Case Report

Wegener`s granulomatosis and mucoromycosis: A case study and review of literature

Sayyed Mojtaba Abtahi, Mohammadreza Omrani[1], Mansoor Karimifar[2], Afrooz Eshaghian[1], Maryam Kachuei[3], Azarmidokht Momeni[4]

Department of Otorhinolaryngology, Kashani Hospital, [1]Department of Otorhinolaryngology, [2]Department of Rheumatology, [4]Department of Pathology, Alzahra hospital, [3]Isfahan Faculty Of Medicine, Isfahan University of Medical Science, Isfahan, Iran

Abstract

Mucormycosis is a fatal invasive infection which mostly involves diabetic or immunosuppressed patients. Early diagnosis, improving immunosuppression, systemic antifungal therapy, and surgical debridement are necessary for successful treatment. In this case study, we represent a known case of Wegener's granulomatosis (WG), with concomitant sinusal mucormycosis mimicking vasculitic disease relapse, which was successfully treated with surgical debridement, amphotericine, and intravenous immunoglobuline.

Key Words: Immunoglobulins, intravenous, mucormycosis, paranasal sinuses, wegener granulomatosis

Address for correspondence:
Dr. Afrooz Eshaghian, Department of Otorhinolaryngology, Al-Zahra hospital, Isfahan University of medical science, Isfahan, Iran.
E-mail: eshaghian@edc.mui.ac.ir
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INTRODUCTION

Wegener`s granulomatosis (WG) is a small-to-medium vessel vasculitis.[1] American College of Rheumatology defined WG by two of this four criteria: nephritic urinary sediment, abnormal chest finding, nasal or oral inflammation, and granulomatous inflammation on biopsy.[2]

WG is treated by combination of corticosteroids and cyclophosphamide or other immunosuppressive agents (like methotroxate and azathioporine).[3]

Mucoromycosis is as a rapidly progressive fatal fungal infection which involves mostly immunocompromised patients. A biopsy of suspected tissue and culture confirmed the diagnosis of invasive fungal rhinosinusitis. Treatment requires reversal of predisposing factors (such as acidosis or immunosuppression), systemic antifungal therapy, and surgical debridement.[4]

Fungal infection mimics signs and symptoms of WG, and could be neglected for an early biopsy. In accordance with literature, this is the first published case of sinus mucoromycosis in a WG patient that was diagnosed and treated successfully.

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CASE REPORT

The patient was a 39-year-old man who was referred to our hospital because of headache, cough, and hemoptysis. The patient is a known case of WG for 2.5 years. His other complains were fever and chills. Dark bloody nasal discharge was also mentioned.

Headache was localized on frontal and maxillary sinuses. It was permanent and not related to light and

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noise. In the frontal region, a pulsatile fashion was described. No exacerbating factor was mentioned. We found no nausea or vomiting history during those days.

Firstly, there were bloody streams in sputum; however it changed to apparent hemoptysis in a few days. It occurred two times and was estimated about 200 cc each time.

He had no previous history of smoking, addiction, any specific allergy, hypertention, diabetes mellitus, or hyperlipidemia. He afflicted a chronic anemia and admitted for pneumonia last year. Vaccination seems to be complete. No history of tuberculosis or any similar problem in the first-degree relatives had been recorded before.

He received cyclophosphamide (100 mg BID), methyl prednisolone (50 mg QID), ceftriaxon (1 g BID), and azithromycine (250 mg per day). Two units of packed cells were administered after his last episode of hemoptysis.

During his stay, he was conscious and obey, a little pale, and had mild coughing without any toxicity. He complained of blurred vision in eyes, nasal stiffness, rhinorrhagi, hearing loss, hematuria, perspiration, artheralgia in wrist and foot.

On the examination of neck mobile, not firm, elastic lymph nodes was palpable. He had a saddle nose and septal perforation with many dark crusts was obvious in nasal examination.

There was redness in his right eye. Vision and pupil reflex were normal; ophthalmologic examination revealed no ophthamologic involvement.

