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Chronical rhino-orbital mucormycosis in an immunocompetent host: A case report

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ABSTRACT

Mucormycosis is an opportunistic fungal infection caused by molds within the order Mucorales. The rhino-orbital-cerebral localization is the most frequent. It is a destructive, necrotizing and potentially fatal disease. The treatment involves aggressive surgical debridement combined with antifungal drugs. The course is quickly fatal in the event of delayed diagnosis and / or treatment. This infection usually affects immunocompromised and diabetic patients, but cases of mucormycosis in immunocompetents are increasingly reported. Chronic mucormycosis is extremely rare and affects both immunocompromised and immunocompetent patients, its clinical evolution is nonspecific and its treatment is not standardized.

We report the case of a destructive rhino-orbital and pulmonary involvement in a 59 years old immunocompetent patient who presented a right periorbital edema associated and a vision loss and a notion of nasal obstruction and progressive onset headache four months before admission. Her condition progressed with rapidly extensive necrosis. She underwent extensive surgical resection but soon succumbed to multiple organ failure. The diagnosis of mucormycosis was confirmed post mortem on the excisional piece.

The purpose of this article is to draw attention to chronic mucormycosis in the immunocompetent and to emphasize the importance of early diagnosis and adequate management of this fatal infection.

1. Introduction

Mucormycosis is an opportunistic fungal infection caused by molds within the order mucorales. The rhino-orbito-cerebral localization is the most frequent form (44–49%), followed by the pulmonary and cutaneous localizations (10%), then the gastrointestinal localization and the disseminated form [1]. The causative agents of mucormycosis are saprophytic fungi commonly found in the environment, so the majority of humans are exposed to them daily. The occurrence of the disease usually in subjects with an underlying immunodeficiency pattern is evidence of the efficiency of the immune system against these agents. The rhino-orbital localization is usually manifested by a rapidly invasive acute rhino sinusitis with ophthalmological and neurological signs. In this article we report the case of chronic rhino-orbital mucormycosis in a 59-year-old non-diabetic and immunocompetent patient with a fatal course and whose diagnosis was only made post-mortem. Only a few cases of chronic immunocompetent mucormycosis have been described where the attenuation of clinical symptoms and the chronicity of the course, inconsistent with the classic description of the disease, divert the diagnostic approach away from mucormycosis.

2. Case report

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Our work is a single case report and has been reported in line with the SCARE criteria [2].

A 59-year-old female patient presented to the ophthalmologic and maxillofacial emergency room with painful right periorbital swelling associated with reduced visual acuity, right nasal obstruction and headache, all of which had progressed for four months in a context of feverish sensations. The patient had no particular pathological history, no medication or toxic habits, no notion of trauma or recent dental care. The clinical examination found a conscious, afebrile patient who presented a reddish, firm and painless swelling of the right periorbital and lateronasal region with closure of the palpebral cleft, chemosis, nasal deviation to the left and right nasal obstruction without oculomotricity disturbances or diplopia. A craniofacial CT scan showed uncollected right orbital cellulitis associated with ethmoid and maxillary sinusitis and extension to the soft tissues from the orbito-nasal angle to the eyelid level. Prob-

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Fig. 1. Facial swelling with palpébro-nasal and labial skin necrosis.

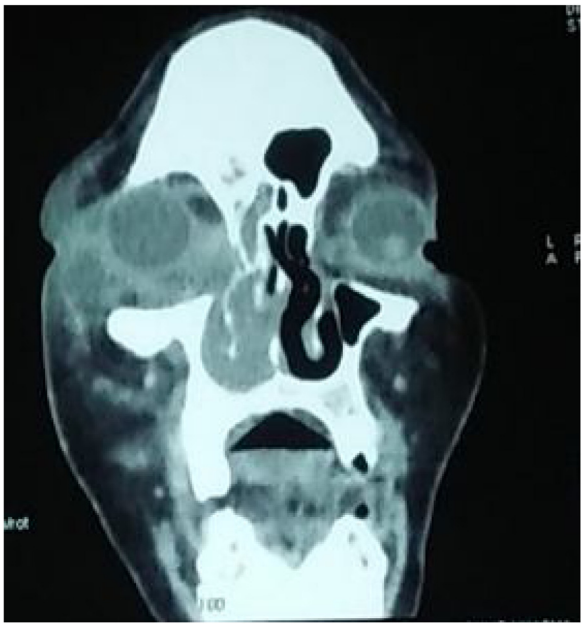

Fig. 2. Facial CT scan: infiltration of the orbital cavity, the nasal cavity and the right ethmoidal sinus.

abilistic antibiotic therapy with Amoxicillin-Clavulanic acid and Metronidazole was started.

When the patient did not improve, a first biopsy was performed on the periorbital fat and the nasal cavity in favor of nonspecific granulomatous inflammatory tissue. The blood count, blood sugar, renal and hepatic functions, plasma protein electrophoresis as well as HIV, syphilis and viral hepatitis B and C serologies were unremarkable, then a granulomatosis assessment was performed, revealing a increased angiotensin converting enzyme (ACE) and bilateral diffuse interstitial lung syndrome.

