Int. Adv. Otol. 2012; 8:(1) 143-147

CASE REPORT

Bilateral Chronic Otitis Media and CSF Otorrhea; the only Presenting Feature of Wegener's Granulomatosis

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Wegener's Granulomatosis (WG) is an autoimmune disease. The lungs, kidneys and upper respiratory organs are major sites of organ involvement. Although nose, ear and throat involvement can be observed in most of cases during the natural course, their individual involvement is quite rare. A case with bilateral refractory chronic otitis media and spontaneous CSF fistula that was diagnosed as WG is presented.

Submitted : 16 May 2011 *Accepted : 15 July 2011*

Introduction

Wegener's Granulomatosis (WG) is an immune mediated disease which is characterized by necrotizing vasculitis of small arteries and veins. The specific organization of necrotized vessels ultimately results with necrotizing granuloma formation in the affected organs [1]. The upper respiratory tract involvement is usually presented by nasal congestion, epistaxis, discharge, sinusitis, cough, stridor, otitis media, hearing deficit and oral ulceration[2]. The other organs which are commonly involved are lungs (45%-70% at presentation and 85% throughout the disease course), kidneys (77% of patients), heart, nervous system and muscles and joints[3].

The nose and paranasal sinuses are mainly affected organs concerning the upper respiratory tractus. They are involved in %85 of cases during disease progress and the ear involvement is seen approximately in 35% of all cases with WG4,[5]. The patients are commonly presented with nonspecific symptoms including upper respiratory tract infections, sinusitis, fever, fatique, myalgias in its early stages. These nonspecific findings may delay the early diagnosis of the disease so that the profound organ involvements are seen later in disease progress. The otologic manifestations occasionally may be the only presenting feature in WG. There are only a few cases reported; chronic otitis media, mastoiditis, facial nerve paralysis, sensorineural hearing loss were the only presenting symptoms[6-9].

The certain diagnosis of WG can be made with the presence of circulating pattern antineutrophil cytoplasmic antibodies (C-ANCA) and anti-PR3 antibodies which are specific to WG in the active phase. Also histological examination of biopsy specimens of specific organs involved may be helpful in certain diagnosis making[10].

This case report presents a patient with WG whose initial only presenting feature was bilateral otitis media with effusion which rapidly progressed to refractory otitis media, sensorineural hearing loss and spontaneous CSF fistula.

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Case report

A 50-year-old woman was admitted at the Department of Otolaryngology of Uludag University with the complaints of severe pain, hearing loss and discharge in the right ear. These symptoms had already been persisting for almost one month. She had already failed to respond to miringotomy and antibiotic regimen of Ceftriaxone followed by oral cephalosporin. She was admitted to ENT department with the diagnosis of refractory otitis media.

Purulent discharge form right ear, thickening of tympanic membrane, edema of external acustic canal and severe otalgia were the main findings during admission. Mixed type severe hearing loss was detected at right ear (BC:58dB, AC:100dB) and mild conductive hearing loss was present on the other side (BC:5dB, AC:28dB). Type B recordings were obtained with immitancemetry bilaterally (Fig1). The high resolution CT x-rays of temporal bone showed a loss of aeration and increased density at mastoid cavities and middle ear especially at right side. There was no bone erosions. Middle ear ossicles were intact bilaterally (Fig2). Magnetic resonance images (MRI) demonstrated mastoiditis on right side with normal inner ear structures .

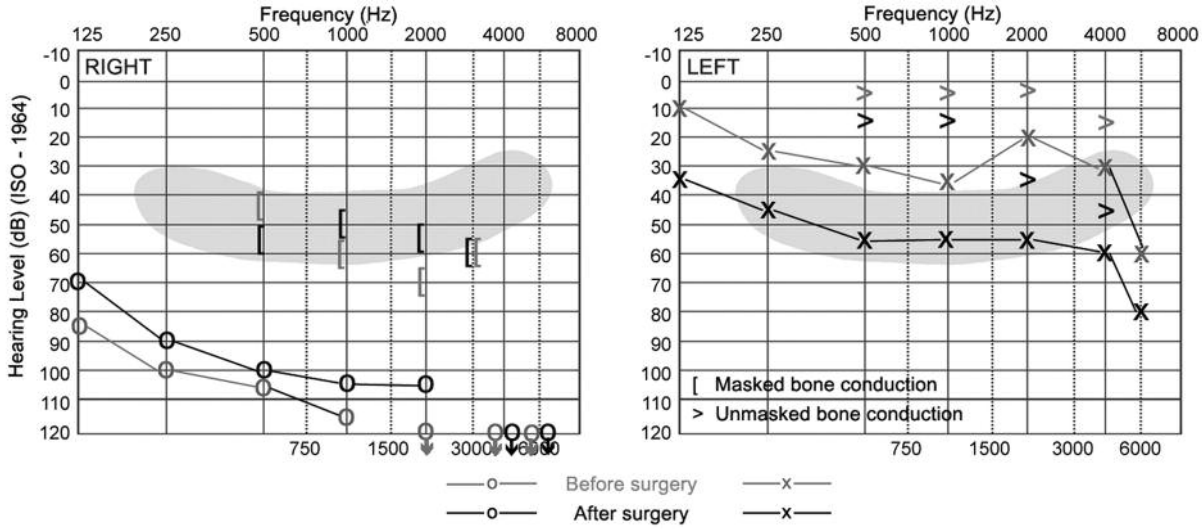

Figure 1. The audiogram showing a severe mixed type hearing loss at right and a mild conductive hearing loss at left ear before surgery; and also a severe mixed type hearing loss at right and a moderate mixed type hearing loss at left ear after surgery.

