CASE REPORT

Sudden cochlear hearing loss as presenting symptom of arachnoid cyst of the posterior fossa

Cisti aracnoidea della fossa cranica posteriore ed ipoacusia improvvisa

G. CADONI, S. AGOSTINO, M. VOLANTE, M.S. SCIPIONE
Department of Otorhinolaryngology, Catholic University of Sacred Heart, Rome, Italy

Key words

Cranial posterior fossa • Arachnoid cyst • Sudden hearing loss

Parole chiave

Fossa cranica posteriore • Cisti aracnoidea • Ipoacusia improvvisa

Summary

Arachnoid cysts account for almost 1% of neoformations located in the cerebellopontine angle. The aetiopathogenesis is unknown. Arachnoid cysts of the cranial posterior fossa may produce symptoms typical of a tumour such as headache, dizziness, tinnitus and progressive sensorineural hearing loss. Management of these lesions is still controversial; if the arachnoid cyst is symptomatic, surgical treatment is usually recommended. The case is described of an adult female with sudden unilateral cochlear hearing loss as presenting symptom of an arachnoid cyst in the cranial posterior fossa.

Riassunto

La cisti aracnoidea rappresenta quasi l'1% delle neoformazioni dell'angolo pontocerebellare. L'eziopatogenesi non è nota. La cisti aracnoidea della fossa cranica posteriore può manifestarsi con sintomi come cefalea, instabilità, acufene ed ipoacusia neurosensoriale progressiva. Il trattamento di queste neoformazioni è ancora controverso, l'indicazione è chirurgica se la cisti è sintomatica. In questo lavoro abbiamo descritto il caso di una donna affetta da cisti aracnoidea della fossa cranica posteriore esordita con ipoacusia improvvisa monolaterale.

Introduction

The cranial posterior fossa is the second most common site for arachnoid cysts (AC) that account for approximately 0.4-0.8% of all cerebellopontine angle lesions [1].
The aetiopathogenesis of AC is unknown; a post-traumatic and infectious aetiology has been suggested, but the most acceptable hypothesis to explain their origin appears to be a congenital developmental malformation resulting in splitting of the arachnoid membrane [2].
AC of the posterior fossa may produce symptoms typical of a tumour, such as headache, dizziness, tinnitus and progressive sensorineural hearing loss [3 4].
Computed tomography (CT) and magnetic resonance imaging (MRI) have led to an improvement in the pre-operative diagnosis of AC even if accurate diagnosis is still difficult and may be mistaken for epideromid cysts or other cystic lesions [5].
The management of AC is still controversial even considering that spontaneous disappearance has rarely been reported [6]. Surgical treatment is recommended for symptomatic AC of the cranial posterior fossa and radical complete resection is the most reliable long-term solution [7].
The case is described of an adult female with sudden unilateral cochlear hearing loss as the presenting symptom of an AC of the cranial posterior fossa.

Case report

A 32-year-old female came to our attention with a 3-day history of right sudden hearing loss and tinnitus. Clinical examination was normal. There was no history of ototoxicity, recent trauma, infectious disease, familiar hearing loss or neurologic disorders. Results of a complete physical head and neck examination were normal.
A normal autoimmune profile was obtained and blood count, serum biochemical screening, urinalysis, free treponemal antibody absorption test were all within normal limits.
Audiometry showed a right mild sensorineural hearing loss at high frequencies (Fig. 1). Eardrum and tympanometric results were normal. Analysis of auditory brainstem responses showed no significant increase in wave V latency (Fig. 2), and right distorsion products of otoacoustic emissions were absent at high frequencies (> 2 KHz) (Fig. 3). No vestibular dysfunction was found.
A combined regimen of steroids (methylprednisone 1 mg/kg/day), plasma expander (low molecular weight dextran 500 ml/day) and aspirin (100 mg/day) was prescribed until MR of the brain was performed.
CT scan of the middle ear showed no morphological alterations.

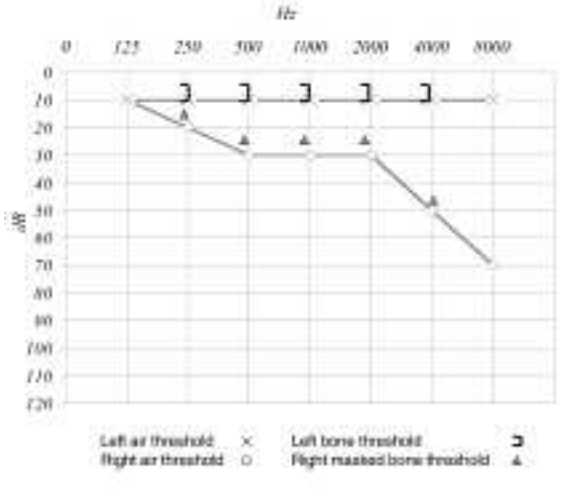

Fig. 1. Audiometry shows a right mild sensorineural hearing loss at high frequencies.

