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(CASE REPORT)

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Case report: Cystic vestibular schwannoma

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Abstract

Background: Cerebellopontine angle tumors are a severe condition because of their life-threatening and functional loss potential, explained by the narrowness of the posterior fossa.

The vestibular schwannoma, or acoustic neuroma, is a benign, well-circumscribed extraaxial tumor that arises from Schwann cells of the vestibulocochlear cranial nerve. The tumor is usually mainly solid, but can sometimes develop small cystic areas, due to degenerative modifications, causing microcyst formation.

The coalescent of small cysts leading to forming a larger one, and responsible of a predominant cystic contingent, is rare, which makes interesting the case we are about to present.

Case report: We are about to report the case of a 64-year-old male patient, with no particular medical history, which clinical interview has noted only tinnitus as a present symptom. He had experienced one year ago several disequilibrium and dizziness episodes, who have subsequently stopped over time, without any particular treatment. He also felt a little loss of hearing, especially in the left ear, but thought it was probably age related. No clinical abnormality including the facial nerve palsy was found.

The MRI showed a stage 4 (Koos classification) left cystic vestibular schwannoma. This lesion was compressing the left vertebral and basilar arteries, the brain stream, the left cerebellar hemisphere, the left middle cerebellar peduncle, and the fourth ventricle, causing a triventricular hydrocephalus.

After reviewing all potential therapeutic options, the patient declined surgery and decided to postpone the treatment.

Conclusion: MRI is definitely the best way to explore the posterior fossa, especially the cerebellopontine angle, and helps us differentiate between all numerous tumors than can develop in this space.

Keywords: Schwannoma; Cystic; Vestibulocochlear nerve; Cerebellopontine angle; MRI

1. Introduction

Cerebellopontine angle tumors are a severe condition because of their life-threatening and functional loss potential, explained by the narrowness of the posterior fossa.

The most frequent tumor in this anatomical area is the acoustic schwannoma. It represents 6 to 8% of intracranial tumors, 25% of posterior fossa tumors, and 80% to 92% of cerebellopontine angle tumors [1,2,3], followed by meningiomas [4].

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The coalescent of small cysts leading to forming a larger one, and responsible of a predominant cystic contingent, is rare, which makes an interesting case we are about to present.

The patient is usually around 30 to 50 years old, and suffers from neurosensory symptoms such as hearing problems, tinnitus, equilibrium abnormalities, and vomiting.

MRI is the main imaging diagnosis tool for cerebellopontine masses, and usually helps differentiating this affection from other differential diagnosis.

Treatment option for cystic schwannomas include a wait-and-see approach, surgery, and radiosurgery, depending on the tumor size, growth rate, vascularization and location.

2. Case report

We report the case of a 64-year-old male patient, with no particular medical history, which clinical interview has noted only tinnitus as a present symptom. He had experienced one year ago several disequilibrium and dizziness episodes, who have subsequently stopped over time, without any particular treatment. He also felt a little loss of hearing, especially in the left ear, but thought it was probably age related. No clinical abnormality including the facial nerve palsy was found.

With the discomfort caused by tinnitus, he decided to consult an otorhinolaryngologist. An audiogram was performed and revealed a sensorineural hearing loss (SNHL), with a 65 dB loss on the left ear, which led the physician to prescribe an MRI (Figures 1 to 8), showing a voluminous extraaxial lesion, located on the left cerebellopontine angle, with smooth and regular margins, measuring 48 x 30 x 48 mm. The lesion was mainly cystic-appearing; moderately hypointense on T1-weighted images (Figure 1), and had a liquid hypersignal on T2-weighted images (Figure 2), which did not disappear on FLAIR sequence (Figure 3). No modification was seen on FAT SAT (Figure 4).

We also individualized a small solid enhanced contingent, developed in the internal auditory canal, causing an enlargement of the porus, and extended into the cerebellopontine angle (Figure 5).

Diffusion showed no restriction and the apparent diffusion coefficient (ADC) was high (Figure 6).

This mass had also hemorrhagic stigmata, revealed on T2* (Figure 7), T2 (Figure B) and FLAIR sequences (Figure 3) with a fluid-fluid level.

This lesion was compressing the left vertebral and basilar arteries, the brain stream, the left cerebellar hemisphere, the left middle cerebellar peduncle, and the fourth ventricle, causing a triventricular hydrocephalus (Figure 8).

The diagnosis retained was a stage 4 (Koos classification) left cystic vestibular schwannoma.

After reviewing all potential therapeutic options, the patient declined surgery and decided to postpone the treatment. He still hasn't have surgery until today, still has the same symptoms but without worsening.