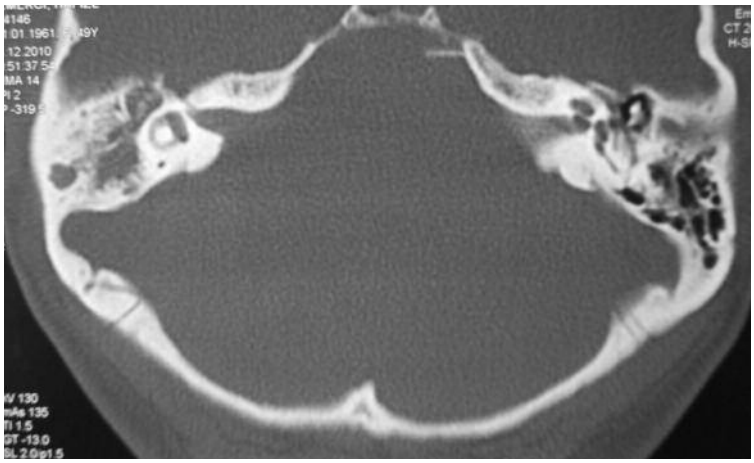

Figure 2. Axial CT scan of the temporal bone shows thickening of the mucosa of the middle ear and mastoid cavity on right side.

The external ear canal was irrigated daily with topical antibiotic applications and a course of ciprofloxasin and tazobactam intravenous administration. Vertigo symptoms arised with the development of polyp tissue soon after.

Mastoidectomy was performed and surgical findings showed thickening of membrane, dense granulation tissues at the attic region and around ossicular chain. The biopsy specimens of middle ear revealed dense mixed type nonspecific inflammation.

The vertigo symptoms persisted with the development of hearing loss and seropurulant drainage at left ear. There was a horizontal left sided nystagmus. A recent audiogram showed severe mixed type hearing loss at right ear (BC:52dB, AC:103dB) and a moderate mixed type hearing loss was detected at left ear (BC:25dB, AC:52dB). Type B recording was obtained again on left ear (Fig1). The videonystagmography (VNG) findings indicated a left sided peripheric vestibular pathology. Subsequently, a grommet type ventilation tube was inserted. The high resolution CT x-rays of temporal bone showed a loss of aeration and increased density at left mastoid cavity and middle ear. The tegmen tympani was thinned but middle ear ossicles were intact at the left side.

During follow-up, a pulsatile spontaneous fluid drainage was observed on left side. The biochemical analysis of this clear fluid indicated a CSF leakage. A regimen of intravenous Vancomycin and Meropenem was immediately administered. Beside a lumbar drain was placed.

Mastoidectomy was performed to left side and a dense granulation tissue which filled the middle ear and mastoid cavity was cleared. There was an active CSF leakage from tegmen tympani. The defect at tegmen timpani was oblitered with adipose tissue and temporal fascia. The biopsy showed fibrin-exudate material which was rich in polymorphic leucocysts.

After surgery, the patient had respiratory difficulties and chest pain that occurs on deep inspiration. The chest X-ray image showed hilar nodular lesions although it was completely normal preoperatively. The patient was positive for c-ANCA and PR3 (1/320 E.P. and 1/1000 respectively). P-ANCA and MPO were negative. And there was a mild elevation of CRP; 1,01 mg/dl. The CT images of thorax revealed consolidated areas at anterior segment of right upper lobe, lateral segment of middle lobe and superior segment of inferior lobe near hilum. And also there were nodules at left lateral segment of inferior lobe with bilateral pleural effusions (Fig3). All these findings were relevant to pulmonary involvement of WG. The biopsy specimens were reevaluated due to suspicion of WG. The definitive diagnosis was WG, because microscopic examination of specimen showed multinuclear giant cells with necrotic areas (Fig4). Any possible risk for renal involvement was eliminated with normal abdominal and urinary system USG and urine tests. Subsequently, the patient was consulted by pulmonologist with the diagnosis of WG.