We performed a second biopsy three weeks later in the maxillary sinus revealing the presence of aspergillary filaments. The evolution was marked by the rapid extension of the swelling with the appearance of plaques of necrosis on the skin and palate (Fig. 1) with dyspnea and total dysphagia.

A second CT scan objectified the lesion extension within the intraorbital level, to the contralateral nasal fossa and the frontal sinus without breaking the base of the skull (Fig. 2).

Based on the clinical, radiological and pathological data, the diagnosis of nasosinus and pulmonary aspergillosis was made and the patient was placed on intravenous Voriconazole (300 mg twice daily). The patient subsequently presented with respiratory distress with multi-organ failure following which the patient was rushed to the operating room where a tracheotomy was performed with an enlarged necrosectomy and exenteration. The patient died 24 h later as a result of her multiple visceral failure. The anatomopathological study performed on the necrosectomy specimen demonstrated the presence of diffuse tissue necrosis extending to the maxillary and zygomatic bones with large non-septate colored mycelial filaments.

3. Discussion

The previously reported case represents an exceptional form of mucormycosis because our patient did not present any risk factor, and due to the chronic course. Mucormycosis is a rare infection caused by a group of filamentous fungi in the orders of Mucorales. It essentially occurs on a particular ground. Risk factors include diabetes mellitus, hematologic malignancies, transplantation of hematopoietic cells and solid organs, immunosuppressive therapy, iron overload and HIV-AIDS [1]. An American study conducted between the years of 2003 and 2010 found that the incidence of mucormycosis is less than 0.01% of all hospitalizations in the United States with only 6.0% of cases of mucormycosis having no identifiable risk factor [3]. The anatomical locations of mucormucosis are mainly rhino-orbito-cerebral, pulmonary, cutaneous, gastrointestinal and disseminated. The mortality rate is around 50% for the rhino-orbital form and can reach 100% in the disseminated form [1,4]. Its high morbidity and mortality is linked to its rapid vascular invasion with tissue necrosis.

Although inhalation is the main way of contamination, responsible for rhino-orbitosinus and pulmonary forms, in immunocompetent subjects the majority of reported cases of mucormycosis were localized to the skin following trauma [5]. The epithelium represents an active barrier against vascular and tissue invasion, the possibility of the development of mucormycosis in an immunocompetent subject can be explained by the presence of an epithelium previously weakened by chromic rhino sinusitis [4,6]. The spores can remain trapped at the level of the nasosinus mucosa. However, if not or badly treated, they can invade the orbit and / or the base of the skull giving the rhino-orbito-cerebral form or pass into the circulation blood and spread throughout the body.

Mucormycosis in immunocompetent patients is rare, with an incidence between 4 and 19% [7]. Both diagnosis and management of this disease are difficult. Several cases have been reported where rhino-orbital mucormycosis has been erroneously treated with antibiotics as being cellulitis [6,8], or by Voriconazol as being aspergillosis [8,9]. While infection spreads rapidly in immunocompromised hosts, it can be slow and chronic in immunocompetent ones. The usual clinical manifestations are exophthalmos, ptosis, diplopia, ophthalmoplegia and reduced visual acuity. The most common early signs are nasal congestion or facial pain [7,10,11].

Diagnosis of mucormycosis is based on histopathology and culture. Mucorals are angioinvasive and necrotic, stain poorly with Gram stain, Grocott-Gomori methenamine silver is the preferred stain [1]. The causative agent identified in 11 to 27% of cases 76]. We can only find a granulomatous inflammatory reaction with the presence of giant cells and polymorphonuclear cells without caseous necrosis [10], which was the case in the first biopsy which only indicated the presence on a nonspecific granulomatosis inflammation.

Amphotericin B is the first-line treatment for mucormycosis. The new generation Triazoles (Posaconazole and Isavuconazole) are used as salvage therapy for patients refractory or intolerant to Amphotericin B. In contrast, Echinocandins and Voriconazole have low activity against Mucorales. Due to the potential for rapid spread of mucormycosis, Amphotericin B should be initiated immediately once the disease is suspected, except that there are no standardized guidelines for the duration of treatment [1,9]. Surgical debridement has extremely important adjunctive roles, considerably improving drug delivery and survival rate [1]. In this case, the diagnosis was misled towards aspergillosis; despite this, the prognosis could have been improved if the medical treatment was based on Amphotericin B instead of Voriconazol since Amphotericin B is the most widely-accepted medication in the treatment of aspergillosis and mucormycosis [12] and if the surgical debridement was performed earlier.

4. Conclusion

Mucormycosis is a rare pathology and it is even more so in immunocompetent patients adopting an atypical and misleading clinical presentation. The essence of the care is based on early diagnosis as well as on multimodal management, both surgical and medical, involving multidisciplinary collaboration.

Declaration of Competing Interest

None.

Sources of funding

None.

Ethical approval

Our study is exempted from ethnical approval.

Consent

Written informed consent was obtained from the patient's family for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Bahaa RAZEM: Study concept, data collection, writing the paper and making the revision of the manuscript following the reviewer's instructions.

Yassine DEMMAI: Study concept, reviewing and validating the manuscript's credibility.

Faiçal SLIMANI: reviewing and validating the manuscript's credibility.

Registration of research studies

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