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Rhinocerebral mucormycosis in a child with leukemia: CT and MRI findings

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Abstract. A case of rhinocerebral mucormycosis in a child, evaluated by serial computed tomography (CT) and magnetic resonance imaging (MRI), is presented, demonstrating the severity and rapidity with which the infection extends from the paranasal sinuses to the orbits and ultimately to the brain.

Mucormycosis refers to acute and often fatal infections caused by fungi of the order Mucorales in the class Zygomycetes [1, 2]. The most common pathogens of this order are *Rhizopus, Absidia* and *Mucor* [1–4]. The fungi have a wide distribution and are found in soil, air, decaying vegetation, foods of high sugar content, and bread mould [1, 3, 5]. Mucormycosis is most frequently encountered in patients with poorly controlled diabetes mellitus (80%) [1, 2]; however, it is also seen in immunocompromized patients and a few normal individuals [1, 4, 5].

The five major clinical presentations of mucormycosis are rhinocerebral, pulmonary, gastrointestinal, cutaneous and disseminated [1, 3]. Rhinocerebral mucormycosis is the most common form of the disease [3].

Case report

M. C., a $6^1/_2$-year-old male with B-cell acute lymphoblastic leukemia in clinical remission, presented with epistaxis and thrombocytopenia. The patient subsequently developed fever, headaches, a clear nasal discharge, and proptosis of the right eye. Within 24 h, right blindness developed. A CT scan (Fig. 1) demonstrated pansinusitis, right proptosis, and thickening of the right medial rectus muscle. There was early evidence of right cavernous sinus thrombosis (Fig. 2). MRI study (Fig. 3) obtained later the same day confirmed changes of cavernous sinus thrombosis. Within hours, the patient developed diabetes insipidus, proptosis and absent vision in the left eye, bilateral third, fourth and sixth cranial nerve palsies and speech impairment. The patient underwent urgent antrostomies for culture and decompression.

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Postoperatively, the child developed decorticate posturing and irregular breathing. Serial CT scans obtained within 48 h after operation demonstrated bilateral cavernous sinus thrombosis, occlusion of the internal carotid arteries (Fig. 4), and increasing cerebral edema. The patient was declared brain dead and respiratory assistance was discontinued. Autopsy results confirmed rhinocerebral mucormycosis with (1) invasion of the paranasal sinuses and the nasal and sphenoid bones; (2) infiltration with thrombosis of the internal carotid, anterior cerebral, middle cerebral, and ophthalmic arteries; (3) infiltration of the cavernous sinuses bilaterally with resultant thrombosis; (4) infiltration of the orbital tissues, meninges, and cerebral cortex; and (5) diffuse cerebral edema with tonsillar herniation. There was no evidence of residual leukemia.

Discussion

The Mucorales, saprophytic fungi frequently found in the upper respiratory tract as innocuous spores, are nonpathogenic in normal hosts. When they convert into hyphae and become invasive they can disperse rapidly through the sinuses into the craniofacial structures such as the orbit and the cavernous sinus and then into the brain [1]. Presenting features of unilateral facial pain, orbital cellulitis, proptosis, ophthalmoplegia, and rapid visual loss are highly suggestive of mucormycosis [3]. The pathological hallmark of this disease is vascular invasion and thrombosis with infarction of affected tissues [1]. Internal carotid artery thrombosis is a well-known complication of mucormycosis [1] and in the patient presented was bilateral, resulting in global brain ischemia. Although both central retinal arteries appeared patent, the ophthalmic artery and its branches showed widespread thrombosis, explaining the sudden blindness.

The diagnosis of rhinocerebral mucormycosis is extremely difficult to make since cultures are often negative [1]. In the patient presented, the fungus was only cultured from the nasal secretions postmortem.