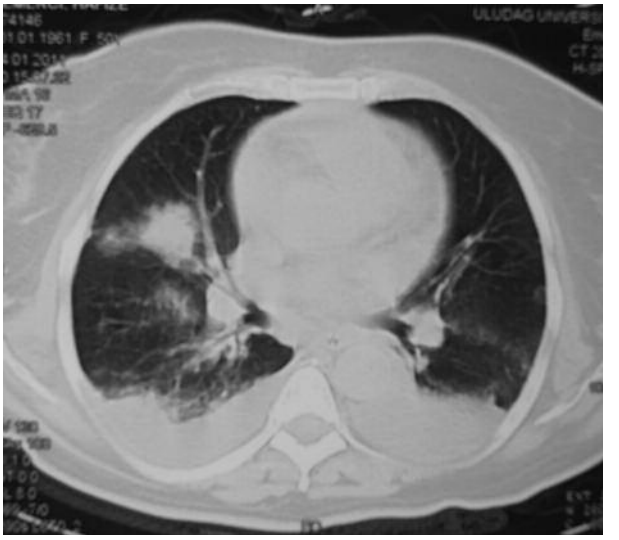

Figure 3. CT scan of the lungs shows multiple consolidated areas and nodules and note also the bilateral pleural effusion.

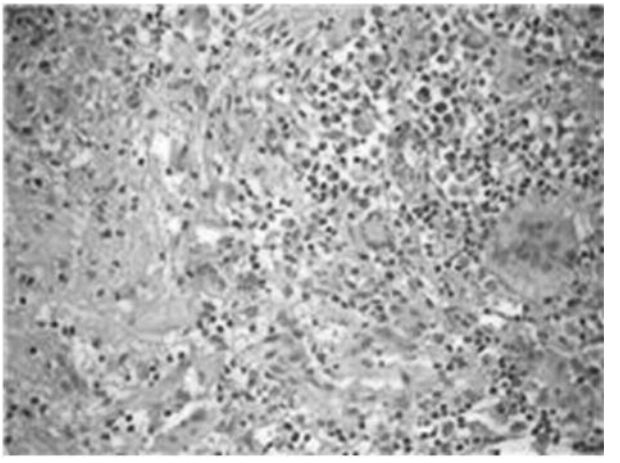

Figure 4. The histological specimen shows multinuclear giant cells and inflammation with neutrophiles, plasma cells, lymphocysts beside necrotic areas consistent with Wegener Granulomatosis.

Later, the patient was referred to rheumatologist and signs and symptoms of the patient were dramatically improved soon after the immunosuppressive treatment started with methylprednisolone, cyclophosphamide and methothrexate. A regimen of Rituximab 1gr per one day was also added to treatment protocol. Unfortunately, a residual bilateral mixed type hearing loss was persisted. There was no discharge in control examinations and the patient is healthy otherwise.

Discussion

Wegener's granulomatosis is an autoimmune disease which mainly affects upper respiratory tract, the lungs and the kidneys[7]. The nose, paranasal sinus and ear involvement can be observed in the majority of cases during the disease course[11]. Furthermore, the otological manifestations were observed in nearly half of patients with WG[12,13]. The most common otological finding was serous otitis media secondary to eustachian tube inflammation[7,14]. Another common finding was chronic otitis media presented by granulomatous lesions at middle ear mucosa and mastoid cavity[6]. And also there are few cases reported in the literature in which cronic otitis media and facial paralysis were the only presenting signs of WG[6,7,9]. Up to our knowledge, this is the first case in English literature which reports a patient with rapidly progressive bilateral chronic otitis media followed by spontaneous CSF fistula.

To make a definitive diagnosis is sometimes difficult especially if the otological manifestations are the only presenting symptoms. In such circumstances, a delay for the initiation of treatment is inevitable. Although there is no consensus regarding the timing of the surgery, it is obvious that early diagnosis and treatment may prevent further complications and improve disease related health of the patient[5,7].

As presented in this case, a delay in diagnosis of WG as well as immediate surgical interventions such as miringotomy, tube insertion or mastoidectomy may worsen the clinical picture. But it is still uncertain that whether this is the natural progress of disease itself or the surgery provoke the disease progress[5-15]. Consequently, the surgery may be delayed until other possible systemic diseases were evaluated.

Since the histopathological examination of granulation tissues at middle ear and mastoid cavity generally reveals nondiagnostic inflammation, the correct histopathological diagnosis can only be achieved if WG is suspected[7,16]. C-ANCA and anti PR3 antibody positivity may be quite helpful especially in the active phase of disease[17]. Furthermore, any change in antibody titers may show the disease activity and reflect the response to treatment[6].

We conclude that the combination of detailed physical examination, positive serological markers and histological confirmation is mandatory to put a definitive diagnosis in patients with WG confined to ear only.

Conclusion

Although otolaryngologists are the medical professionals who are most familiar with otological diseases, they in some cases may not recognize a related systemic disorder. This unfortunately causes a delay in diagnosis and prevents early treatment. In general, a sudden onset of ear drainage with granulative tissues emerging from tympanic cavity, hearing loss, and severe otalgia is not common in the absence of previous history of ear disease. One must consider the possibility of an underlying systemic disease, especially if the case is also refractory to medical treatment and rapid progression with complications is observed. Thus, otolaryngologists should be highly suspicious and aware of the presence of WG beside other autoimmune diseases as causative factors.

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