

CASE SERIES AND REPORTS

Audiological findings in a case of cerebellar angioreticuloma

Risvolti audiologici nell'angioreticuloma cerebellare

A. SERRA¹, P. DI MAURO¹, S. COCUZZA¹, I. CHIARAMONTE², L. MAIOLINO¹¹ Department of Otorhinolaryngology, University of Catania, Italy; ² Department of Radiology, Unit of Neuroradiology, University of Catania, Italy

SUMMARY

The aetiology of neurosensorial damage with unilateral hearing loss and/or tinnitus and dizziness can often be difficult to determine because they may be caused wide variety of pathologic processes and a variety of diagnostic tests are needed in initial evaluation. In this paper, the authors describe, the techniques and indications of neuroimaging for evaluation of auditory symptoms. Auditory brainstem response (ABR) testing is still the choice when condition is suspected. We present a study of one patient with unilateral tinnitus, with no significant hearing loss and normal ABR: the patient underwent MRI of the district brain and the internal auditory channel (AIC) that showed the presence of a rare intracranial neoplasm, namely cerebellar angioreticuloma.

KEY WORDS: MRI • ABR • Tinnitus • Cerebellar angioreticuloma

RIASSUNTO

Nella pratica clinica l'eziologia della sintomatologia audiologica e/o vestibolare può spesso essere difficile da determinare, poiché svariati processi patologici possono esserne responsabili e numerosi test diagnostici vengono utilizzati nella valutazione iniziale. In questo lavoro, gli autori descrivono in dettaglio un caso clinico caratterizzato da assenza di significativa ipoacusia neurosensoriale, presenza di acufeni monolaterali e normalità dell'esame ABR. Il paziente è stato sottoposto a Risonanza Magnetica Nucleare del distretto encefalico e dei condotti uditivi interni. L'indagine neuroradiologica ha rilevato la presenza di una neoplasia intracranica rara, l'angioreticuloma cerebellare.

PAROLE CHIAVE: Risonanza Magnetica • Potenziali evocati uditivi • Acufeni • Angioreticuloma cerebellare

Acta Otorhinolaryngol Ital 2015;35:212-214

Introduction

Despite the low incidence of retrocochlear diseases, diagnosis of these conditions is frequent in clinical practice because about 30% of ENT patients refer audiology symptoms such as unilateral hearing loss or balance disorders. Unilateral sensorineural hearing loss, unilateral tinnitus and balance disorders are necessary to suspect a retrocochlear pathology: the most common cause is acoustic neuroma, which represents approximately 80-90% of all tumours of the cerebellopontine angle (PCA)¹, with an incidence ranging from 1/50,000 to 1/81,000 new cases a year².

The evolution of the traditional diagnostic protocol, represented by physical ENT examination, tonal and vocal audiometry, acoustic stapedial reflexometry, ENG, TEOAE and auditory evoked potentials (ABR)³.

Thanks to the excellent morphological definition, there is potential ability to identify lightly symptomatic disease characterised by normal electrophysiological pattern. Thus, radiologic methods have high specificity and sensitivity in early diagnosis of retrocochlear disorders⁴.

Despite the technical evolution of computed tomography (CT) and magnetic resonance imaging (MRI), and the consequent easier diagnosis of diseases of the skull base and petrosal bone, it is not possible to exclusively rely on these imaging techniques in the evaluation of retrocochlear pathology, also considering that these techniques are expensive. For these reasons, the investigation by MRI cannot be considered as a valid screening method.

Herein, the authors present a clinical case of a patient with unilateral tinnitus with no significant hearing loss and normal ABR: the patient was to undergo MRI of the brain district and internal auditory channel (AIC).

Case report

The study was conducted on a male patient 38 years of age at the Department of Otorhinolaryngology of the University of Catania. The patient complained of the presence of unilateral high tone tinnitus in the left ear, with progressive increase in the time of the intensity of tinnitus. Tinnitus over time had become pulsatile.



Fig. 1. Cerebellar angioreticuloma (FSE-T2 weighted image MRI).



Fig. 2. Left anterior inferior cerebellar artery hyperplasia (angio MRI 3D).

The patient underwent otoneurological anamnesis, tonal audiometry, stapedal reflexometry, TEOAE and auditory evoked potentials (ABR).

The patient underwent neuroradiological evaluation of the brain and internal auditory canals (IAC) with MRI.

The MRI study was conducted with GE equipment (Signa Excite HD) 1.5 T with FSE T2-weighted sequences, FSE T1-weighted, FLAIR, DWI, T2 * GE on the three orthogonal planes for the brain with a thickness of layer equal of 5 mm and interval of 0.5.

For study of IAC, sequences on axial and coronal planes with a slice thickness of 2 mm without interval, with technical FSE T2-weighted, T1-SE, with saturation of the fat signal, acquired on axial and coronal plans were used.

3D-FIESTA sequences were acquired in axial plans, and these images were subsequently processed in MPR 1 mm of para-sagittal and coronal images. The parameters of the 3D-FIESTA sequences were the following: in patients with likely expansive lesions, after obtaining informed consent, we proceeded to study with intravenous administration of gadolinium contrast (Dotarem) at a dose of 0.2 ml per kg of body weight⁵, and acquisition of axial and coronal SE T1-weighted sequences and sagittal 3D - FSPGR and coronal and para-sagittal MPR. In patients who could not or refused to undergo MRI, we proceeded with high-resolution CT of the petrosal bone with GE equipment (Brilliance 64) with the following parameters: FOV 18 × 18, electronic window for bone, thickness 0.65 mm, layer 0.65 mm, rotation time 1 sec, 140 kV, 240 mA. One mm coronal and para-sagittal MPR images were constructed (for a more accurate evaluation of IAC) and the original images were reformatted with

a FOV of 22 × 22 cm, an electronic window for soft tissue thickness 1.2 mm and layer 0.65 mm.