Imaging findings in the child presented correlated with CT and MRI appearances of mucormycosis documented in the literature as well as autopsy results [4, 6]. Findings of pansinusitis, orbital spread, subsequent development of cavernous sinus thrombosis and occlusion of the internal

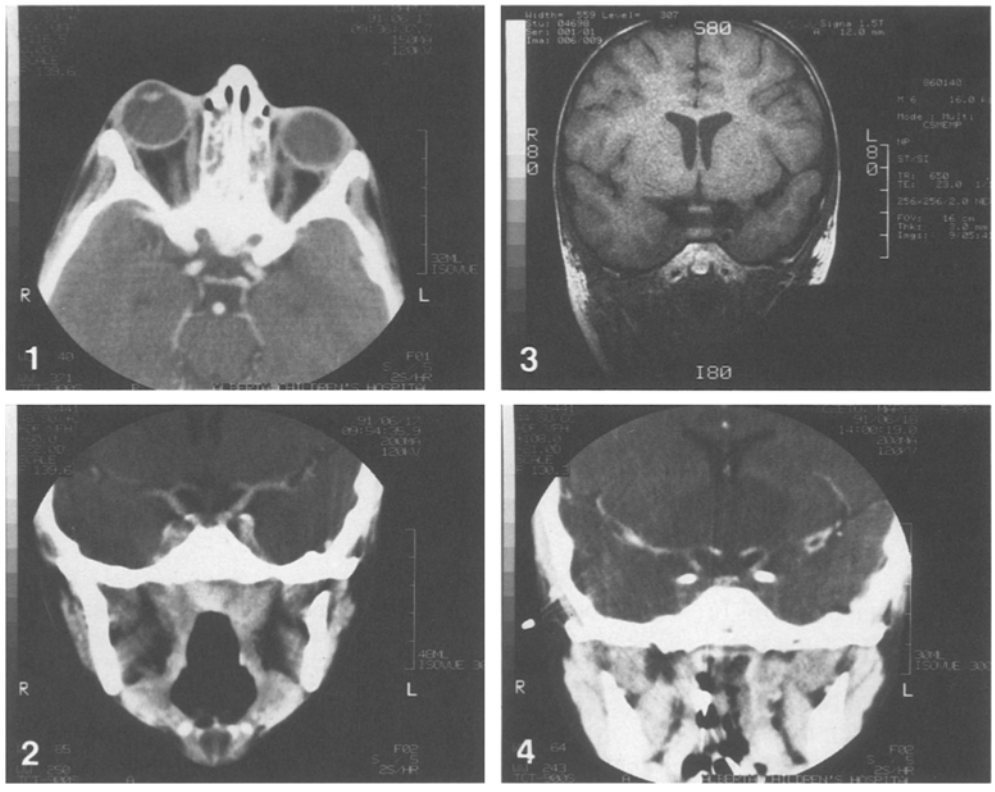

Fig. 1. Enhanced axial CT of the orbits demonstrates right proptosis without evidence of preseptal edema or orbital fat infiltration. The right medial rectus muscle is thickened and slightly laterally displaced. Ethmoid air cells are opacified

Fig. 2. Enhanced coronal CT scan of the cavernous sinuses demonstrates filling defects within the enlarged right cavernous sinus. Findings are indicative of cavernous sinus thrombosis

Fig. 3. Coronal T1-weighted MRI scan (TR 650 ms, TE 23 ms) at the level of the cavernous sinuses demonstrates almost complete absence of the normal signal void within the right intracavernous carotid artery. The cavernous segment of the left internal carotid artery is patent but narrowed. Findings are consistent with evolving internal carotid artery thrombosis

Fig. 4. Enhanced coronal CT scan of the cavernous sinuses demonstrates intraluminal filling defects, thrombi, occupying the cavernous sinuses bilaterally. Hypodensity of the thalami and temporal lobes medially is noted, consistent with infarction

carotid arteries with resultant cerebral infarction were clearly delineated on the serial CT and MRI studies. Both modalities can effectively demonstrate the wide spectrum of findings in rhinocerebral mucormycosis.

Current therapy consists of the administration of antifungal chemotherapy (amphotericin B) surgical intervention (biopsy, debridement of necrotic tissue or intracranial decompression) and control of the underlying immune deficiency [1, 5]. Recently, adjunctive hyperbaric oxygen therapy has been used with favorable outcome [5].

Prompt diagnosis and aggressive therapy may decrease the 40–85% mortality rate currently associated with rhinocerebral mucormycosis [5, 6].

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