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CASE STUDY

Otologic Wegener's Granulomatosis☆

Granulomatosis de Wegener del oído

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Introduction

Wegener's granulomatosis (WG) is a systemic vasculitic disease characterized by necrotizing granulomas and vasculitis of small vessels.[1,2] Upper airway tract involvement is frequent over the course of the disease.[1,3] Cytoplasmatic pattern antineutrophil cytoplasmic antibodies (c-ANCA) are highly specific for WG in the active phase.[1,3] Proteinase 3-ANCA (PR3-ANCA) positivity is usually associated to progression from localized to systemic disease.[2]

Case Report

A 51-year-old woman presented with bilateral hearing loss with one month of evolution associated with otalgia and tinnitus. After antibiotic, short-term systemic corticosteroids and topical nasal corticosteroid she remained symptomatic. Eardrums were thickened with effacement of normal morphology. She was admitted in Otorhinolaryngology department for tympanostomy tubes placement. Pure-tone audiogram showed severe/profound progressive bilateral mixed hearing loss with air-bone gap of 50 dB on right ear and 40 dB on the left, with neurosensorial worsening at high frequencies (Fig. 1). Computerized tomography (CT) scan revealed complete bilateral medial ear filling (Fig. 2a).

She developed a right peripheral facial palsy (stage II/VI in the House–Brackman scale). She presented mild leukocytosis (11.3×10^9/L – 88.5% neutrophils) with sedimentation rate of 69 mm/h and C-reactive protein of 5.01 mg/dL. Mastoidectomy of the right ear revealed caseous material in the middle ear and hypertrophy of the mucosa; mastoid was aerated. Biopsies of ear mucosa were inconclusive (Fig. 2b). Bacteriological, mycological and mycobacteriological cultures were negative.

There was a worsening in lung function showing bilateral pleural effusion, with elevation of inflammatory parameters except leukocytosis. Enzyme-linked immunosorbent assay (ELISA) serology testing for c-ANCA and PR3-ANCA was positive. Herpes simplex and Epstain–Barr virus antibody serology were negative for active infection. Mantoux skin test was negative. Biopsy of the nasal mucosa showed no granulomas. The diagnosis of WG was made based on these findings. She started on methylprednisolone 1 g/day therapy. During the hospitalization the patient developed a non-ST-elevation myocardial infarction, from which recovered. She was discharged on oral methylprednisolone and three monthly cycles of cyclophosphamide. Four months later she developed right per

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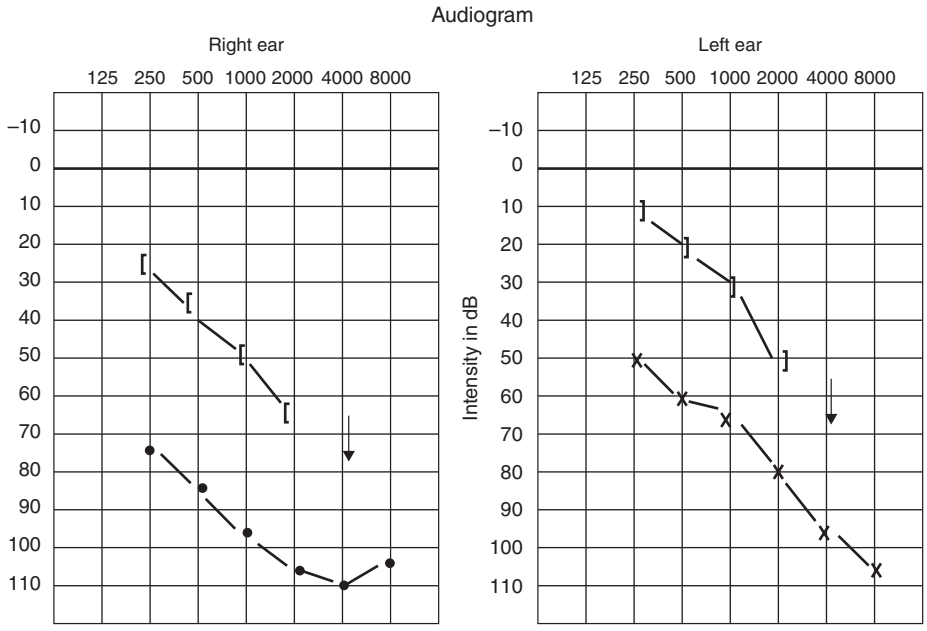

Figure 1 Pure tone audiogram before mastoidectomy.