Vascular evaluation was conducted with Siemens angiography with selective catheterisation of the carotid and vertebral arteries.

The audiological evaluation by tonal audiometry revealed the presence in the patient of mild sensorineural hearing loss in the left ear. The mild hearing loss was centered on high frequencies. Impedanzometric examination detected the presence of tympanogram of type "A" and normal stapedal reflexes. The otoacoustic emissions were present bilaterally. Examination by ABR detected the normal morphology of the track and normality of the relative and absolute latency of the waves.

Neuroradiological imaging recognised the presence of a neoplastic formation in the cerebellar with characteristics similar to those of cerebellar angioreticuloma (Fig. 1). MRI examination was completed with 3D angiographic sequences (Fig. 2) and subsequently with selective angiography (Fig. 3). Angiographic sequences identified the presence of hyperplasia of the left anterior inferior cerebellar artery (AICA), a phenomenon which makes it the case even more rare just for the type of unusual vascularisation of the cerebellar angioreticuloma. Selective angiography was also subsequently performed that identified the presence of hyperplasia of left cochlear artery, which was normally not visible in the other conditions.

The patient was subjected to neurosurgical removal of angioreticuloma. Audiological examinations, after neurosurgery, were unchanged from the initial assessment. The



Fig. 3. Left anterior inferior cerebellar artery hyperplasia and cochlear artery hyperplasia (selective angiography MRI).

patient also noticed the reduction and disappearance of pulsatility of tinnitus.

Discussion

Unilateral audiological symptoms are among the most common causes of ENT evaluation, and dizziness and imbalance are also frequent in clinical practice. Many retrocochlear pathologies, such as vestibular tumours, congenital and acquired demyelinating diseases (e.g. multiple sclerosis) and neurovascular conflicts, can be associated with ENT symptomatology during their natural history. ABR permits the diagnosis of retrocochlear disorders that are otherwise not recognisable without diagnostic imaging. From a review of the literature there are contrasting results about the specificity and sensitivity of ABR for detection of intra- and extra- canal tumours⁶. In our study, a patient presented with cerebellar angioreticuloma that was diagnosed by neuroradiological evaluation. Examination by ABR did not detect the presence of morphological abnormalities or timing of absolute and relative latency of the waves. This consideration does not diminish the validity of ABR for the diagnosis of retrocochlear disorders, and the same finding may be present when the damage affects only the CNS structures with balance function, even without audi-

tory system alteration. Other studies attribute a large number of false positives to ABR⁷.

Thus, MRI with Gd permits better reliability in the identification of retrocochlear and central auditory pathway diseases for its excellent morphological definition, especially for some symptomatic lesions with normal electrophysiological pattern, and the MRI can also evaluate cochlear and retrocochlear disorders, such as IAC and PCA alterations⁸. For patients with a high clinical suspicion, MRI can and should remain the gold standard of diagnosis as ABR may miss smaller and/or intracanalicular tumours⁹.

Initial symptoms are sometimes underestimated and the patient is not always referred to radiological investigation. It is always necessary to maintain a high index of diagnostic suspicion to decrease the interval between the first symptoms and final diagnosis.

We believe that this strategy allows adequate diagnosis of retrocochlear disorders, such as small lesions of the IAC and PCA, with the possibility of effective and early medical and surgical techniques aimed at the preservation of hearing.

References

- 1 Jackler RK, Pitts LH. *Acoustic Neuroma (Vestibular Schwannoma)*. In: *Neurotology*. St Louis: Mosby; 1994. p. 729-85.
- 2 Tos M, Thomsen J. *Incidence of vestibular schwannoma*. *Laryngoscope* 1999;109:736-40.
- 3 Hendrix RA, Robert M, Sclafani AP. *The use of diagnostic testing in asymmetric sensorineural hearing loss*. *Otolaryngol Head Neck Surg* 1990;103:593-8.
- 4 Sedwick JD, Gajewski BJ, Prevatt AR, et al. *Magnetic resonance imaging in the search for retrocochlear pathology*. *Otolaryngol. Head Neck Surg* 2001;124:652-5.
- 5 Plaza G, Lopez Lafuente J, Aparicio JM, et al. *Resonancia Magnética: Prueba de elección en el depistaje de tumores del conducto auditivo interno y ángulo pontocerebeloso*. *Acta Otorrinolaringol Esp* 2001;52:651-6.
- 6 Day AS, Wang CT, Chen CN, et al. *Correlating the cochleovestibular deficits with tumor size of acoustic neuroma 1*. *Acta Otolaryngol* 2008;7:756-60.
- 7 Furman JM. *Otoneurology an Issue of Neurologic Clinics*. Saunders Ed; 2005.
- 8 Oh JH, Chung JH, Min HJ, et al. *Clinical application of 3D-FIESTA image in patients with unilateral inner ear symptom*. *Korean J Audiol* 2013;17:111-7.
- 9 Koors PD, Thacker LR, Coelho DH. *ABR in the diagnosis of vestibular schwannomas: a meta-analysis*. *Am J Otolaryngol* 2013;34:195-204.

Received: December 1, 2014 - Accepted: March 8, 2015

Address for correspondence: Agostino Serra, University of Catania, Department of Otorhinolaryngology, A.O. "Policlinico Vittorio Emanuele", via Santa Sofia 78, 95123 Catania. Tel. +39 095 3781093. Fax +39 095 7335738. E-mail: serra@policlinico.unict.it