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Original article

ENT mucormycosis. Report of 4 cases

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ABSTRACT

Objectives: Mucormycosis is an opportunist infection usually affecting immunocompromised patients. It is rare and often fatal. The pathogen is filamentous fungus of the mucorales order. Studies discuss the clinical, diagnostic and therapeutic aspects of ENT mucormycosis, insisting on early clinical diagnosis, laboratory data not being contributive within satisfactory time limits.
Material and methods: A retrospective study included 4 patients with ENT mucormycosis diagnosed over a 13-year period, from January 2000 to December 2012.
Results: The study included 2 male and 2 female patients, aged from 3 to 77 years. Two patients were diabetic. There were 2 cases of sinonasal mucormycosis and 2 of otologic involvement. Diagnosis was founded on anatomopathologic and mycologic examination. A fatal issue occurred in 1 case with otocerebral involvement.
Conclusions: Otorhinolaryngologic mucormycosis is a rare fungal infection, which needs to be borne in mind. Rhinocerebral lesions are the most common clinical manifestations. Involvement of the ear is very rare. Diagnosis is often difficult, but should be as early as possible. Treatment, initiated urgently, associates antifungal treatment, surgical resection and control of risk factors. The prognosis remains in all cases severe.

1. Introduction

Mucormycosis is a rare opportunistic aggressive fungal infection usually associated with severe immune deficiency. Extension is rapid, with a high rate of mortality. The pathogen is a filamentous fungus of the zygomycete class of the Mucorales order [1,2].

Transmission is air-borne, with predominantly sinonasal tropism (40% of cases). Other locations (digestive, pulmonary or cutaneous) are possible but much rarer [1,3]. Diagnosis is founded on anatomopathologic and mycologic examination [4,5]. Prognosis is severe, depending essentially on early diagnosis and treatment [2,6].

The present study and review of the literature updates clinical, paraclinical, evolutive and therapeutic aspects of ENT mucormycosis.

2. Material and methods

A retrospective study included 4 cases of mucormycosis (2 rhinosinus and 2 otologic), diagnosed and treated in our ENT department between January 2000 and December 2012.

3. Results

3.1. Case 1

A 3-year-old girl, born to first cousins, with history of chronic diarrhea and recurrent bronchopneumopathy, was admitted for right retroauricular swelling of 3 weeks' evolution, associated with fever.

On examination, general health status was poor; fever was 38.7 °C. Height and weight were retarded: weight, 9 kg (−3.5 SD); height, 83 cm (−3.5 SD); and cranial perimeter, 45 cm (−4 SD). Otologic examination found right retroauricular swelling with facing local inflammatory signs. On otoscopy, the external auditory canal showed narrowing and inflammatory walls; the tympanum could not be visualized.

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Mastoiditis in a context of likely immune deficiency was suspected and the patient was put under wide-spectrum antibiotherapy (cefotaxime 50 mg/kg/d + fosfomycin 100 mg/kg/d) associated to fluconazole 6 mg/kg/d.

Biologically, blood count found 19,000 leukocytes/mm^3 (89% PNN; 4% lymphocytes: 760/mm^3). Similar lymphopenia was found on all subsequent counts, suggesting cellular immune deficiency. Protein electrophoresis found hypoalbuminemia (26 gr/L) and hypoproteinemia (47 gr/L) but not hypogammaglobulinemia. Immunoglobulin weight assay was normal and HIV serology negative. Cell immune study found an inverted CD4/CD8 ratio, but this deficit could not be classified.

Cerebral CT screening for intracranial infection was normal.

The patient underwent right mastoidectomy. Intraoperatively, the mastoid cortex was found to be intact; the right external auditory canal had a fistula on the posterior wall, with abundant granulation tissue. Right necrotizing external otitis with mastoid reaction was diagnosed. Histologic study of a surgical specimen found mycelial spores and large ramified filaments without septa, visible on PAS and Grocott staining, suggesting external auditory canal mucormycosis. Direct mycologic examination was negative; culture isolated *Rhizopus oryzae*.

Amphotericin B was initiated in the light of the histology findings, at progressive dose. However, the patient went into anaphylactic shock, requiring transfer to pediatric intensive care, cessation of amphotericin B and resumption of fluconazole.

Subsequent evolution was unfavorable, with onset of right peripheral facial palsy at postoperative day 5 and extension of the retroauricular necrotic lesion toward the parotid space. Temporal CT found right lateral sinus thrombosis (Fig. 1). The patient died 4 days later, from severe sepsis and multi-organ failure.

