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CASE REPORT

Misdiagnosis of Otitis Media in a Patient with Wegener's Granulomatosis

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Wegener's granulomatosis is an idiopathic systemic disease characterized by necrotizing vasculitis with the formation of granulomatous lesions that involves the upper respiratory tract, the lungs and the kidneys. Limited forms of Wegener's granulamatosis confined to only head and neck are not rare where as it is unusual that the otologic symptoms are the primary manifestations of the disease. These patients can also be misdiagnosed as otitis media which can cause the deterioration of the disease. We presented here a patient admitted with bilateral otitis media and facial palsy as the first manifestation of Wegener's granulomatosis and was treated with antibiotics for nearly three months before the diagnosis. We conclude that early diagnosis of Wegener's granulomatosis requires a suspicion of the disease in the patients with otologic symptoms and/or facial paralysis if there is otitis media refractory to treatment.

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Wegener's granulomatosis (WG) is an idiopathic systemic disease characterized by necrotizing vasculitis with the formation of granulomatous lesions. The vasculitis affects small and medium-sized vessels. The etiology of this disease is still unclear but appears to be autoimmune. Classically, WG involves the upper respiratory tract, the lungs and the kidneys. Although the limited forms of WG confined to only head and neck are not rare, it is unusual that the otologic symptoms are the primary manifestations of the disease [1, 2]. Paralysis of facial nerve that occurred during the course of the WG is also very rare. These patients can be misdiagnosed as otitis media which can cause the deterioration of the disease. Therefore, the aim of this case report was to document a patient presenting with bilateral otitis media as the first manifestation of WG and no other systemic signs and who suffered from facial palsy during the course of the disease. The patient was treated with antibiotics for nearly three months before the diagnosis of WG.

Case Report

A 31-year-old man admitted to different outpatient clinics with a history of right otalgia, mild hearing loss and discharge of the right ear lasting for nearly three months and he had prescribed different kinds of antibiotics. However, there was no improvement under the medication with local and systemic antibiotics and besides the same symptoms started in the left ear of the patient two weeks ago. So, he presented to our outpatient clinic with bilateral otalgia and discharge, severe hearing loss and subfebrile fever.

On examination, bilateral tympanic membranes were hyperemic, thickened and bulging with a pulsatile mucopurulent discharge. The Rinne test (512 Hz tuning fork) was bilateral negative and Weber test was lateralized to the left ear. Fiberoptic nasopharyngoscopy and laryngoscopy showed no abnormalities in the rhinopharynx and larynx. The audiogram revealed bilateral mixed hearing loss 83dB for the left ear and 81dB for the right ear (Figure 1).

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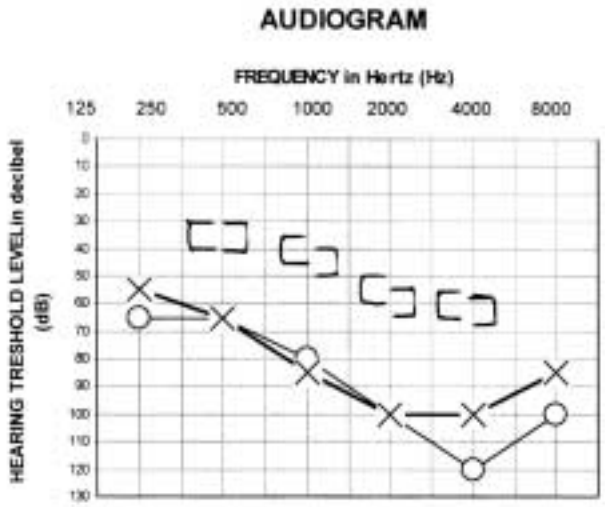

Figure 1. Audiogram shows bilateral severe mixed hearing loss before treatment.

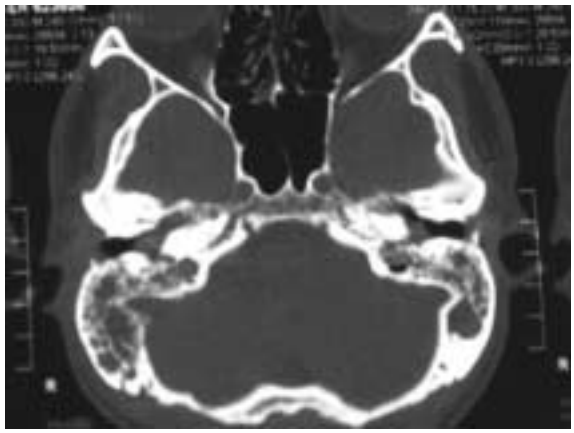

Figure 2. Axial CT scanning of the temporal bone shows bilateral inflammatory material occupying the mastoid air cells and the middle ear.

