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Otitis Media as the First Manifestation of Wegener's Granulomatosis

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Abstract: We report a case of a 48 years old woman with bilateral otitis media as the first manifestations of WG who had been given different kinds of antibiotics due to mistakenly diagnosed otitis media. The patient failed to respond to the different outpatient treatments with local and systemic antibiotics. She had redness and buldging in right and left tympanic membrane and the audiogram revealed moderate C.H.L(Conductive Hearing Loss) in right ear and severe mixed hearing loss(MHL) in left ear. The cytoplasmic antibody (cANCA) was positive in high titer. She had two lesions in lung parenchyma in chest –X ray that characteristic necrosis,granulomaand capilaritis were seen in lung tissue biopsies.

Key words: Wegener Granulomatosis, Otitis Media, Cytoplasmic Antibody

INTRODUCTION

Wegenergranulomatosis(WG)is a systemic vasculitis characterized bygranulomatous inflamation involving upper and lower respiratory tract and kidneys. Thevasculitis affect smalland medium sizedvessels. The diagnosis of Wegener granulomatosis isperformed on the basis of the clinical picture, serum cANCA and histologic examination of biopsies. Theantineutrophil cytoplasmic antibodies(cANCA),is reported for the first time in 1985 by Van der Woude,are highly specific for WG,especially in the active phase [1].

Apopulation-based study from Norfolk, England, reported incidences of 8.5 cases per million for WG [2]. Approximately 50% of WG patients have otologic finding, including both sensorineural hearing loss(SNHL) and conductive hearing loss(CHL)[3].The incidence of SNHL in WG varies between 2.8% and 13% [4]. SNHL is more important, because cranial nerve involvement suggests a neurodegenerative process that might warrant aggressive therapy. It is unusual that the otologic symptoms especially SNHL are the primary presentation of the disease.The difficulty of diagnosis often delays the initiation proper treatment and it occasionally progresses to the irreversible phase.Therefore ENT surgeon should be familiar with the disease and to make appropriate referral for treatment.This case report characterized by unusual clinical presentation of WG with the leading sign of hearing loss.

Case Report: A 48 years old woman without previous illness presented with common cold symptom followed by headache and left sided otalgia, tinitus and ear fullness since two month ago.Ten days later ear fullnessand discharge of the right ear was added.She was treated with oral antibiotic for acute otitis media by ENTsurgeon.The patient failed to respond to the different outpatient treatment with local and systemic antibiotic.Due to progressive hearing loss, an audiometry was done. The audiogram revealed moderate C.H.L(Conductive Hearing Loss) in right ear and severe mixed hearing loss(MHL) in left ear.Sinusoidal Computed tomography scaning of the temporal bone and paranasal sinus endoscopy was normal (Figure 1,2). She was admitted to the hospital and intravenous antibiotics were administered.In routine hospitalization process and because of mild hemoptysis, a chest-X ray was done.It showed two lesion inlung parenchyma that one of them sharp,well defined with 4cm in diameter in left hilar area and others in right superior lobe with diameter of 2 cm. For ruleout of metastasis, spinal and pulmonary MRI was done and the patient was counseled with us. Pulmonary MRI (Figure 3) revealed

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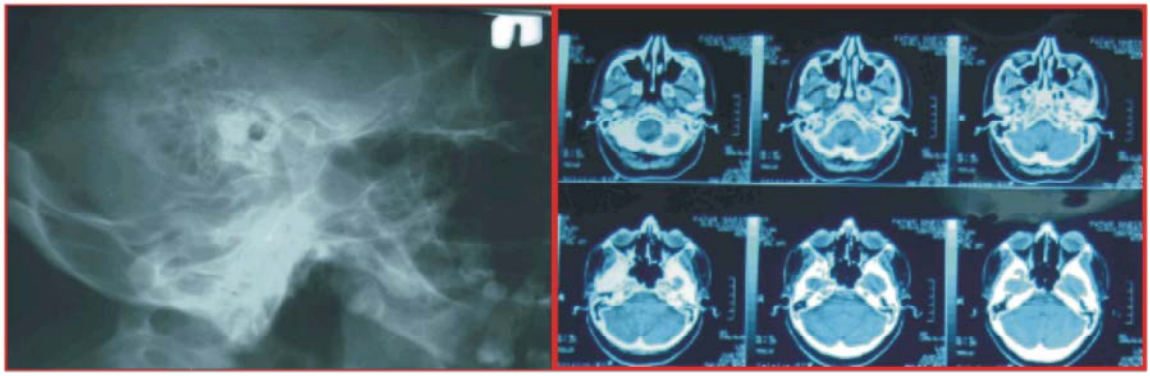

(1) (2)

Fig 1,2: The graphy of temporal and paranasal sinus CT (was normal)

