Chronic otitis media and facial paralysis as a presenting feature of Wegener's granulomatosis

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ABSTRACT

Upper airway disease, especially nasal and paranasal sinus involvement, is the most common manifestation of Wegener's granulomatosis. Chronic otitis media and facial palsy are rare but well known presenting features of Wegener's granulomatosis. We report a 40-year-old woman who presented with complaints of ear discharge, deep-seated ear pain and loss of hearing in her right ear. Early diagnosis demands heightened suspicion in a patient with otological symptoms and facial paralysis.

Keywords: chronic otitis media, facial nerve palsy, hearing loss, Wegener's granulomatosis

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INTRODUCTION

Wegener's granulomatosis (WG) is a clinicopathological syndrome characterised by extravascular granulomatous inflammation, granulomatous vasculitis of small-sized vessels, necrosis of the upper and lower respiratory tracts, and pauci-immune glomerulonephritis.[1] WG and other antinuclear cytoplasmic antibodies related vasculitis are difficult to diagnose because of low prevalence and variable presentations, signs and symptoms, mimicking common illnesses like infections, malignancies, thromboembolism and connective tissue disorders.[2] WG of the middle ear can be due to primary disease in the middle ear leading to chronic otitis media, or secondary to Eustachian tube involvement causing middle ear effusion.

CASE REPORT

A 40-year-old woman presented with complaints of ear discharge, deep-seated ear pain and loss of hearing in her right ear for the past six months. Generalised weakness, joint pain, fever and nasal stuffiness preceded the ear symptoms. The ear discharge was serous initially; however, for the last one month, there was bloody discharge. She complained of deviation of the angle of her mouth. She had consulted an otolaryngologist and underwent simple mastoidectomy with mastoid exploration 15 days previously, with a presumptive diagnosis of chronic suppurative otitis media with complications. She developed a fever that was intermittent and high grade, with profuse sweating. There was an increase in the bloody discharge from the ear with redness of the eye one week before presenting to our hospital. The patient had no record of previous admission. On examination, her vital signs were stable, ear discharge was bloody, with gaping of the surgical scar, intact graft and granulation tissue on the tympanic membrane on the right side. Nasal examination revealed bilateral large crusts. Episcleritis of both eyes was noted. No arthritis, nasal deformity, rashes, thyroid swelling, joint deformity, hair loss, oral or genital ulcers or photosensitive reactions was seen. Systemic examination was essentially normal except for a lower motor neuron type of facial nerve palsy on the right side.

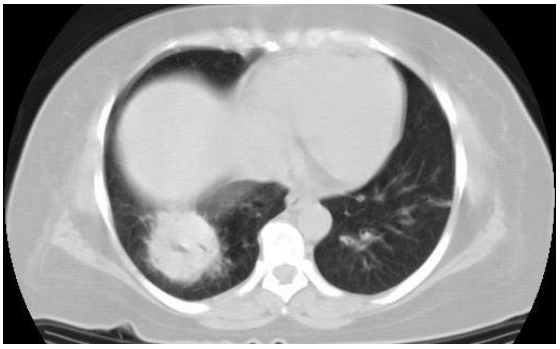

Fig.1 CT image of the chest shows a large nodule with cavitation.

Laboratory investigations revealed a haemoglobin level of 9.7 g/dL, haematocrit 29.8%, leucocyte count 20,800/μL with neutrophilia, platelet count of 431,000/μL, and erythrocyte sedimentation rate of 140 mm/1st hour. Urine examination showed 2+ proteins, 30–35 dysmorphic red blood cells/high-power field. Her microbiological evaluation was negative for malaria, dengue, leptospira and tuberculosis. Blood culture did not grow any bacteria. Human immunodeficiency virus, hepatitis B and C, and ANA serology were negative. Her random blood sugar was 91 mg/dL, blood urea 151 mg/dL, serum creatinine 7.1 mg/dL, Na+ 138 mEq/L, K+ 5.1 mEq/L, serum bilirubin 0.8 mg/dL, aspartate transaminase 28 U/L, alanine transaminase 33 U/L, alkaline phosphatase 183 U/L, and serum amylase 92 U/L. Her bleeding and coagulation profiles were normal. Chest radiograph showed a mass lesion in the right lower zone. Computed tomography of the chest

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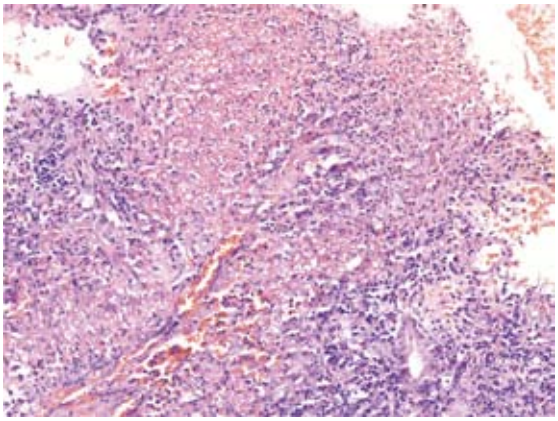

Fig.2 Photomicrograph of the nasal biospy shows granuloma (Haematoxylin & eosin, × 40).

