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Case Report

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Fatal Rhinocerebral Mucormycosis Mimicking Giant Cell Arteritis (Horton's Disease) In a Diabetic Patient

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Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy

Summary

Introduction

Mucormycosis is an infection with vascular inotropism causing thrombosis. A secondary vasculitis is rare. We report a case of giant cell vasculitis due to a mucormycosis infection. Case It was a 56-year-old man with type 2 diabetes presenting brutal blindness after dental extraction surgery. The initial diagnosis was Horton's disease. The biopsy of the temporal artery revealed a giant cell vasculitis. The patient was treated with corticosteroid therapy. His state was worsening with occurring of bilateral blindness and cutaneous necroses. A mucormycosis was suspected. A cutaneous biopsy of a necrotic zone with a parasitologic analysis confirmed it. Treatment with amphotericin B was initiated, but it did not prevent mortality.

Conclusion: Mucormycosis is a rare infection. The diagnosis is often late. The prognosis remains severe.

1. Introduction

Rhinocerebral mucormycosis is a mycotic infectious disease. It is caused by ubiquitous saprophytic organisms of the subphylum Mucormycotina, including genera such as Absidia, Mucor, Rhizomucor, and Rhizopus [1].

This infection occurs mainly in immunosuppressed, diabetic, and immunocompetent patients weakened by a trauma or a burn. Usually, the vascular involvement in mucormycosis is the arterial thrombosis by invasion from near to near. More rarely secondary vasculitis has been reported [2-4].

We report a case of rhino-Orbito-cerebral mucormycosis complicated with a giant cell vasculitis. The clinical course was fatal.

2. Case report

Mr. A. H. 56-year-old man with type 2 diabetes on oral antidiabetic drugs for seven years, and chronic renal failure for four years, was hospitalized for bilateral blindness in a poor general state.

The medical history began one month ago with mandibular pain and facial edema. A facial mass CT scan revealed left maxillary sinusitis and granuloma of the 27th tooth. A dental extraction surgery was then performed.

Four days after the surgery, the patient developed sudden blindness in the left eye. The ophthalmological examination was normal. The orbito-cerebral MRI was normal too. There was an inflammatory syndrome and hyperleukocytosis. The temporal artery biopsy showed an infiltrated media with lymphocytes, plasmocytes, and histiocytes with giant cells in contact with the internal elastic lamina. The diagnosis of giant cell arteritis (Horton's disease) was retained. The patient was treated with methylprednisolone bolus and high-dose corticosteroid therapy. Unfortunately, he developed blindness in the other eye. The ophthalmological examination showed occlusion of several branches of the central artery of the retina.

On arrival at our department, the patient was in a poor general state. Clinical examination showed bilateral ptosis and bilateral exophthalmia. There were several black and dry lesions without purulent material at the forehead (4 cm on the large axis) (figure 1) and the ear lobes (0.5 cm on the large axis). In biological findings, there was an important inflammatory syndrome with a reactive C protein at 144 mgl, a sedimentation rate at 144 mm (at the first hour), and severe pancytopenia. The myelogram identified a macrophage activation syndrome. Mucormycosis was suspected. A cutaneous biopsy in the necrotic zone showed a large cell thrombotic. Subsequent, direct examination confirmed the

diagnosis. Cultures were positive for Rhizopus oryzae. Treatment with amphotericin B was initiated. Unfortunately, the evolution was quickly fatal, with a multi-visceral failure with death.

3. Discussion

We reported a fatal rhinocerebral mucormycosis in a 56-year-old man with type 2 diabetes mellitus mimicking systemic vasculitis with histologic findings of giant cell arteritis. Mucormycosis incorporates a range of infections caused by Zygomycetes, and rhino-cerebral mucormycosis is usually an acute and fulminant disease that develops first and foremost in the orbital region then progresses from near to near. This localization is the most common (40-49 % of cases). Other localizations of mucormycosis are possible but more seldom (digestive, pulmonary, and cutaneous) [5].