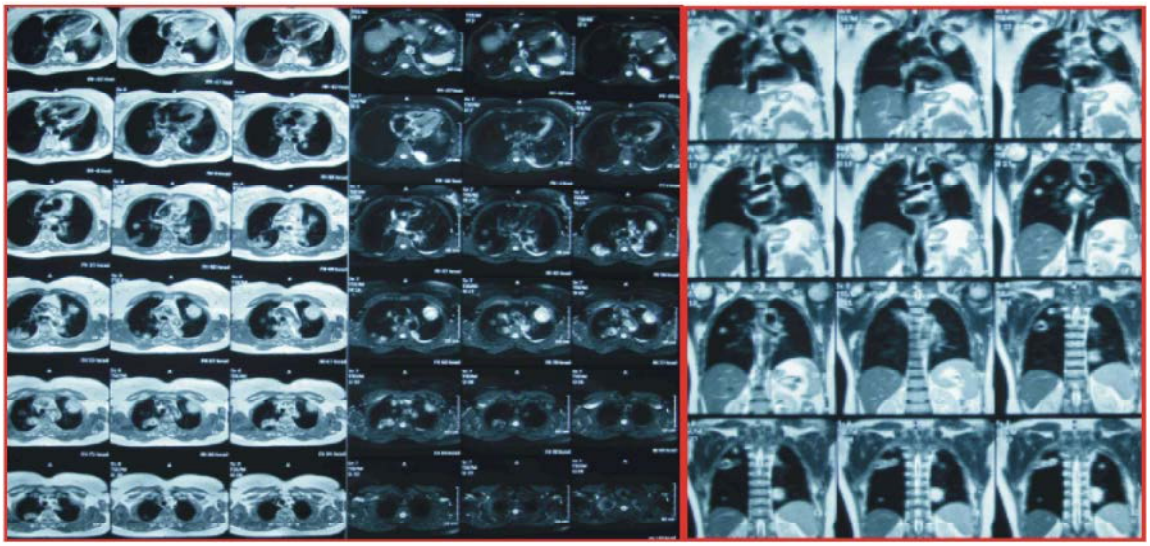

Fig. 3: Pulmonary MRI revealed multiple speculated intrapulmonary mass(41x44mm) and(15x9mm) in left upper lobe, (36x30mm)in left lower lobe and (22x15mm),(16x16mm),(46x16mm),(27x26mm), (29x12mm)in right middle lobe and upper lobes of lungs.

multiple speculated intrapulmonary masses in left upper lobe and left lower lobe and multiple mass in right middle lobe and upper lobes of lungs.Intralesional necrotic foci in some of the above mentioned lesions are also seen and the mediastinal space was normal. Systemic involvement of the disease was evaluated after she was referred to internal ward and on the physical examination,she had redness and buldging in right and left tympanic membrane.

On admission, the leukocytic count was 7800/mm3 with 71% neutrophil fraction and the other routine hematological and biochemical test of the blood was normal.Second audiogram revealed severe bilateral MHL.CT of the lungs revealed multiple cavitary nodules, one mass in left lung,mild bilateral pericardial effusion and right paratraceal lymphadenopathy.

The further clinical course was characterized by sensorymotor peripheral neuropathy in both legs. The cytoplasmic antibody (cANCA) was positive in high titer.For tissue diagnosis an open lung biopsy was done.Characteristicnecrosis,granulomas and capilaritis were seen in all tissue biopsies.Upon this findings prompt treatment with pulse methylprednisolone therapy 1gram for three days and then oral prednisolone(60 mg daily) and cyclophosphamide(100 mg daily)was started. Two months after the treatment, her hearing loss diminished butpersisted.The chest CT scan was completely normal. The motor neuropathy improve gradually but sensory deprivation continues. Elevated ESR and CRP reverted to normal range and the titer of c-ANCA decreased and was negative.One month later cyclophosphamide was discontinued and changed to MTX 15mg weekly and

prednisolone was 10mg daily. Four months after treatment she complained of hip pain.MRI study revealed corticosteroid induced avascular necrosis of the hip. Three years after the therapy,she is alive and doing well and was taken off all medications and remains under control of the outpatient followup.

DISCUSSION

Majority of the patients with WG present with upper airway disease, occurring in more than 90% of cases. Common upper airway presentations include recurrent rhinosinusitis, nasal polyposis, nasal chondritis leading to saddle nose deformity, nasal septal perforation, serous otitis media, hearing impairment and stridor due to subglottic stenosis [5].The limited forms of WG, confined to only head and neck, are not rare and it is not unusual that the otologic symptoms are the primary manifestations of the disease. Therefore, these patients can be misdiagnosed as otitis media which can cause the deterioration of the disease because of the delay in the therapy [6].

The otologic manifestations of WG are common and range from 19% to 45%. McCaafrey [7] classifiedotologic 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.Illum and Thorling[8] demonstrated multiple neuropathies of cranial nerves VI, VII, IX,XII in two patients with otologic involvement of WG. The mechanism of sensorineural hearing loss in WG isunclear. It was suggested that vasculitic or granulmatous involvement of the auditory nerve isresponsible for the hearing loss [7,9,10]. 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. 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[11]. 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[12]. As cyclophosphamide can be associated with dose dependent toxicities including cystitis, opportunistic infections, malignancies, infertility, cytopeniaandthe patient did not have renal involvement; methotrexate was the choice of therapy. Regardless ofwhether cyclophoshamide or methotrexate is used for remission-induction therapy, glucocorticoids are given concurrently and are an essential part of treatment.If untreated, the disease usually runs a rapidly fatal course and 82% of patients die within one 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 [13,14].

In conclusion, 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 [15,16]. 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.The management of the disease includes medical treatment with systemic steroids, methotrexateor cyclophosphamide.

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