Table I. Otological and nasal manifestations of Wegener's granulomatosis.[11]

Manifestations of Wegener's granulomatosis
Otological
Otitis media with serous effusion
Chronic otitis media
Chronic suppurative ptitis media
Cholesteatoma
Tympanic membrane and temporal bone granuloma
Facial nerve palsy
Meningitis
Sensorineural hearing loss
Vertigo
Nasal
Crusting
Granulation, destruction
Rhinorrhoea, pain, epistaxis, hyposmia
Epiphora
Saddle nose deformity
Perforated nasal septum
Chronic sinusitis

revealed multiple nodular lesions bilaterally, with one large nodule with cavitations in the right basal region (Fig. 1). Pure tone audiometry revealed bilateral mixed hearing loss, which was more severe on the right side. She was positive for rheumatoid factor and C-reactive protein. Serology was strongly positive for circulating antineutrophil cytoplasmic antibodies.

The diagnosis of WG was confirmed when a nasal biopsy revealed features suggestive of granuloma (Fig. 2). The biopsy was negative for acid-fast bacillus. The patient was started on oral cyclophosphamide (2 mg/kg/day) and prednisolone (1 mg/kg/day), along with haemodialysis. However, the response to dialysis was unsatisfactory with increasing creatinine levels. She underwent plasma exchange (40 ml/kg/day). After five consecutive cycles, serum creatinine levels started decreasing. In spite of immunosuppression and steroid therapy, there was minimal improvement in the hearing loss or facial palsy. The patient died after four months due to drug-induced febrile neutropenia and Gram-negative sepsis.

DISCUSSION

Otological involvement may occasionally be the first and only sign of WG.[3] The prevalence of ear involvement varies from 19% to 45% of all cases.[4] Otological involvement may be divided into: (1) serous otitis media, (2) chronic otitis media, (3) sensorineural hearing loss, (4) vertigo, and (5) facial nerve palsy.[5] Serous otitis media and conductive hearing loss are respectively observed in 25% and 14% of patients at presentation and 44% and 42% of patients in the course of treatment.[1] The most common cause is Eustachian tube obstruction from luminal granuloma or nasopharyngeal inflammation and ulceration.[6] Chronic otitis media occurs from the direct involvement of the ear by WG. Less than 10% of the patients with chronic otitis media may present as otorrhoea and conductive deafness due to ossicular damage by destructive granulomata. The inner ear disease causes sensorineural hearing loss (8%), vestibular dysfunction or both, but the mechanism is poorly understood. Proposed mechanisms are the deposition of immune complex in the cochlea, granulomatous compression of the cochlea and vasculitis of the vasa vasorum and cochlear vessels.[4,7] In one of the largest surveys done on 701 patients, 68% had sinus and 62% had lung involvement at diagnosis.[8] Joint pain (51%), sinusitis (48%), nasal discharge (43%), cough (34%) and hearing loss (36%) were the presenting symptoms at diagnosis. Fauci et al reported that 25% of patients presented with serous otitis media and 6% had hearing loss.[9] Takagi et al and D'Cruz et al respectively reported chronic otitis media and conductive deafness as the most frequent finding.[5,10] In 35% of patients, hearing loss was noted as the permanent disease-related morbidity.[3] These were related to recurrent serous or suppurative otitis media, sensorineural impairment or a combination of them.

Systemic diseases affecting the middle ear and temporal bone include granulomatous and infectious diseases, bone disorders, neoplastic diseases, collagen vascular diseases, immune deficiency and AIDS. Differential diagnoses of middle ear WG include tuberculosis, syphilis, Lyme disease, mycotic infections, sarcoidosis, polyarteritis nodosa, Churg-Strauss disease, microscopic polyangitis and malignancy of the temporal bone, epipharynx or parotid gland.[11] Facial nerve paralysis in association with WG is rare, being present in about 5% of patients alone or in combination with hearing loss, and rarely, may be the presenting feature. Bilateral facial palsy has been reported only once. It is secondary to compression of the nerve in the middle ear, especially in the presence of a dehiscent fallopian canal or due to vasculitis of

its microvasculature.[4] Facial nerve decompression is not useful but may aggravate the problem. The nose and paranasal sinus are the most the commonly affected sites, having been noted in 40%–64% of patients. Nasal mucosa and paranasal tissue biopsy provide the most favourable results and are easier to access than other sites (Table I).[11]

Specific causes of adult-onset otitis media include paranasal sinus disease, nasopharyngeal carcinoma, tumours and post-radiation sequelae. WG should be considered as the differential diagnosis in cases of atypical inflammatory states of the ear. The combination of sensorineural hearing loss and facial nerve palsy is extremely rare, and its association with acute or chronic middle ear inflammation should be considered in the early diagnosis of WG. The suspicion of the disease could be raised in situations in which there is granulomatous proliferation in well-pneumatised mastoid cavity with rapid progress or otitis media refractory to treatment. For new-onset otitis media in an adult, consider WG.

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