Orbital Complications of Fungal Pan-Sinusitis in Uncontrolled Diabetes

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ABSTRACT

The purpose of our clinical case presentation is to emphasize the role of ear, nose and throat specialist in early recognition and urgent treatment of mucormycosis, which is a rare infection caused by fungus belonging to the order Mucorales. They are known opportunistic organisms, which potentially invade and infect a host with depressed immunity. In our paper we present a case of an uncontrolled diabetic male with orbital complications caused by a fungal pan-sinusitis. The typical presentation of rhino-orbital fungal infection is that of anterior orbital inflammation, severe visual loss, external ophthalmoplegia and fever. Our diagnostic was based on an otolaryngological, ophthalmological, imagistic but especially biopsy exam, which is the only one that can make the certain diagnostic in this case. We followed the standard treatment for these situations. Early recognition and treatment with urgent surgical debridement and systemic antifungal therapy is the key to the management of rhino-orbital mucormycosis and is necessary to limit the spread of infection, which can lead to high morbidity and mortality. Therefore, health practitioners should be familiar with the signs and symptoms of the disease.

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INTRODUCTION

Rhino-orbital mucormycosis is a rare, opportunistic disease which affects, at first the paranasal sinuses and nasal cavity, being induced by the saprophytic fungi and it can quickly evolve towards death. The early signs and symptoms of rhino-orbital mucormycosis are non-specific that’s why early diagnose is really difficult. Onset is typically acute. Nasal discharge of blood may be present, and nasal examination may reveal a black, crusty material. The most frequent signs of this disease are represented by: headache, facial pain, lethargy, decrease visual acuity affecting one eye, unilateral exophthalmia, modified color vision and visual field. The infection spreads, invading the nerves, blood vessels, cartilages, bones, per neural space, inducing thrombosis of the Cavernous Sinus, the Carotid Artery and Jugular Vein and also nervous dysfunction.

Mucormycosis must be suspected in all diabetic patients, particularly those in ketoacidosis.
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 sis, and any debilitated or immunocompromised individual with multiple cranial nerve palsies with or without proptosis. It requires immediate hospitalization because this is a rapidly progressive and possibly life-threatening disease, and the treatment is complex both medical and surgical and must be conducted in a multidisciplinary team.

**OBJECTIVES**

The purpose of our paper is to present a clinical case of a rare, potentially lethal disease, of rhino-orbital mucormycosis and our clinical approach regarding diagnostic and treatment. We also wish to emphasize the important role of the clinician in early detection of mucormycosis, by recognizing the signs and symptoms of this disease and also in adapting the treatment of this serious infection. The immune status of the patient is also an important key, knowing the fact that mucormycosis is an opportunistic disease, that affects the immunosuppressed patients, in our case uncontrolled diabetes mellitus with ketoacidosis. The idea of boarder pathology is stressed, by showing in our presentation the involvement of more than one of medical specialties: ears, nose and throat specialist, ophthalmologist, neurologist, internist, infection disease specialist and radiologist.

**CASE REPORT**

We present the case of a 53-years old male with uncontrolled diabetes mellitus that presented himself at the Ears, Nose and Throat ward in Coltea Hospital with a huge palpebral edema that induced a complete ptosis with the impossibility to open the right eye, with protrusion of the right eyeball. The patient accused violent retroorbital pain, depressed general status and he had fever 39.2 degrees Celsius. From the history of the disease it is important to note the fact that for the last two months, the patients felt a slight pain on the right half of his face, with "on and off" discrete palpebral edema, headache with right hemicranias, and deterioration of visual acuity of his right eye. His diabetes was diagnosed four years ago, is non insulin dependent, and within the last few months was not well controlled, with glycemia values between 290 and 350 mg/dl (according to the patient's own measurements at home).

The ophthalmological exam, revealed: visual acuity of the right eye was 0.5; intraocular pressure of his right eye 16 mm Hg; right upper eyelid edema with complete ptosis and the impossibility to maintain the eye open, axial exophthalmia, painful, non-reductable, limitation of the ocular movement (complete paresis of the right eye and non-reactive mydriasis). The anterior pole of the right eye examined with a biomicroscope showed moderate conjunctival edema (chemosis) and normal in rest and eye fundus examined with a Volk lens revealed a normal optical disc, and a back-ground diabetic retinopathy. The left eye has 20/20 visual acuity with a back-ground diabetic retinopathy.

The results of the CT scan was: "expansive tissue that invades the right nasal wing, filling the nasolabial and nasopalpebral angles, invading the maxillary sinus and the anterior floor of the nasal fossa and expanding along the outer wall of the maxillary into the pterygopalatine fossa. We can note a cranial extension and also of the right hemiethmoid bone and intraorbital, retroocular, filling the orbital cone and enlarging the optic meatus. A same density tissue invasion is seen on the right side of the sphenoidal and frontal sinuses; inducing changes in bones structure and periostosis of the right wall of the maxillary sinus, internal wall of the orbit and the adjacent ethmoid" (Figure 1).