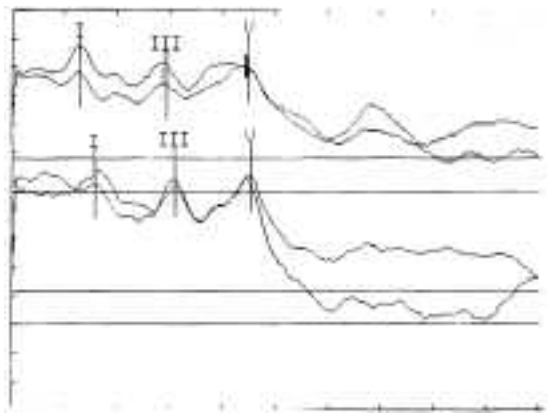

Fig. 2. Analysis of auditory brainstem responses showed no significant increase in wave V latency (left ear: superior traces, right ear inferior traces).

MR imaging revealed a posterior fossa AC (diameter 15 mm) located at the right cerebellopontine angle with compression of the VIII cranial nerve and postero-inferior cerebellar artery (Fig. 4).

We perform MRI using the FLAIR sequences technique. These sequences confirmed the diagnosis, and excluded other disorders such as epidermoid cysts. While epidermoid cysts appear hyperintense with the FLAIR sequences, ACs appear hypointense.

The patient was then examined by the neurosurgeons who chose conservative management with routine radiological monitoring to identify eventual growth of the cyst. Follow-up revealed no deterioration in the clinical conditions of the patient and a second MRI scan, one year after the initial diagnosis showed no change in the size of the AC.

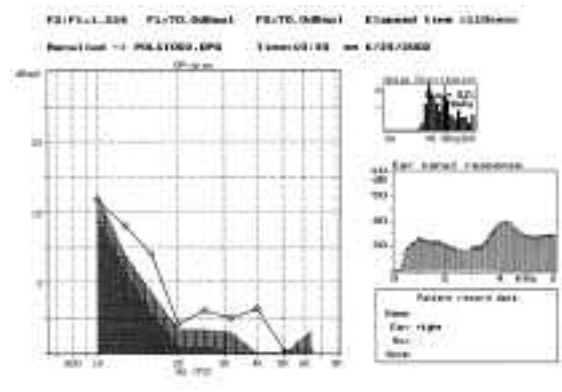

Fig. 3. Distorsion products otoacoustic emissions (DPOAEs) were absent at high frequencies (2 KHz) in right ear according to a cochlear SNHL.

Discussion

Sudden sensorineural hearing loss (SSHL) has been defined as a sensorineural hearing loss of > 30 dB over at least 3 contiguous audiometric frequencies, that develops in a period of a few hours to 3 days [9].

A specific cause of spontaneous SSHL can be identified in only 10-15% of cases. Numerous clinical and laboratory investigations have attempted to identify the cause of this disorder which, in fact, has numerous possible causes, which can be classified as infectious, traumatic, immunologic, toxic, circulatory, metabolic, neurologic and neoplastic [10].

Furthermore, sudden sensorineural unilateral hearing loss may result from causes affecting cochlea, eighth nerve, or more central auditory tracts.

In the present case, the analysis of auditory brainstem responses showed no significant increase in wave V latency, but distortion products otoacoustic emissions were absent suggesting a cochlear hearing loss. MRI (FLAIR sequences) of the brain permitted us to make a diagnosis of cochlear hearing loss accompanying a cranial posterior AC. Possible mechanisms responsible for the effect on the cochlea include degenerative changes due to chronic partial obstruction of the blood supply by the mass, biochemical alterations in the inner ear fluids, loss of efferent control of active mechanical tuning, and hair cell degeneration secondary to neuronal loss in the eighth nerve [11].

Few Authors have reported cases of AC located in the posterior fossa, the first clinical symptom of which was progressive retrocochlear hearing loss without any other symptoms typical of a cerebellar localization [12 13].

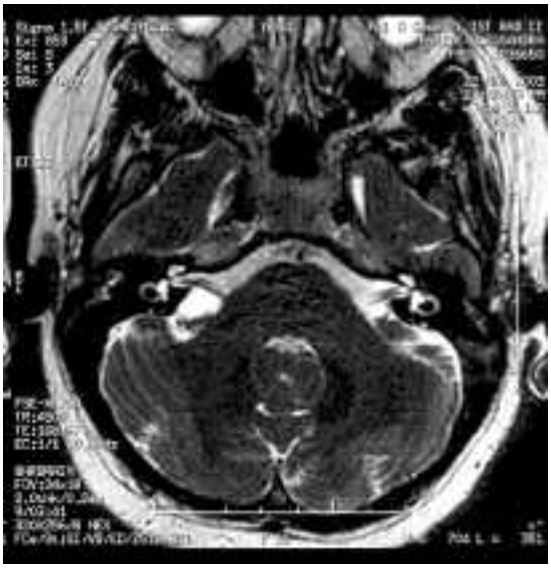

Fig. 4. Brain MR scan with gadolinium shows arachnoid cyst of cranial posterior fossa (diameter 15 mm) displacing both the right VIII cranial nerve and postero-inferior cerebellar artery (PICA) forward and laterally.

