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Rhinocerebral Mucormycosis

Rinoserebral Mukormikozis

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Rinoserebral mukormikozis nadir görülen, ilerleyici ve ölümcül olabilen oportunistik fungal bir infeksiyondur. Yüksek mortalite ve morbiditeye sahiptir. Erken tanı, medikal ve cerrahi tedavi prognozun düzelmesi için önemlidir. Biz, predispozan faktörü aplastik anemi ve akut myeloblastik lösemi olan iki olgu sunduk. Mukormikozis, olgularımızın etkilenen gözünde oküler hareketlerde bozulmaya, propitozise, kemozise ve hiperemiye neden olmuştur. Ayrıca ilk olgumuzda mukormikozis nedeni ile trigeminal sinirin birinci ve ikinci dalıda etkilenmiştir. Her iki olgumuzda da mukormikozis infeksiyonu histopatolojik olarak doğrulanmıştır. İlk olgumuzun manyetik rezonans görüntülemesinde sol taraftaki maksiller, sfenoid ve etmoid sinuste yumuşak doku artışı ve etkilenen bölge çevresindeki dura ve ekstraokuler kaslarda kontrast tutulumu mevcuttur. İkinci olgunun orbitofrontal bilgisayarlı tomografisinde sol frontal, maksiller, etmoid ve sfenoid sinuste radio-opasite ve sol etmoid sinusun kemik duvarında delikli destruksiyon vardır. Her iki olgumuzda antifungal tedavi verilmesine rağmen ölmüştür. Mukormikozisin başarılı tedavisi için erken tanı, altta yatan predispozan risk faktörün düzelebilir olması, cerrahi debridman ve erken antifugal tedavi gerekmektedir.

Anahtar Sözcükler: Rinoserebral mukormikozis, oküler palsi, presdispozan faktör, erken tanı, antifungal tedavi

Rhinocerebral mucormycosis is a rare, progressive and fatal opportunistic fungal infection. It has high morbidity and mortality. Early diagnosis, medical and surgical management is necessary for improving prognosis. We reported two cases. Aplastic anemia and acute myeloblastic leukemia were predisposing factors in our cases. Mucormycosis caused ocular movement disorders, amaurosis, proptosis, chemosis and hyperemia on the affected eye in our cases. Furthermore, the first case had trigeminal palsy involving first and second divisions of the nerve due to mucormycosis. We confirmed mucormycosis infection with histopathological examination in both cases. In our first case, magnetic resonans imaging revealed increase of soft tissues in the maxillary, sphenoid and ethmoid sinuses and contrast-enhancement of dura around the affected area and of extraocular muscles on the left side. Orbitoparanasal computed tomography of the second case revealed radiopacity involving the left frontal, maxiller, ethmoid, sphenoid sinuses and spotty destruction of bony walls of the left ethmoid sinus. Although we gave prompt antifungal therapy in both cases, they died. Successful treatment of mucormycosis requires early diagnosis, reversal of underlying predisposing risk factors, surgical debridement and prompt antifungal therapy.

Key Words: Rhinocerebral mucormycosis, ocular palsy, predisposing factor, early diagnosis, antifungal therapy

Rhinocerebral mucormycosis is a rare, progressive and fatal opportunistic fungal infection (1). Mucormycosis was first described by Paultauf in 1885 (1, 2). The majority of cases have diagnosed in patients with immunologic and metabolic disorders such as diabetes mellitus, lymphoma, leukemia, myelodysplastic syndromes, prolonged corticosteroid therapies, acute renal failure, and severe burns (1-11).

Rhinocerebral mucormycosis has high morbidity and mortality. Early diagnosis, medical and surgical management is necessary for improving prognosis (7, 8).

Case Report 1

A 50-year-old male who had aplastic anemia was referred to our neurology clinic because of amaurosis and pain on the right eye. He had proptosis, chemosis and mydriatic pupil with no response to light and

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complete visual loss with no light perception on the right eye. Horizontal movements were impaired on the right eye. He had hypoesthesia within the first and second division of the trigeminal nerve. Magnetic resonance imaging (MRI) of brain and orbita revealed soft tissue in the maxillary, sphenoid and ethmoid sinuses and contrast-enhancement of dura and extraocular muscles (Figure 1). We thought he had mucormycosis and started amphotericin B 3mg/kg daily. The biopsy of maxillary sinuses confirmed our diagnosis of mucormycosis. Because of pancytopenia the patient could not be operated and died in one week.

