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Rhino-Orbital Mucormycosis Two Case Reports

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Résumé:
La mucormycose est une infection fongique opportuniste causée par des moisissures de l'ordre des Mucorales. La localisation rhino-orbito-cérébrale est la plus fréquente. C'est une maladie destructrice, nécrosante et potentiellement mortelle. Le traitement implique un débridement chirurgical agressif associé à des médicaments antifongiques. L'évolution est rapidement fatale en cas de retard de diagnostic et/ou de traitement. Cette infection touche généralement les patients immunodéprimés et diabétiques, mais des cas de mucormycose chez les immunocompétents sont de plus en plus rapportés. La mucormycose chronique est extrêmement rare et touche aussi bien les patients immunodéprimés que immunocompétents, son évolution clinique est non spécifique et son traitement n'est pas standardisé. Nous rapportons deux cas d'une atteinte rhino-orbitaire chez deux patients de statut immunitaire different, le premier de 59 ans qui présentait un œdème périorbitaire droit associé à une baisse de l'acuité visuelle et une notion d'obstruction nasale et de céphalée d'apparition progressive quatre mois avant son admission. Son état a évolué avec une nécrose rapidement étendue. Elle a subi une résection chirurgicale approfondie mais a rapidement succombé à une défaillance multiviscérale. Le 2éme patient est âgé de 65ans, qui présentait aussi une baisse de l'acuité visuelle de l'oeil droit avec un œdème palpébral inflammatoire important et des zones de nécrose avec exophtalmie stade 3 et une ouverture du globe était impossible, un bilan a été demandé objectivant une hémopathie maligne de découverte fortuite. Le diagnostic de mucormycose a été confirmé sur la pièce d'excision. Le but de cet article est d'attirer l'attention sur la mucormycose chronique chez les immunocompétents et de souligner l'importance d'un diagnostic précoce et d'une prise en charge adéquate de cette infection mortelle.

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I. 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 gastro intestinal 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.

II. Case Reports

Our work is about two cases, our first patient is 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. Probabilistic 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 an 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.

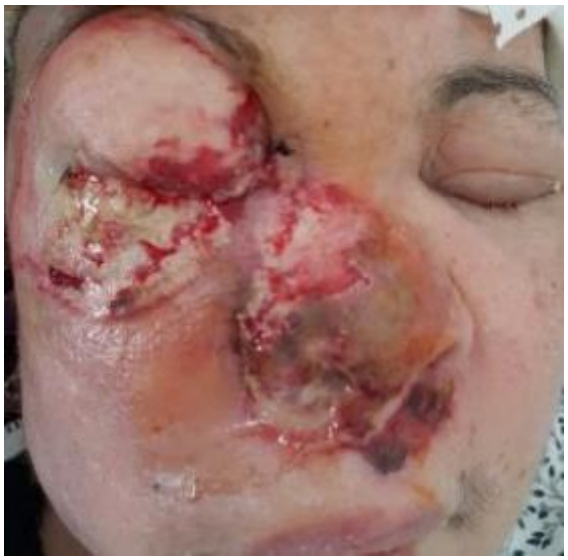

Fig. 1. Facial swelling with palpebral-nasal and labial skin necrosis.

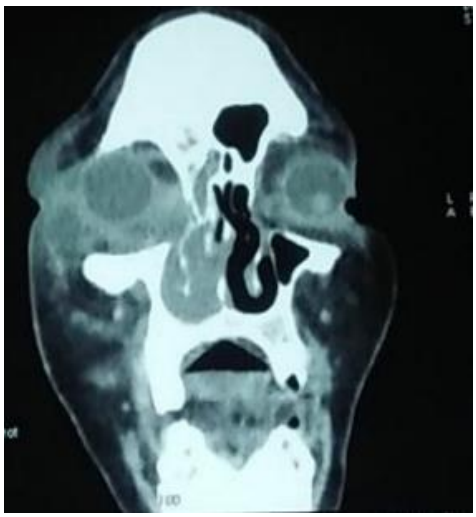

Fig. 2. Facial CT scan: infiltration of the orbital cavity, the nasal cavity and the right ethmoidal sinus.