Much controversy exists regarding the treatment of ACs [7 13 14]. Symptomatic cysts of the posterior fossa may be treated by various surgical procedures following a sub-occipital craniectomy, such as drainage, total or partial removal of the cyst, shunting or fistulization of the cyst to the sub-arachnoid space. Stereotactic puncture and endoscopy decompression have also been successfully employed in the treatment of AC. Surgical treatment frequently improves vestibular symptoms but auditory deficits are less likely to respond to surgery.

As in our case, conservative management with regular radiological monitoring may be suggested in order to identify those cases with gradual enlargement of the cyst requiring surgical treatment.

In conclusion, occasionally, unilateral sudden sensorineural hearing loss (SSHL) could be diagnosed in association with an AC of the cranial posterior fossa. That confirms the importance of diagnostic imaging and, in particular, contrast-MR of the brain, in all cases of SSHL patients, even if a specific cause of spontaneous SSHL can be identified in only 10-15% of cases.

References

1 Yamakawa H, Pearlstein AE, Eiber A. *MR imaging of intracranial arachnoid cysts*. J Comput Assist Tomogr 1987;11:236-41.

2 Di Rocco C. *Arachnoid cysts*. In: Youmans JR, editor. *Neurological surgery, Edn 3*. Philadelphia: Saunders; 1990. p. 1299-325.

3 Schievink WI, Piepgras DG, Nichols DA. *Spontaneous carotid-jugular fistula and carotid dissection in a patient with multiple intracranial arachnoid cysts and hemifacial atrophy: a generalized connective tissue disorder?* J Neurosurg 1995;83:546-9.

4 Haberkamp TJ, Monsell EM, House WF, Levine SC, Piazza L. *Diagnosis and treatment of arachnoid cysts of the posterior fossa*. Otolaryngol Head Neck Surg 1990;103:610-14.

5 Tamas LB, Wyler AR. *Intracranial mucocele mimicking arachnoid cyst: case report.* Neurosurgery 1985;16:91-7.

6 Pandey P, Tripathi M, Sarat Chandra P, Singh VP, Mehta VS. *Spontaneous decompression of a posterior fossa arachnoid cyst: a case report.* Pediatric Neurosurgery 2001;38:162-3.

7 Samii M, Carvalho GA, Schuhmann MU, Matthies C. *Arachnoid cysts of the posterior fossa*. Surg Neurol 1999;51:376-82.

9 Pappas DG, Brackmann DE. *Arachnoid cysts of the posterior fossa*. Otolaryngol Head Neck Surg 1981;89:328-32.

10 Wilson WR, Byl FM, Laird N. *The efficacy of steroids in the treatment of idiopathic sudden hearing loss.* Arch Otolaryngol 1980;106:772-6.

11 Hughes GB, Freedman MA, Haberkamp TJ, Guay ME. *Sudden sensorineural hearing loss*. Otolaryngol Clin North Am 1996;29:393-405.

12 Prasher DK, Tun T, Brookes GB, Luxon LM. *Mechanisms of hearing loss in acoustic neuroma: an otoacoustic emission study.* Acta Otolaryngol 1995;115:375-81.

13 Ottaviani F, Neglia CB, Scotti A, Capaccio P. *Arachnoid cyst of the cranial posterior fossa causing sensorineural hearing loss and tinnitus: a case report.* Europ Arch Oto-Rhino-Laryngol 2002;259:306-8.

14 Hadley MN, Grahm TW, Daspit CP, Spetzler RF. *Otolaryngologic manifestations of posterior fossa arachnoid cysts*. Laryngoscope 1985;95:678-81.

15 Choi JU, Kim DS, Huh R. *Endoscopic approach to arachnoid cyst*. Childs Nerv Syst 1999;15:285-91.

16 Boltshauser E, Martin F, Altermatt S. *Outcome in children with space-occupying posterior fossa arachnoid cysts.* Neuropediatrics 2002;33:118-21.

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■ Address for correspondence: Dr. G. Cadoni, Istituto di Clinica ORL, Università Cattolica del Sacro Cuore, largo A. Gemelli 8, 00168 Rome, Italy. Fax: +39 06 3051194
E-mail: gabriella.cadoni@rm.unicatt.it