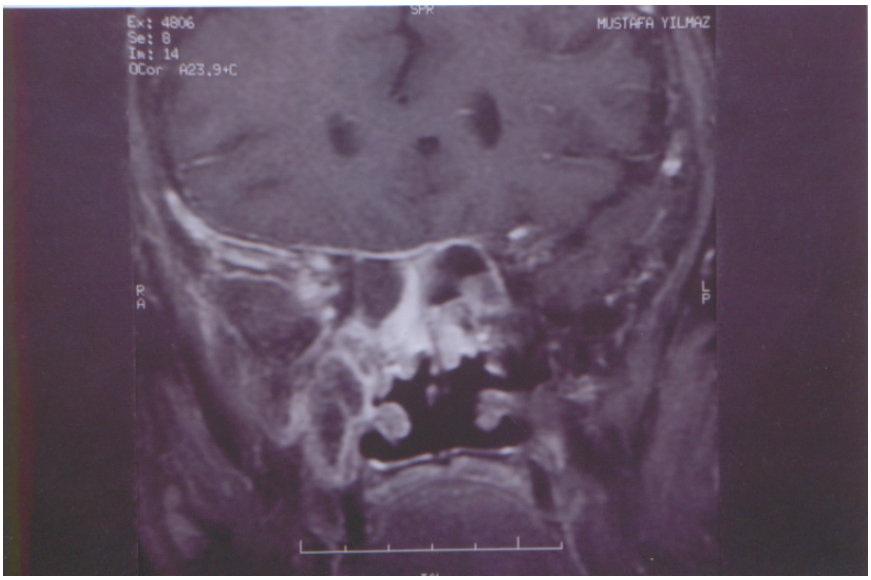

Figure 1: Soft tissue increased of maxillary, sphenoid and ethmoid sinus and contrasten-hancement of dura and extraocular muscles in MRI.

Case Report 2

A 46-year-old female referred to our clinic from haematology with the chief complaint of headache, pain over the left side of his face and left eye, nasal obstruction and left eye swelling. She had acute myeloblastic leukemia. On physical examination, there was edema around the left eye, her left eye was hyperemic and swollen. There was no ocular palsy at once. Orbitoparanasal CT scan revealed radiopacity involving the left frontal, maxiller, ethmoid, sphenoid sinuses, spotty destruction of bony walls of the ethmoid sinus (Figure 2). Surgical debridement of paranasal inflamed tissue was done; mucormycosis was diagnosed by histopathological examination and amphotericin B treatment was started. Despite of early diagnosis and treatment, the patient got worse. Total ophtalmoplegia developed on the left eye and the patient died one week later.