For the second, it is a 65-year-old man, with no particular pathological history, suffering from a flu syndrome that has been ongoing for 45 days, associated with a red and painful right eye for which he consulted.

On examination, we noted significant inflammatory eyelid edema of the right eye with areas of necrosis with stage 3 exophthalmos, opening of the globe was impossible and the rest was not visible. When opening the mouth, we note the presence of oral candidiasis with areas of necrosis extended to the palate, the inner side of the cheeks and the cavum. An orbital facial CT scan is requested showing uncollected preseptal cellulitis, a biopsy of the lesions performed showing large non-septate filaments of irregular diameters with angular branches. on the right suggestive of mucormycosis. A biological assessment was requested which revealed a hight leukocytosis with blasts.

The patient is referred to hematology for adequate management with antifungal treatment started with voriconazole 300 mg x2 per day (IV) with daily excision of necrosis, the evolution is marked by the hemodynamic worsening of the patient who was transferred in intensive care then died of septic shock of fungal origin.

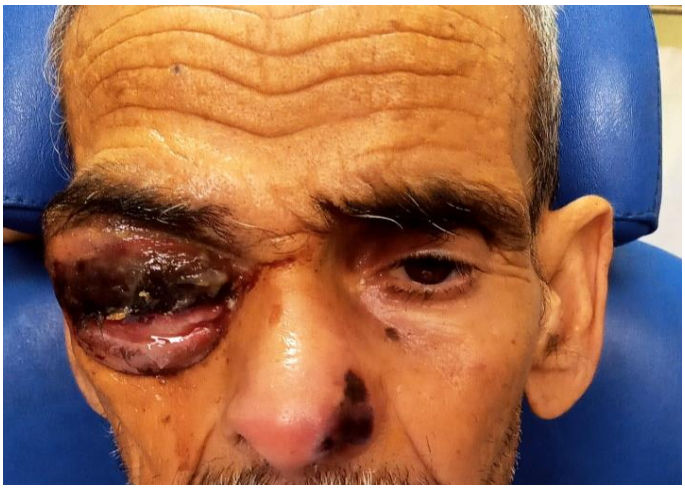

Fig. 3: Eyelid edema with diffuse necrotic plaque, associated with necrosis of the contralateral ala of the nose

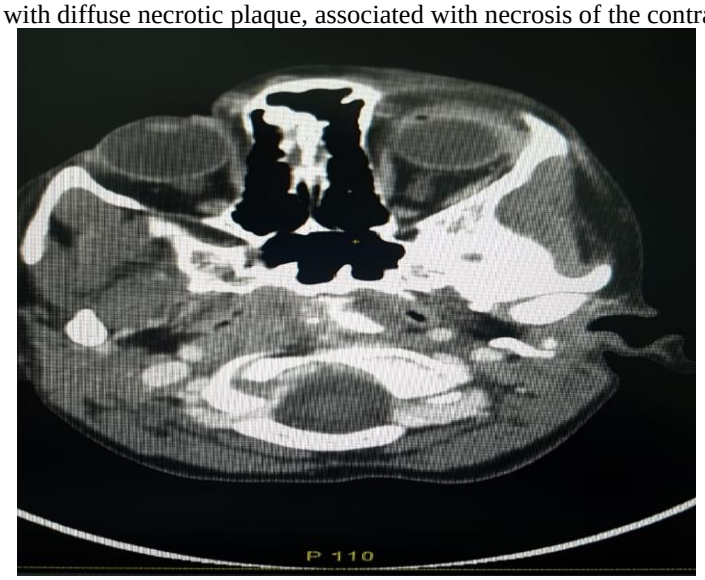

Fig. 4: Non-collected preseptal cellulite objectified on orbital facial CT

III. Discussion

The cases previously reported represent forms of mucormycosis where our first patient presented no risk factors and the second revealed a malignant hematological disease. 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 immuno competent 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 potentialfor rapid spread of mucormycosis, Amphotericin B should be initiated immediately once the disease is suspected, exceptthatthere are no standardized guidelines for the duration oftreatment[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 Amphoericin 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.

IV. 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.