The leukocyte count was 14,800/mm3 with a 60% neutrophil fraction and the other routine hematological and biochemical tests of the blood and urine were in the normal ranges. Human immunodeficiency virus and hepatitis B virus screening were negative. The x-ray of the paranasal sinuses and the chest had no pathological findings. Computed tomography scanning of the temporal bones showed bilateral inflammatory material occupying the mastoid air cells and the middle ear (Figure 2). Antibiotic treatment (third generation cephalosporin and metronidazole) was given intravenously. Myringotomy with insertion of ventilation tubes performed and yielded purulant fluid bilaterally. There were no acido-resistant bacilli (ARB) and any other bacterial growth on culture of the myringotomy material.

He failed to respond to the medical therapy lasting for ten days and underwent right cortical mastoidectomy for diagnosis. Granulomatous and polypoid changes of the mastoid cells were observed and all granulation tissue and polyps were removed. The operation materials were sent for pathological and microbiological examinations. Facial nerve paralysis was observed on the opposite ear two days after the operation, and facial nerve function degenerated to a House-Brackmann Grade IV. The bacteriological and mycobacteriological examinations of the materials were negative. The histopathologic examination showed inflammatory granulomatosis suggesting the possibility of WG. Systemic involvement of the disease was evaluated after he was referred to a rheumatologist. The cytoplasmic pattern of anti-neutrophil cytoplasmic antibody (cANCA) was positive. High resolution computed tomography of the lungs revealed multiple nodules and patchy alveolar infiltration with a central necrosis in both lower lobes indicating the involvement of lungs. The renal ultrasonography and urinary tests showed no renal involvement.

Treatment with methylprednisolone and methotrexate was started as recommended by the rheumatologist. There was dramatically improvement in his symptoms and audiogram findings after ten days treatment. The ear examination of the patient showed bilateral small inferior-posterior retraction pocket and a small inferior-anterior perforation at the tympanostomy site in the right ear. Besides, paralysis of the facial nerve was improved and achieved a House-Breckmann Grade I and the audiogram showed bilateral minimal hearing loss, 38dB for the left ear and 40dB for the right ear at three months after hospital discharge (Figure 3).

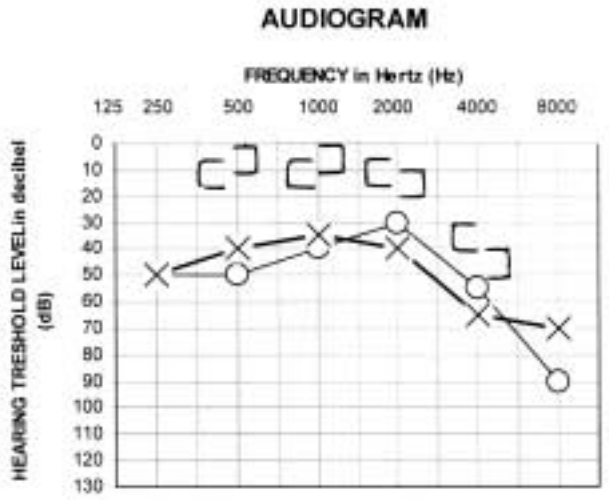

Figure 3. Audiogram shows improvement of hearing at three months after hospital discharge.

Discussion

WG is a disease that was described by Friedrich Wegener in 1936 and characterized by necrotizing vasculitis with the formation of graulomatous lesions. The vasculitis affects small to medium-sized vessels. Classically, it involves the upper respiratory tract, the lungs and the kidneys [2]. Involvement of at least one of three systems and a biopsy confirming necrotizing granulomatous inflammation, usually with vasculitis, or the presence of cANCA confirms WG [3]. The limited forms of WG confined to only head and neck are not rare and it is unusual that the otologic symptoms are the primary manifestations of the disease[2]. Therefore, these patients can be misdiagnosed as otitis media which can cause the deterioration of the disease because of the delay in the therapy. Paralysis of facial nerve that occurred during the course of the WG is also very rare. We presented here a patient admitted with bilateral otitis media and paralysis of the facial nerve as the first manifestations of WG and no other systemic signs. Before the diagnosis and treatment of WG, he was given different kinds of antibiotics by the misdiagnosis of otitis media for nearly three months.