The predisposing factors are mostly chronic pathologies, especially diabetic ketoacidosis decompensation like our patient, severe immunosuppressive conditions such as prolonged neutropenia, steroid or T-cell suppressor therapy, solid organ transplantation or allogeneic hematopoietic stem cell transplantation, cirrhosis, prolonged treatment with antibiotics or cytotoxic drugs, and patients with protein-energy malnutrition, trauma, burns or surgery. Exceptionally, immunocompetent patients may be affected [1,2,6].

Its presentation can be confused initially with those viral infections or sinusitis. In the early stages, the symptoms are not specific (fever, headache, rhinorrhea, etc.). The progression is quick, within a few hours or days, towards necrotic lesions of the oto-rhino-laryngology sphere and involving one or more cranial nerves [7].

Acute sinusitis associated with a blood-tinged nasal discharge and facial cellulitis may be present. Like this observation, various ocular findings may be associated, including proptosis, exophthalmos, ophthalmoplegia, periorbital edema, and chemosis. Facial necrotic eschar and neurologic signs may appear later. Differential diagnoses of lesions should include squamous cell carcinoma, chronic granulomatous infection such as tuberculosis, tertiary syphilis, midline lethal granuloma, and other deep fungal infections [8]. In rare cases specific bacterial infections can cause rapid necrosis tissue lesions in immunocompromised patients [9].

The inflammatory response associated with rhinocerebral mucormycosis is characterized by the presence of neutrophils among large areas of necrosis and ischemia due to mycotic arterial thrombosis. This infection has a recognized vascular tropism. It has a very strong angio-invasive character; it invades the vessels' walls and light, creating parietal necrosis and endoluminal thrombosis. This mechanism can mimic systemic vasculitis, essentially granulomatosis with polyangiitis [10,11]. But true secondary vasculitis is exceptional. Pathologically, the appearance is one of necrosis, perivascular infiltration with polymorphonuclear leukocytes, and hyphae penetration into blood vessel walls, with subsequent thrombosis. The organism grows profusely along the internal elastic lamina and spreads by direct extension through blood vessels to the eye and central nervous system [3]. Exceptionally, granulomas with multinucleated giant cells have been reported in association with mucormycotic infections [12-14]. To our knowledge, only two cases of giant cell granulomatous vasculitis secondary to the rhino-mucormycosis brain occurring in diabetic patients were reported [3,4].

The diagnosis is based on a body of clinical and epidemiological arguments, on the one hand, and histological or mycological evidence on earliest and deep biopsy samples, on the other. The histological examination makes it possible to diagnose by showing short, irregular, very large filaments, measuring from 5 to 20 microns in diameter, not septated with ramifications at right angles. Culturing of biopsy fragments allows the identification of genus and species. The most frequently isolated are Rhizopus (47%), Mucor (18%) and Absidia (5%) [14]. The treatment is urgent. It is based on anti-fungal treatment (intravenous amphotericin B 1-1.5 mgkg per day) and surgical resection of necrotic tissue. Factors for a favorable prognosis are a rapid diagnosis, the use of amphotericin B lipid complex, and extensive ablation of necrotic tissue. It is essential to jointly treat the predisposing factors such as unbalanced diabetes or other underlying comorbidities [16].

The prognosis of mucormycosis remains severe due to immunocompromised terrain and the delayed diagnosis. Even after well-conducted treatment, the results are still disappointing. Survival during mucormycosis decreases from 76% in case of management within less than seven days to about 40% if the delay is longer than two weeks of the first symptoms [17].

This case illustrates the severity of mucormycosis in diabetic patients, the diagnostic difficulties related to the lack of specificity of the clinical features, and its potential to mime systemic vasculitis. It is necessary to know how to suggest the diagnosis and its rapid confirmation by biopsy and the identification of the mucoral by a parasitological examination.

4. Conclusion

Mucormycosis is a life-threatening infection that occurs mainly in immunosuppressed patients and those with diabetes. Despite a loud clinical expression, the diagnosis remains difficult and, therefore frequently late. It relies upon the identification of the agent in tissue or by culture. Management involves anti-fungal treatment (mainly amphotericin B) and sometimes debridement of affected tissues. The prognosis is consequently terrific even if there were recent therapeutic advances and mortality rates remain high.

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