3.2. Case 2

An 18-year-old male with type 1 diabetes was admitted to the pneumology department with 38.5 °C fever, a poorly-systematized left inferior lobe infection site, ipsilateral purulent pleurisy and ketoacidosis decompensation. Antibiotherapy with cefotaxime 100 mg/kg/d and metronidazole 1.5 g/d was initiated, with pleural aspiration. Evolution showed onset of an ulceronecrotic lesion of the left hemi-palate and left periorbital cellulitis. Examination of a palate necrosis sample found Mucorales filaments. Culture of the palate and pleural samples isolated *Rhisopus arrhizus*. Facial CT and MRI found left maxillary sinus filling with left peripalpebral and jugal soft-tissue thickening and fatty infiltration of the adjacent infratemporal fossa, without cerebral extension.

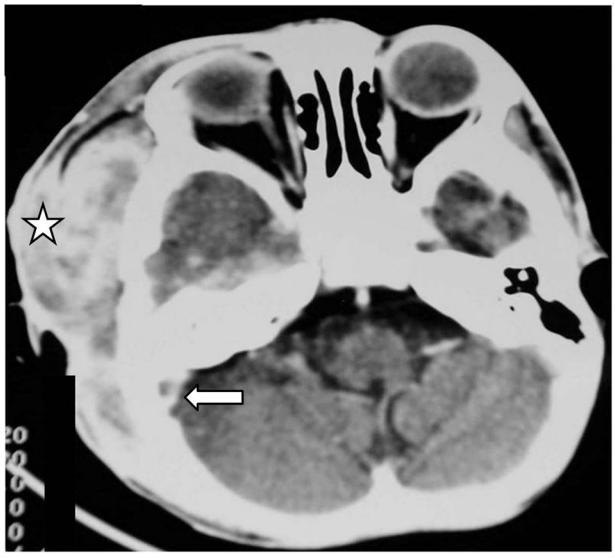

Fig. 1. Cerebral CT, axial slice: severe right temporal soft-tissue infiltration, with intense heterogeneous uptake (☆). Thrombophlebitis of right lateral sinus (⇦).

Amphotericin B was initiated, with surgical curettage of the palatine lesion, performed twice. Evolution showed progressive onset of generalized urticaria due to amphotericin B allergy, confirmed by discontinuation and reinitiation of treatment, and onset of left grade-IV peripheral facial palsy on the House-Brackmann classification. Liposomal amphotericin B was progressively initiated at 3 mg/kg/d for 8 weeks, with good clinical tolerance and improved local and general health status. Posaconazole relay was introduced at 800 mg/d for 2 months. The patient was in good health at the last check-up, 9 months after end of treatment, with good rhinosinus and pulmonary status but persistent grade-II peripheral facial palsy.

3.3. Case 3

A 24-year-old female with type 1 diabetes was admitted for right peritonsillar phlegmon complicated by ipsilateral lateral cervical cellulitis. On admission, she showed 38 °C fever, and was placed under antibiotics (cefotaxime 50 mg/kg/d + metronidazole 1.5 g/d) with reduction of phlegmon and surgical drainage of the cellulitis. Diabetic ketoacidosis decompensation led to transfer to intensive care. Evolution showed persistent fever and glycemic index disorder after 4 days' insulin and antibiotic therapy. ENT check-up found favorable evolution of phlegmon and cellulitis, but nasal endoscopy found a necrotic aspect in the right inferior turbinate, with blackish secretion in the ipsilateral nasal cavity. Mucormycosis was suspected and amphotericin B was initiated at 1 mg/kg/d after inferior turbinate biopsy and mycologic analysis.

Facial CT found ethmoid cell filling with thickened mucosal framework in the right maxillary sinus. Facial MRI found no intra-orbital or neuromeningeal extension.

Pathology found typical Mucorales perivascular angulated filaments without septa; culture isolated *Rhisopus arrhizus*.

The patient underwent complete surgical treatment, with right inferior turbinectomy, ipsilateral ethmoidectomy and right maxillary sinus debridement. Evolution was favorable after 12 weeks' amphotericin B. At 12 months' follow-up, there had been no recurrence.

3.4. Case 4

A 77-year-old male with hypertension was admitted for otalgia associated with left purulent otorrhea and ipsilateral peripheral facial palsy of 15 days' evolution without improvement under antibiotherapy (amoxicillin + clavulanic acid 3 g/d) and short-course corticosteroids (prednisolone 1 mg/kg/d). Evolution showed deteriorated health status with onset of vertigo and vomiting.

Otologic examination found erythematous ulcerated lesions in the left auricle. Otoscopy found purulent otorrhea and a congested external auditory canal, bleeding on slightest contact, with anterior wall polyp and non-marginal subtotal tympanic perforation. The patient showed grade-III peripheral facial palsy. Vestibular examination found a pulsating right horizontal nystagmus and left segmental deviations. Neurologic examination was otherwise normal.

Temporal CT found left middle ear filling, scutum erosion, tegmen tympani lysis, and pneumolabyrinth with stripping of the third part of the facial nerve (Fig. 2). Pure-tone audiometry found total left hearing loss.