The otologic manifestations of WG are common and range from 19% to 45%. Mc Caafrey et al [4] classified otologic involvement into three major patterns; serous otitis media (90%), sensorineural hearing loss (43%) and chronic otitis (24%). Paralysis of the facial nerve should also be added to the otological manifestations. The granulomatous process in the temporal bone and/or rhinopharynx causes the otologic manifestations in the middle ear. However, the patients with WG do not usually refer with ear symptoms on admission and otologic symptoms as the primary manifestations of the disease was reported as 10% [5]. Thus, these patients can be diagnosed as otitis media by mistake and this may cause a delay in the management of the disease. Our present case was admitted with bilateral otitis media as the first manifestation of WG and no other systemic signs. Therefore, he was given invalid antibiotic treatment for nearly three months before the diagnosis of WG.

As mentioned above, another important otologic manifestation of WG is facial palsy (5%) [4]. It is much rarer than the rest of the otological findings alone or in combination with hearing loss. It is caused by either compression in the temporal bone or vasculitis of the vasa nervosum [5, 6]. Illum and Thorling [7] demonstrated multiple neuropathies of cranial nerves VI, VII, IX, XII in two patients with otologic involvement of WG. Both patients had large cranial base lesions with destruction of the petrous portion of the temporal bone. In our patient.

The mechanism of sensorineural hearing loss in WG is unclear. It was suggested that vasculitic or granulmatous involvement of the auditory nerve is responsible for the hearing loss [4, 8, 9]. The sensorineural hearing loss is the result of the primary involvement of the ear by the disease whereas conductive hearing loss is caused by the serous otitis media. The audiogram of our patient revealed bilateral mixed hearing loss which was assessed as the result of ear involvement and sensorineural hearing loss was decreased by the three months treatment.

Clinical suspicion of WG should prompt determination of cANCA levels which is 99% specific

for the disease. However, it should be kept in mind that the sensitivity of the test depends on the activity of the disease and in case of loco-regional involvement the sensitivity of the method is 60%, whereas 93% in the generalized form [10]. On admission we did not measure cANCA level in our patient. After WG was diagnosed by the histopathological examination of the operation specimen, cANCA was measured and found to be positive.

Treatment of WG includes cyclophosphamide, methotrexate and systemic steroids. In patients with limited forms of WG, methotrexate is a choice for remission-induction treatment. A recent large study supports the use of methotrexate in limited forms of the disease showing comparable remission-induction rates to those treated with cyclophosphamide [11]. As cyclophosphamide can be associated with dose dependent toxicities including cystitis, opportunistic infections, malignancies, infertility and cytopenias and the patient did not have renal involvement; methotrexate was the choice of therapy. Regardless of whether cyclophoshamide or methotrexate is used for remission-induction therapy, glucocorticoids are given concurrently and are an essential part of treatment. Surgical management of WG related to middle ear disease is not suggested as patients may become worse after myringotomy and mastoidectomy [12]. Facial nerve decompression may also aggravate the problem by the damage of the nerve. However, surgical procedures can be performed for the diagnosis of the disease like in our patient who underwent mastoidectomy.

If untreated, the disease usually runs a rapidly fatal course and 82% of patients die within a year. Overall survival rates of WG have improved over the last decades, reversing the poor and often fatal prognosis of the disease, since the widespread institution of early therapy. Remission rates of 70-85% have been achieved, depending on the extension of disease, particularly a major renal involvement [5,6].

In conclusion, we presented a case with bilateral otitis media and facial palsy as the first manifestations of WG who had been given different kinds of antibiotics due to mistakenly diagnosed otitis media. Early diagnosis of WG requires a suspicion of the disease in the patients with otologic symptoms and/or facial paralysis if there is otitis media refractory to treatment [13, 14]. The deterioration of the clinical symptoms of otitis media should alert the physician, especially in case of severe sensorinoeural hearing loss and facial palsy. In these circumstances, cANCA titers should be measured and histopathological examination should be performed. The management of the disease includes medical treatment with systemic steroids, methotrexate or cyclophosphamide and surgical therapy is not suggested unless necessary for the diagnosis.

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