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Case Report

Right Maxillary Sinus Mucormycosis with Intracranial Extension in a 14-Year-Oldmale

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Abstract

Rhino-cerebral fungal infection is rare in immunocompetent patients without a history of underlying predisposing risk factors. Signs of this infection can mimic other diseases such as vasculitis or intracranial tumors. We present a 14-year-old male patient with diplopia, headache, and strabismus after blunt facial trauma. The first clinical impression was that of vasculitis such as Wegener's granulomatosis,but histopathological examination of abiopsied specimen confirmed the presence of rhino-cerebral mucormycosis.

INTRODUCTION

Mucormycosis is a fungal infection caused by fungi in the Mucoralesorder [1]. It is one of the most rapidly progressing and lethal human fungal infections that typically begins in the nose and paranasal sinuses. Mucormycosis frequently involves the sinuses and brain as the primary sites of infection [2]. It can also present in pulmonary, gastrointestinal, cutaneous, or disseminated forms. This fungus invades the arteries, forms thrombi within blood vessels, thus reducing blood supply, and causes necrosis of tissues [3].

In most cases the patient is immunocompromised, although rare cases have occurred in which the subject was not identified. Such rare cases are typically caused by a traumatic inoculation of fungal spores [3,4]. Predisposing factors for mucormycosis include Acquired Immune Deficiency Syndrome (AIDS), diabetes mellitus, malignancies (e.g. lymphomas), renal failure, organ transplantation, long term corticosteroid and immunosuppressive therapy, cirrhosis, energy malnutrition,and deferoxamine therapy [5]. In the present study, we describe the clinical and imaging-based findings of an immunocompetent patient with invasive rhino-cerebral mucormycosis after blunt facial trauma without bones fracture or skin injury. To our knowledge,this is the first case report of rhino-cerebral mucormycosis in an immunocompetent young adult published in Iran to date. Our case is also particularly interesting given the unknown route of infection and pattern of chronic progression.

CASE PRESENTATION

A 14-year-old male was referred to Otorhinolaryngology-Head and Neck Surgery (ENT) ward for evaluation of numbness in the right maxillary region since two months prior. After obtaining his medical history, he was found to have blunt trauma to his anterior region of right ear and facial area without any facial or skull bones fractures following a bicycle accident two months prior.He explained that in the two weeks prior, he suffered from pain at the traumatic site that was aggravated by chewing food and extended to the right temporal fossa. On physical examination he had pain in the anterior of his right ear with right side strabismus and nystagmus. The past medical history was other wise unremarkable. The patient had no history of fever and purulent nasal discharge was taken. A conventional radiograph (paranasal sinus view) was normal. A Computed Tomography (CT) scan of the paranasal sinuses showed mucosal thickening and soft tissue density in the right maxillary sinus with lateral wall destruction (Figure 1).

Magnetic Resonance Imaging (MRI) of the brain was performed and the findings were interpreted as negative except for mucosal thickening and retained secretions in the right paranasal sinuses due to sinusitis.

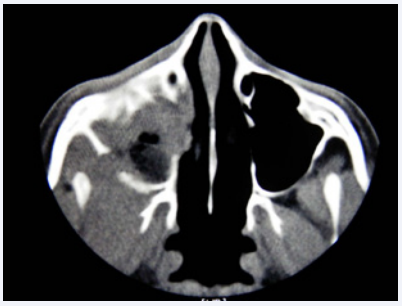

Figure 1 Computed Tomography (CT) scan of the paranasal sinuses shows mucosal thickening and soft tissue density in right maxillary sinus with lateral wall destruction.

After three months, the patient was again evaluated because of aggravation of symptoms. Repeat MRI scans revealed mucosal thickening and destruction of the right maxillary sinus wall involving the right middle cranial fossa dura matter and the right cavernous sinus (Figure 2). Further, a signal abnormality in the right temporal lobe white matter was observed (Figure 3). From these findings, a fungal infection or invasive condition (e.g., lymphoma) was suspected and a biopsy advised. Biochemical assays for C_3, C4, CH50, C-ANCA, and P-ANCA were within normal ranges. Under local anesthesia, a biopsy of the sinunasal mucosa was obtained via a rigid nasal endoscopy, and the specimen was sent to the pathology ward.

Histopathological examination showed fibro vascular connective tissue lined by pseudostratified columnar epithelial cells, stroma severely infiltrated by inflammatory cells mostly lymphoplasma cells, eosinophils, and neutrophils. Numerous multinucleated giant cells were admixed between inflammatory cells. These cells contained fungal hyphae which were non-septatewith right-angle branching, and had a positive reaction by Periodic-Acid-Schiff (PAS) staining (Figure 4). Based on histopathological findings, Zygomycosis was reported. The patient received Amphotericin B (25 mg daily, iv injection). One month later, the patient's symptoms improved. A follow-up brain MRI was performed one month after initiation of treatment and was normal.