In chest examination crackles in the upper part of lungs, and generalized expiratory wheezing was heard. Other physical examination revealed no abnormal findings. Spiral-computed tomography (CT) with contrast from the chest revealed multiple cavitary lesions with alveolar infiltration.

In coronal sinus CT scan, a bilateral mucosal thickening in frontal, sphenoidal, ethmoidal, and maxillary sinuses was present. Bony erosion of the medial wall of both maxillary sinuses and nasal septum was seen [Figure 1].

His laboratory data contained WBC: 12000 (NEUT: 79%); hemoglobine: 9.1; urine sediment: RBC+, WBC+, WBC cast, granuler cast; C-ANCA was positive(1/40).

As the patient had complained of hearing loss, a pure tone audiometry was done that showed left conductive hearing loss (40 db, air bone gap).

A biopsy of the nasal septum with culture of crusts was done by the otolaryngologist. Nasal examination revealed necrotic materials [Figure 2]. The specimen was ready in 1 week; during this time the patient became ill, altered mental status, and drowsy.

Pathology and culture revealed a necrotic material with nonseptated fungal hyphae, consistent with mucormycosis [Figures 3a and b], so amphotricine was administered immediately and an endoscopic sinus surgery (ESS) was done to accomplish the diagnosis and treatment. Massive necrotic fungal debris in his maxillary and ethmoid sinuses was brought out completely. The Eustachian tube was eroded in the left side (that justified his unilateral conductive hearing loss).

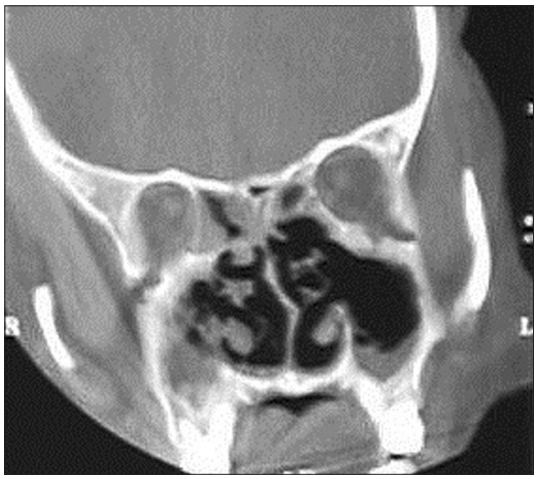

Figure 1: Coronal sinus CT scan of paranasal sinuses (mucosal thickening of ethmoidal sinuses and both maxillary sinuses and destruction of the medial wall of maxillary sinus and nasal septum)

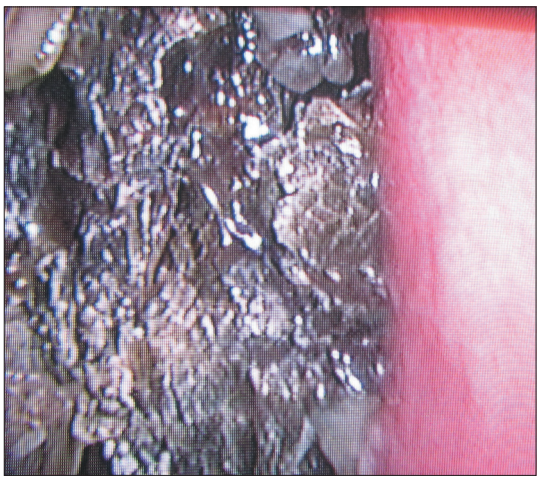

Figure 2: Nasal Endoscopic view: Nasal cavity contained necrotic materials and crusts

