maxillary antrostomy was revised with a right medial maxillectomy extending to the nasolacrimal duct anteriorly, the floor of the nasal vault inferiorly, and posteriorly to include takedown of the posterior maxillary wall as well as the orbital floor. On entering the infratemporal fossa, the adipose tissue displayed non-brisk bleeding which suggested fungal invasion, but not complete destruction, and frozen sections were suspicious for fungal hyphae. However, due to clinical suspicion of limited infratemporal involvement, repeat biopsy showed mature adipose tissue with no fungi. The remaining ethmoid cells were taken to complete a total ethmoidectomy, facilitating removal of the lamina papyracea and exposure of the orbit. Decompression of the orbit revealed necrotic fat and allowed for biopsy of the orbital apex, which would also return positive for mucor infection. The sphenoid sinus was then debrided of necrotic tissue and dark, non-bleeding, necrotic bone was encountered throughout. The nasal cavity floor was necrotic and debridement revealed dark, bony involvement, however it was not as dark as areas previously encountered. Biopsy from the hard and soft palate junction at the margin of debridement would return negative for fungal elements.

From the left nasal cavity, it was observed that the superior and anterior septum was necrotic necessitating total septectomy, extending anteriorly to within approximately one finger width of the columella where fresh, clean cartilage was encountered. The left middle turbinate was necrotic and removed. After a left maxillary antrostomy the sinus appeared to be in good condition with healthy bleeding. The pursuit of involved tissue lead to a left total ethmoidectomy and sphenoidotomy which showed healthy, bleeding mucosa anteriorly and necrotic tissue posteriorly which was removed. The palatal ulcer was further debrided, and biopsies returned positive for fungal involvement. At this time orbital exenteration had been deferred by the family and care team until the extent of intracranial disease could be further evaluated.

On HD 11, MRI confirmed involvement of the inferior frontal lobes and base-of-skull and a palliative right orbital exenteration was planned with the hope that decreasing the fungal burden could affect the patients poor prognosis. However, on HD 14 healthy-appearing, bleeding tissue was encountered in the nasal cavity and yellow, bleeding fat was seen upon re-entering the right orbit. Biopsy of the extra-conal fat and medial rectus muscle would return negative for invasive fungal disease. Recent imaging had demonstrated increased enhancement of the left maxillary sinus yet revision of the maxillary antrostomy revealed nothing of concern. There was continued bleeding without signs of invasive fungal disease throughout the left nasal cavity. The left medial

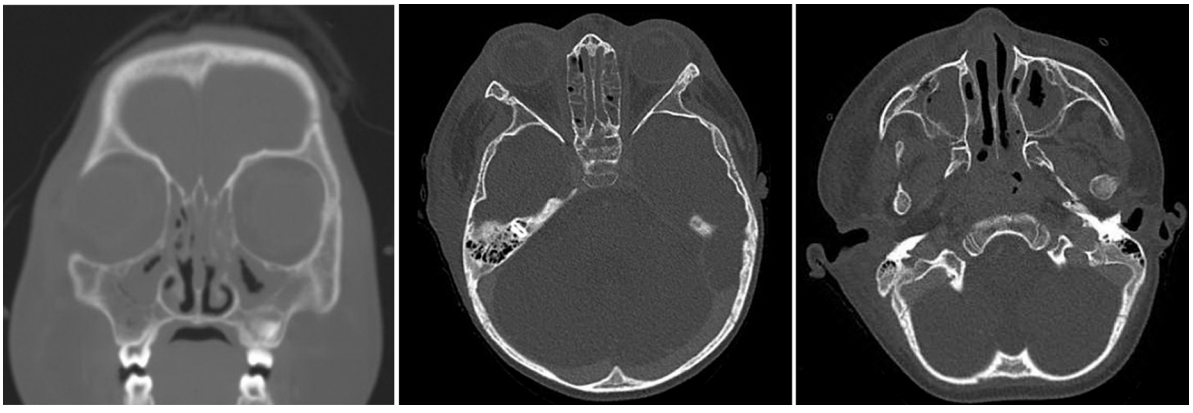

Fig. 2. Initial CT prior to any surgical intervention.