DISCUSSION

Mucormycosis is a rare infection caused by organisms that belong to the Mucoromycotinafungi in the Mucorales order. Various conditions can predispose patients for mucormycosis such as an immunocompromised state, diabetes mellitus, malignancies such ashematologic neoplasms, neutropenia, broad-spectrum antibiotic use, direct or indirect trauma, and chemotherapy with corticosteroid therapy [6,7]. Mucormycosisis found in soil and decaying organic matter. The route of infection is via inhalation of conidia, ingestion, or traumatic inoculation

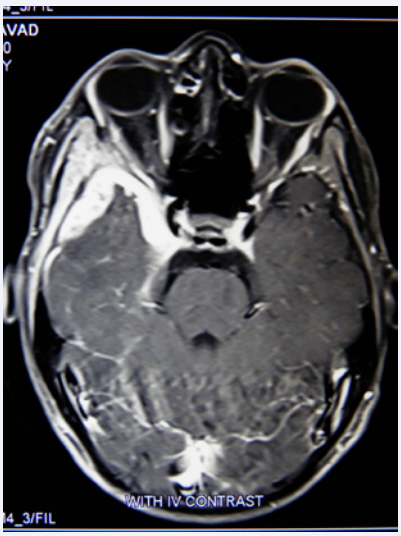

Figure 2 MRI reveals mucosal thickening and destruction of right maxillary sinus wall, with involvement of dura mater in the right middle cranial fossa and right cavernous sinus.

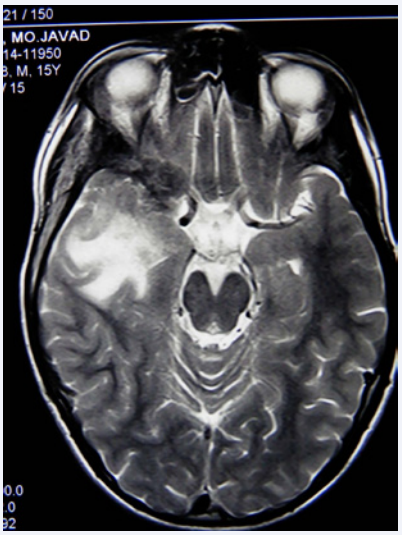

Figure 3 MRI reveals signal abnormality in right temporal lobe white matter.

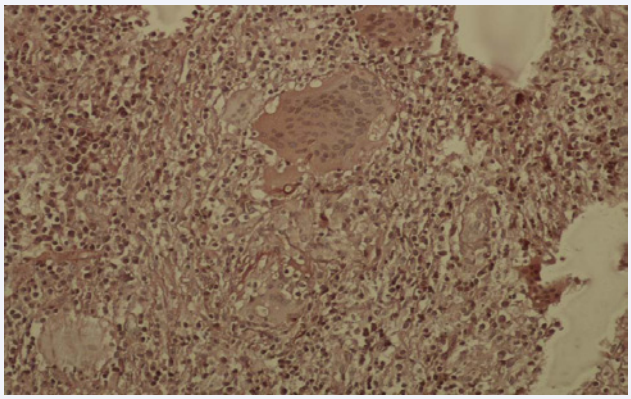

Figure 4 Multinucleated giant cell engulf the fungi hyphae (PAS staining, objective X40).

[2-8]. Our patient had a history of blunt trauma to the anterior region of right ear and facial area without any bone fracture or skin ulcer.Patients affected by mucomycosismay exhibit various clinical symptoms based on the target organ infected [1-6]. Mucormycosis commonly affects gastrointestinal, pulmonary, cutaneous/subcutaneous, rhino-cerebral, and maxillary sinuses tissues.Patients with affected sinuses experience symptoms such as headaches, fever, and sinus pain [6]. Co-infection of rhino-cerebral mucormycosismay occur with other agents presenting with atypical symptoms such as purulent nasal discharge,indicative of bacterial infection [8]. Our patient experienced headaches, strabismus, and nystagmus without rhino-sinusitis symptoms. Although clinical recommendations for identifying fungal infections are often useful,histopathological examination of the affected tissues are necessary for a definitive diagnosis from culture.Mbarek and colleagues reported a series of patients with mucormycosiswho all presented with diabetes mellitus with various symptoms, but interestingly, our patient did not have a history of diabetes mellitus [2]. A case study on mucormycosis in immunocompetent patients by Mignogna and colleagues revealed that the rhino-cerebral infection can occur

via inhalation of sporangiospores from the atmosphere or contamination of skin by physical injuries. Importantly, our patient did not have ulcers on his skin following blunt trauma [9]. Also, in that study, the mean age of infected persons was 54.5 ± 8.6 years, but our patient was 14 years old.Histopathological examination of affected tissues revealed an infiltration of mixed inflammatory cells includingeosinophils, neutrophils, lymphoplasma cells and a foreign body reactioncomposed of multinucleated giant cells ingestingfungal hyphae. PAS staining was an accurate method for the detection of fungi. For a differential diagnosis of acute and chronic inflammation with giant cell infiltration, Wegener's granulomatosis, tuberculosis, and fungal infections could be considered. Wegener's granulomatosis is characterized by geographic necrosis with granulomatous inflammation and vasculitis. The C-ANCA assay can also detect Wegener's granulomatosis in up to 80% of patients [10,11]. Tuberculosis can be ruled out by caseating granulomatous inflammation and Ziehl–Neelsen staining, which detects the presence of acid-fast bacilli in a granuloma. Mucormycosis infection is typically detected by MRI scanning. This infection can appear as an isointense lesion in T1-weighted images, but in T2-weighted images, the signal intensity is variable and occasionally exhibits a hyperintensity pattern [11]. Taken together, treatment is based on patient symptoms and predisposing factors, but rapid intervention can effectively decrease the mortality and morbidity rates in patients.

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