Late Diagnosis of Isolated Agenesis of Cranial Nerve VIII

Agenesia aislada del VIII par de diagnóstico tardío

M. José Lesmas Navarro,∗ Carlos De Paula Vernetta, Laura Cavallé Garrido

Servicio de Otorrinolaringología, Hospital Universitario La Fe, Valencia, Spain

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Hearing loss is one of the most frequent reasons for attending an otorhinolaryngology clinic. When suspected, it is essential to conduct a complete audiological study and complement this with suitable imaging techniques. Computed tomography (CT) is extremely sensitive for the assessment of the internal auditory canal (IAC), while nuclear magnetic resonance (NMR) provides much more detailed information on its contents, as a normal-sized IAC on a CT scan might present absence of cranial nerve VIII and NMR is the only means for its detection.

We report the case of an 11-year-old boy referred to our department due to suspected hearing loss in the left ear (LE), detected by chance by his parents. He presented no familial history or risk factors for hearing loss. Language acquisition was normal and his adaptation to schooling was adequate for his age. There was no prior history of recurrent...
otitis media or vestibular symptoms. Otoscopy was normal in both ears.

In the pure tone threshold audiometry, normal hearing was observed in the right ear (RE) and cophosis in the LE. Transitory otoacoustic emissions were positive in RE and negative in LE.

The NMR scan requested revealed correctly shaped vestibular and cochlear apparatuses with patency of the membranous labyrinths and a normal-sized IAC, but the absence of any visibility of the cochlear nerve (CN) was observed inside the left IAC on both the coronal (Fig. 1) and axial (Fig. 2) slices, corresponding to stage IIb in Casselman’s classification.

NMR has currently displaced CT scans for the imaging diagnosis of hearing loss, as aetiologies that previously went unnoticed, such as the agenesis of the cochlear nerve for example, can today be diagnosed.

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