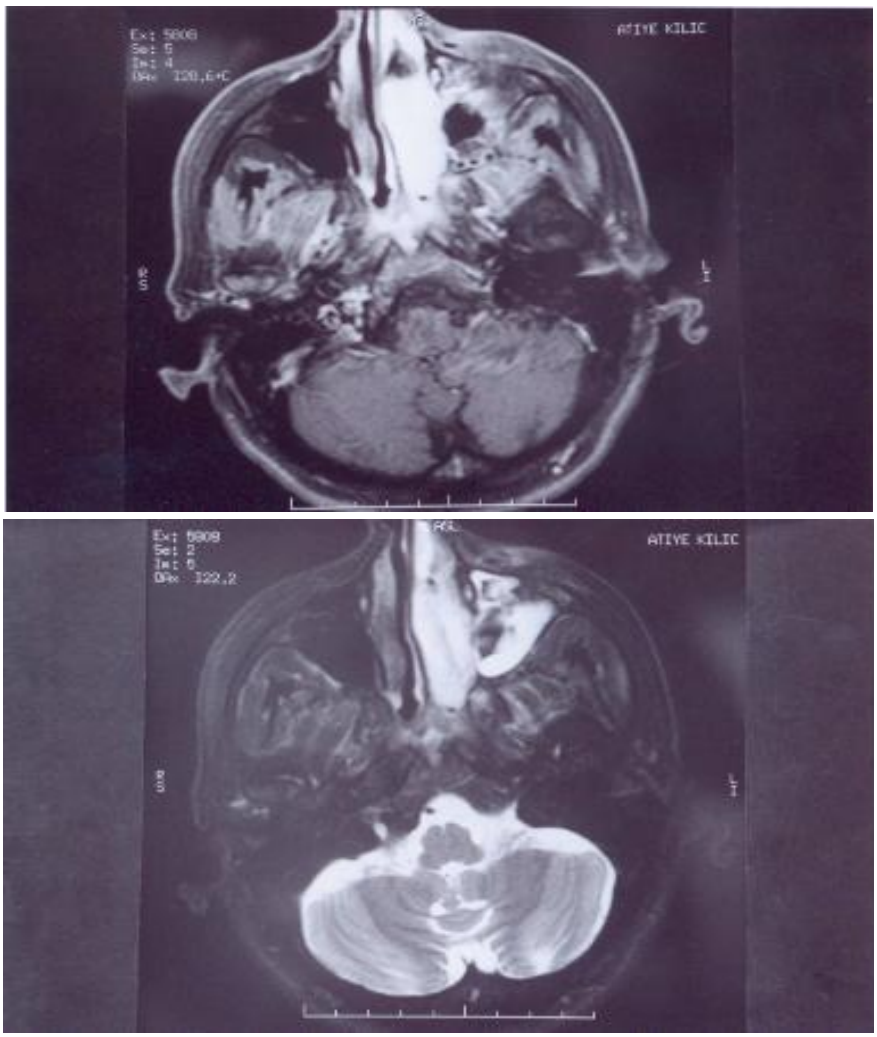

Figure 2: Orbitoparanasal CT scan revealed radiopacity involving left frontal, maxiller, ethmoid, sphenoid sinuses, spotty destruction of bony walls of ethmoid sinus.

Discussion

Zygomycetes (mucormycosis) is a malignant opportunistic fungus that cause central nervous system infection in the setting of obvious

immune dysfunction or anatomic abnormalities (5, 12, 13). It occurs as a rare complication in patients with diabetic acidosis, in drug addiction, and in patients with leukemia and lymphoma, particularly those treated with corticosteroids and cytotoxic agents, severe burns, cirrhosis, severe dehydration, asthma, nephrotic syndrome, necrotizing gingivitis (1-13). Some of the cases have no predisposing factors (14, 15). Aplastic anemia and acute myeloblastic leukemia were the predisposing factors in our cases.

Zygomycetes can invade cerebral blood vessels, causing an arteritis that can trombose or rarely, rupture. Zygomycetes infection often extends from an initial sinus or nasal infection and spreads along infected vessels to the retro-orbital tissues and then to the adjacent brain, producing a localized mass with adjacent cerebral infarctions due to thrombosis from hyphal invasion of arteriole vessel walls (5, 12, 13). Numerous hyphae are present within trombi and vessel wall, often invading the surrounding parenchyma. Sometimes, it may cause hemorrhagic infarction (5).

Zygomycetes location involves the cavernous sinus with the presentation of facial pain, cranial nerve palsies (cranial nerves III, IV, V and VI), proptosis and edema of the lids and retina (1, 3-15). Palatal ulcer was reported with rhinocerebral mucormycosis in literature (9). Mucormycosis caused ocular movement palsies, amaurosis, proptosis, chemosis and hyperemia on the affected eye in our cases. Furthermore, the first case had trigeminal palsy involving first and second divisions of the nerve due to mucormicosis.

Fungi are difficult to isolate or identify from cerebrospinal fuid (CSF). Culturing the fungus from CSF occurs in less than 5% for Zygomycetes (12, 13). Histopathological examination shows aseptate and irregular fungal hyphae branching with neutrophilic infiltrate invading the smaller blood vessels and causing necrosis (1, 6, 9-11, 14). We confirmed mucormycosis infection with histopathological examination in both cases.

Radiological studies such as CT or MRI show contrast enhancing masses within sinuses, cavernous sinus thrombosis, intracranial masses, brain abscesses and ischaemic infarcts in cases with mucormycosis (1, 6, 9-11, 14). The our first case, MRI revealed increase of soft tissues in the maxillary, sphenoid and ethmoid sinuses and contrast-enhancement of dura around the affected area and of extraocular muscles on the left side. Orbitoparanasal CT of the second case revealed radiopacity involving the left frontal, maxillary, ethmoidal, and sphenoidal sinuses and spotty destruction of bony walls of the ethmoid sinus.

The cerebral form of mucormycosis is usually fatal in short order. Rapid correction of hyperglycemia and acidosis, and treatment with amphotericine B have resulted in recovery in some patients (5). Amphotericine B is a antifungal that has been proven to be efficacious. The daily dosage is slowly increased from 0.5 to 1 mg/kg/day or higher for treatment of Zygomycetes infections. Amphotericine B is frequently nephrotoxic. Patients should be followed with frequent serum creatinine and urine analysis; red or white blood cells can be seen in urine analysis. Liposomal amphotericine B should be preferred because of least nephrotoxic effect (13). Surgical management is necessary for improving prognosis (7, 8). Although we started antifungal therapy as early as possible in both cases, they died.

Successful treatment of mucormycosis requires early diagnosis, reversal of underlying predisposing risk factors, surgical debridement and prompt antifungal therapy. However, mucormycosis is frequently a fatal disease despite early diagnosis and prompt treatment such as in our cases. ophtalmology, otorhinolaryngology, neurology, hematology and internal medicine specialists should be aware of the disease and begin amphotericine B treatment as soon as possible when the first manifestations of the disease occur. The treatment should be begun even before histopathological confirmation of the disease for not being late for effective treatment.

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