The cranio-cerebral MRI exam revealed: third degree right exophthalmia; important inflammatory changes of the right maxillary and right ethmoid sinuses and partial involvement of the right sphenoidal and frontal sinuses. The above mentioned areas appear with hyper intensity on T2 and they are heterogeneous captant, the inflamed mucous membrane alternate with small liquid collections non-captant. On the right orbit we notice a typical aspect of right orbital abscess with myositis changes on the medial and lateral rectus muscles. The ophthalmic vein has no vascular anomalies; the right cavernous sinus appeared enlarged but with no signs of thrombophlebities.

The histopathological exams were performed after prelevation of the biological material from the sinus's mucous membrane during the surgery, thus certifying the diagnostic. The results of the histopathological exams revealed enlarged necrosis with rare PMN and mitotic colonies, abundant inflammatory infiltrate with many PMN and fibrosis, pavimentous metaplasia of the glandular epithelium and periodic
acid-Schiff (PAS) staining identify fungus from tissue debris, that's typical for mucormycosis.

After corroborating the results from the ear, nose and throat, ophthalmological, imagistic but especially biopsy exam, we finally confirmed the following diagnostic: Right rhinosinusal mucormycosis, right orbital fungal abscess, and uncontrolled non-insulin diabetes mellitus.

Because this is a serious, potentially lethal disease, we had to act quickly, and surgery was imminent and emergency. First, we tried to equilibrate the general status of the patient, especially the glycemic values, for this we cooperated with the diabetologist and the intensive care specialist. The surgery consisted of large debridement of the necrotic tissues (the areas with osteolysis and chondrolysis) until we reached plain healthy tissue. We administrated systemic (endovenous) Amfothericine B Dezoxycolat using progressive doses, starting with 0.6 mg/kg/zi and finally we adjusted to 1.2 mg/kg/zi, for 6 weeks, followed by Posaconazole for 30 days. During the first treatment, with Amfothericine we monitored the hepatic and renal functions and also the potassium levels (1). In recent studies, the efficiency of Posaconazole, FDA approved drug is revealed, as profilactic treatment for Aspergillus and Candida infections in immunosuppressed patients, dose 400 mg twice a day, success rate being 50-70% in curing mucormycosis (2-5).

The purpose of the surgical treatment was to slow the evolution of the disease and to perform the biopsy for certain diagnostic. We performed extensive debridement in order to remove the necrotic tissue. Even if the orbital involvement is a bad sign, the exenteration of the orbit should not be performed from the very beginning (6,7).

The evolution of the patient was good, no relapses were reported and the diabetes was controlled. After 15 days the palpebral edema was in remission, the patient could move the right eye and open the eyelid, though the ocular motility was reduced, the visual acuity of the right eye was 0.7. After two years from this episode with a normal glycemic values during this period, the patient presented for a check-up in our clinic. The ophthalmological exam revealed, good visual acuity of the right eye 0.9, normal ocular movements, the equality of both palpebral fissures.

DISCUSSIONS

The fungi from the Mucor family can be found ubicuitary in nature, in the soil, vegetation etc. They follow a rapid multiplication, through spores that can have an aerogenic transmission. A normal person comes in contact with this fungi almost daily, though mucormycosis is a relatively rare disease, as it affects the immunosuppressed people: uncontrolled diabetes mellitus, lymphoma, leukemia, neoplasia, AIDS, neutropenic patients, those with different transplants and also after steroidic treatment (8). The great majority of the infections appear by direct inhalation or ingestion of the spores, or if a traumatic part of the skin makes contact with an infected area. The infection spreads really quickly via blood vessels producing tissues necrosis. Very often both treatment and diagnosis are late or inadequate. The mortality of mucormycosis is really high, the highest being in patients with transplants 80% (9), and lowest in uncontrolled diabetic patients with ketoacidosis 20-40% (10), as the diabetes can be equilibrated relatively quickly. This last situation being the case of our patient, for whom the correction of all metabolic and immune deficits was of great importance along with Amfothericine and surgery, the treatment and follow-up of the patient needed involvement from a multidisciplinary team of specialists.
CONCLUSIONS

In 70% of cases mucormycosis appears in diabetes mellitus patients, the percent is increasing proportional with the use of chemotherapy or with the association of other immunosuppressive conditions. The symptoms are nonspecific and the diagnostic is not that prompt, sometimes other associated pathology can misled the clinician. If left untreated, mucormycosis has a fulminate evolution towards death. The survival rate of immunosuppressed patients with rhino sinus mucormycosis without cerebral involvement is between 50-80% and only 10% if the infection spreads into the brain (11). In uncontrolled diabetes mellitus patients with ketoacidosis that are diagnosed with rhino-orbital mucormycosis we should suspect a cerebral spread of the fungi if after 24 hours since the beginning of treatment, the patient is still confused.

Regarding our patient’s situation I would like to stress the idea, that even we encountred a difficult situation, immunocompromised patient with mucormycosis, the early and correct combined (medical and surgical) treatment saved patient’s life and determined a good recovery of the visual system.

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REFERENCES