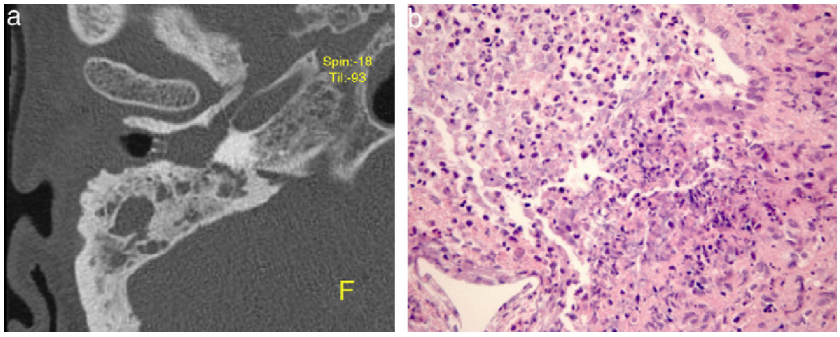

Figure 2 (a) Right ear CT scan. (b) Middle Ear mucosa microscopy – ''*unspecific inflammation*''.

orbicular pain and the CT scan was compatible with WG involvement of the lachrymal gland. She restarted cyclophosphamide and systemic corticosteroids and was discharged with oral prednisolone with stable bilateral profound neurosensorial hearing loss with no other symptoms.

Discussion

WG is a systemic vasculitic disease, although some forms are limited to the respiratory tract.[1,2] If untreated, WG usually is a rapidly progressive disease with 82% mortality within a year.[1,3]

The ear is involved in 19%–61% of all cases but is unusual to be the first and only sign of disease in WG.[1,3] Facial nerve palsy has been reported during the course of the disease but is extremely rare as a presenting feature.[3] Pulmonary involvement is frequent, including pleural effusion.[4] In localized cases, it is frequently difficult to make a definite histologic diagnosis.[1,3,5]

ANCA are detectable in nearly 100% of patients with active generalized disease WG, but only 60% in localized form. High-sensitive PR3-ANCA ELISA yields a sensitivity of 96% and a specificity of 98.5%.[2,5] ESR and c-ANCA levels are useful markers for monitoring disease activity.[3]

A combination of glucocorticoids and cyclophosphamide is the standard treatment for WG. It is important to start the treatment before irreversible changes occur.[4]

This case revealed an aggressive progression, starting with otological manifestations, diagnosed by serological and clinical features.

In conclusion, WG is a challenging diagnosis and should be considered in refractory inflammatory states of the ear.

References

1. Takagi D, Nakamaru Y, Maguchi S, Furuta Y, Fukuda S. Otologic manifestations of Wegener's granulomatosis. Laryngoscope. 2002;112:1684–90.
2. Moosig F, Lamprecht P, Gross WL. Wegener's granulomatosis: the current view. Clinica Rev Allerg Immunol. 2008;35:19–21.

3. Ferri E, Armato E, Capuzzo P, Cavaleri S, Ianniello F. Early diagnosis of Wegener's granulomatosis presenting with bilateral facial paralysis and bilateral serous otitis media. Ausis Nasus Larynx. 2007;34:379–82.
4. Pero MM, Sivasothy P. Vasculitis of the upper and lower airway. Best Pract Res Clin Reumatol. 2009;23:403–17.
5. Alicandri-Ciufelli M, Melteni G, Mascia MT, Genovese E, Pressuti L. ''Hide and seek'' with antineutrophil cytoplasmic antibodies. Am J Otolaryngol. 2010;31:397–8.