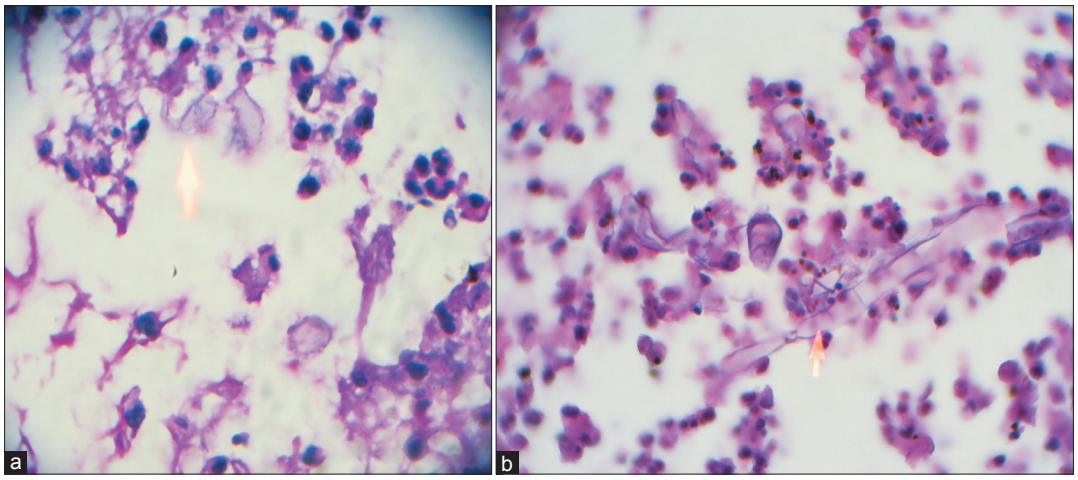

Figure 3: (a) PAS staining, (b) H&E staining (40 HPF) Sections show necrotic material administered with neutrophils and nuclear debris, with some nonseptated fungal hyphae

Immediately after clinical and pathologic establishment of diagnosis of mucormycosis, prednisolone was tapered and administration of cyclophosphamide was stopped. On the other hand, intravenous immune globulin (IVIG) was started (2.5 g for 5 days). Besides, he was also taken vancomycine (500 mg BID), amphotricine (30 mg per day), tab coterimoxazole, tab acid folic, and atrovent spray.

His cough and headache became better. The patient`s mental status improved, and he did not remind any visits or surgery in 2 weeks before. He experienced a second sinus examination by endoscopic sinus surgery. He had complete remission of his fungal infection. Six-week administration of amphotericine was accomplished. At the time of discharge he complained of mild bilateral blurred vision and left conductive hearing loss.

DISCUSSION

This article presents a known case of WG that was superimposed by sinus Mucoromycosis. This is the first reported case of sinus mucoromycosis in a WG patient that was treated successfully. Two previous articles had reported pulmonary mucoromycosis in WG[5] and necrotizing vasculitis.[6]

WG is an ANCA-associated vasculitis with wide variety of symptoms in multiple organs. Nasal symptoms include nasal crusting, ulcer, sinusitis, purulent or bloody rhinorrhea, and saddle nose deformity. Eye involvement can lead to visual loss, retrobulbar orbital masses, proptosis, diplopia, conjunctivitis, keratitis, and uveitis. Hearing loss, oral ulcers, subglotic stenosis, cough, wheezing, stridor, kidney involvement, joint complaints are explained. Skin, nervous system, and heart are also affected, occasionally.[7]

The signs of WG such as septal perforation and bloody discharge of nose and blurred vision are also found in mucoromycosis which makes the diagnosis complex.

Wegener relapse, bacterial sinusitis, nonmucor fungal sinusitis, cavernous vein thrombosis (CVT), tuberculosis, and diffuse pneumonia should be considered as differential diagnosis.

Mucoromycosis is a serious invasive and frequently fatal infection in immunosuppressive patients. The sinus, lugs, brain, and gastrointestinal system are mostly affected. It has been established that before compromising immunosuppression, FESS and aggressive debridement of sinus have not enough efficacy, and the patient`s survival reflects the ability to overcome immunosuppression.[4]

IVIG is an unproven alternative for ANCA-associated vasculitis that is unresponsive to standard therapy.[8] We tapered immunosuppressive agents and change our treatment protocol (IVIG instead of cyclophosphamide); so we were able to help our patient to overcome the mortal mucoromycosis successfully.

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