CASE STUDY

Severe Vestibulocochlear Involvement in Wegener’s Granulomatosis: A Rare Presentation

Afectación vestibulococlear grave en granulomatosis de Wegener localizada: una presentación atípica

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Clinical Case

We present the clinical case of 61-year-old female, lacking relevant antecedents, whose condition began with tinnitus and hearing loss of the right ear (RE). At 4 months she presented an episode of sudden vertigo, followed by continuous dizziness and imbalance. She received a diagnosis of otitis media with effusion (OME), but did not improve after various treatments. After that, she developed hearing loss of the left ear, with a sensation of fluctuating blockage, tinnitus and right facial paresis. One year later, the hearing loss had progressed toward bilateral cophosis and the imbalance worsened. However, the facial paralysis facial subsided. Otomicroscopy exploration: image compatible with OME. Ear and neurological examination: no spontaneous nystagmus, Halmagyi test with corrective ocular with saccades on both sides, instability in Romberg and Unterberger test, absence of response to caloric stimulation (even with ice water) and in the kinetic vestibular rotating chair test (at both low and intermediate frequencies), the patient presented only minimal response to high frequency stimulation, with a gain of merely 0.12–0.64 Hz of rotation. Imaging tests: computed axial tomography (CAT) scan revealed occupation of the maxillary sinus and of bilateral middle ears (ME). On magnetic resonance imaging (MRI), there was thickening of the left posterior wall of the cavum and signs of bilateral damage, stronger in RE (Fig. 1). Immunological analysis: This confirmed positivity for p-ANCA type antibodies with titers of 1:64. Histological analysis: biopsy of ME, cavum and maxillary sinuses showed granulation tissue, small foci of necrosis, fibrinoid degeneration, inflammatory infiltrate and extravasation of red blood cells (Fig. 2). Treatment was initiated with prednisone at 1 mg/kg/day and

Figure 1 3D T-2 weighted FIESTA MRI sequence of the labyrinth. MIP reconstruction of IE: damage to the basal turn of the cochlea (*) and lateral semicircular canal (#) with absence of normal hyperintensity.


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Figure 2  Histopathology. Biopsy of ME and cavitum: infiltrate of chronic inflammatory cells, small foci of necrosis, fibrinoid degeneration, granulation tissue and extravasation of red blood cells (hematoxylin–eosin staining, 40×).

Azatioprine at 100 mg/day. At 4 months the patient was clinically stable and drug treatment was suspended: The vestibular sequelae were treated with vestibular rehabilitation vestibular and the auditory ones with placement of a cochlear implant (CI) (Medel) in the left ear at 18 months after onset of clinical picture. A year later, she walks without help and presents an audiological threshold in free field of 40 db (mean: 225–4000 Hz) with 48% of two-syllable discrimination.

Discussion

Wegener’s granulomatosis (WG) is a systemic vasculitis necrotizing with granulomatous inflammation of small and medium blood vessels. It normally has a local presentation, in which the clinical findings are limited to the upper airway and lungs, respecting the kidneys. This represents approximately 25% of the cases. In the otorhinolaryngological area, it mainly involves the paranasal sinuses and nose. Otological compromise is described in up to 38% of the cases, with ME involvement being more frequent than that of the inner ear (IE). When the IE is involved, the anterior labyrinthus anterior is usually compromised and, exceptionally, the posterior. Sensorineural hearing loss (SNHL) occurs in 8% of the cases and progression to cophosis is rare. Nevertheless, in a series of 19 patients diagnosed with WG by biopsy, SNHL was observed in 12 of the 13 patients with auditory symptoms. Theories are proposed for this disorder, such as immune complexes, granulomatous compression of the cochlea, vasculitis of the vasa vasorum or nerve damage from polyneuritis. In a recent review on otorhinolaryngological manifestations in 25 patients with WG carried out by Morales-Angulo et al., not a single case of vestibular involvement was described.

In less recent literature we have found only a few cases that compromise both the anterior and posterior labyrinth and none with total cochleovestibular compromise. This patient presents extraordinary characteristics of almost total cochleovestibular involvement and, based on the MRI findings, the lesion would be located at the level of the IE. Cyclophosphamide is the most effective treatment and is generally the initial therapy, associated with corticoids. However, due to its powerful side effects and to the fact that there was not generalized WG in our patient, azathioprine was used. The literature describes one case of WG with bilateral moderate-severe SNHL, treated with unilateral CI, in which a mean auditory threshold of 40 db and two-syllable discrimination of 20% were obtained in free field audiometry. Our patient has achieved a level of discrimination that lets her function acceptably well in daily life. The CI would be a therapeutic possibility for rehabilitating cases of SNHL in WG.

The case presented here is very rare and atypical, not previously described in the literature, of a form of localized WG, with bilateral ear involvement. The patient displayed severe vestibular hypofunction with bilateral cophosis as the main symptoms. Knowing the atypical presentations is important to avoid delay in diagnosis and treatment, and consequently avoid possible sequelae of WG itself.

Conflict of Interests

The authors have no conflicts of interest to declare.

References