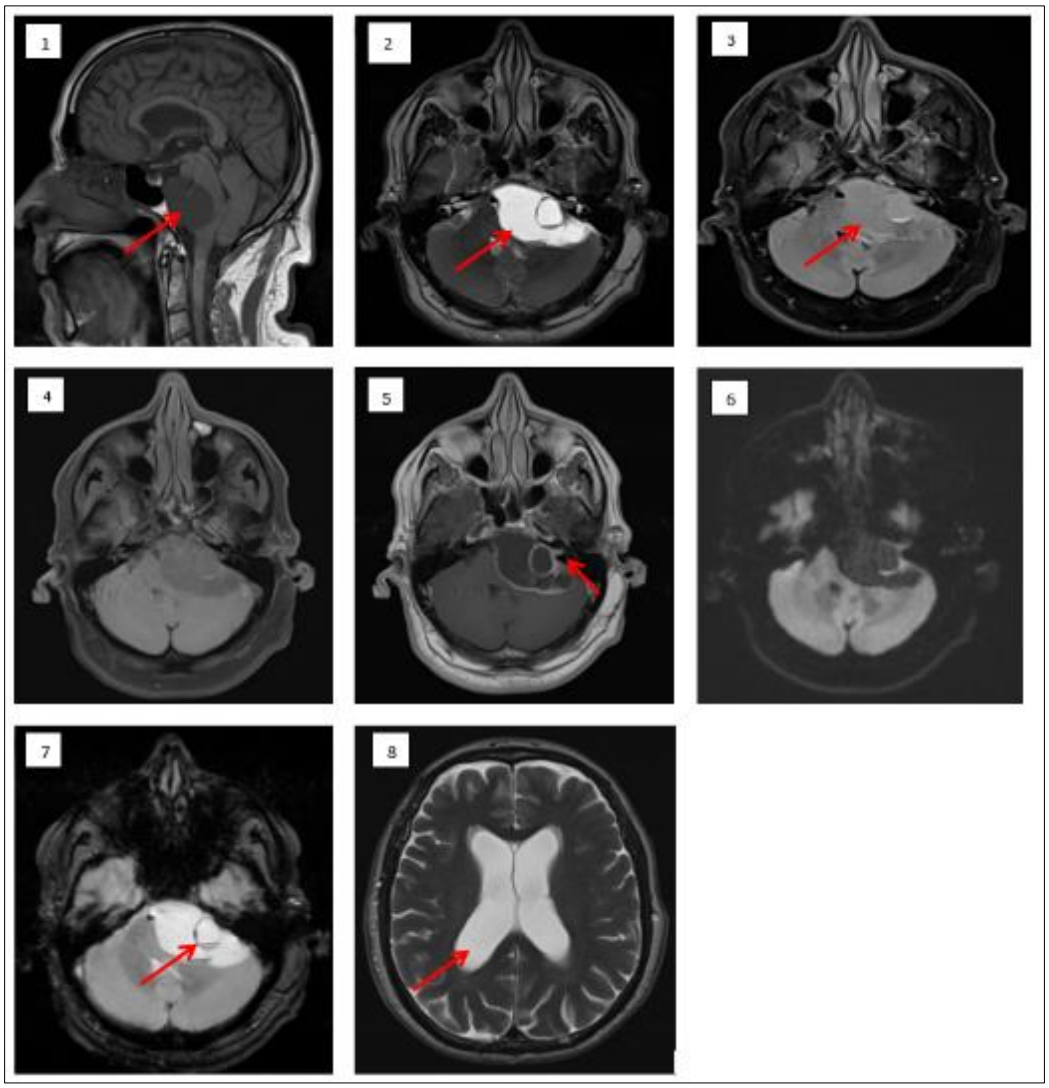

Figure 1 Voluminous mainly cystic extraaxial lesion, located on the left cerebellopontine angle, moderately hypointense on T1-weighted images (1, arrow), with a liquid hypersignal on T2-weighted images (2, arrow), which didn't disappear on FLAIR sequence (3, arrow). No modification was seen on FAT SAT (4). Small solid enhanced contingent (E, arrow), developed in the internal auditory canal, causing an enlargement of the porus, and extended into the cerebellopontine angle. Diffusion (6) showed no restriction and the apparent diffusion coefficient (ADC) was high. Hemorrhagic stigmata revealed on T2* (7, arrow), T2 (2) and FLAIR sequences (3), with a fluid-fluid level

This lesion is compressing the left vertebral and basilar arteries, the brain stream, the left cerebellar hemisphere, the left middle cerebellar peduncle, and the fourth ventricle (1, 2, 3, 4, 5) , causing a triventricular hydrocephalus (8, arrow).

3. Discussion

The cerebellopontine angle (CPA) is a triangular or prismatic subarachnoid space, whose boundaries are made up by the brain stream (pons and medulla) medially, the petrous portion of the temporal bone laterally, the tentorium superiorly, the occiput inferiorly, and the anterior part of the cerebellum.

This space contains CSF, cranial nerves (Higher group: V and VI, middle group: VII, VIII, lower group: IX, X, XI) and vessels (superior cerebellar artery, anterior inferior cerebellar artery AICA, posterior inferior cerebellar artery PICA, labyrinthine artery, and veins draining into the lower petrous sinus or directly into the internal jugular vein).

A spectrum of usual and unusual lesions exists in the CPA [4], and to set them apart, it is imperative to analyze their center, origin, shape, and extent. The symptoms related to CPA lesions are usually not related to the histology of the mass, but to the structures that the lesion affects.

The vestibulochochlear (VC) nerve (or eighth cranial nerve) is purely sensory, responsible for balance and hearing [5]. It has two major parts, the cochlear and the vestibular nerve. Unlike other cranial nerves, the VC nerve can be best described from outside to inside [5].

The cochlear nerve arises from the spinal ganglion in the cochlea. Its fibers pass into the internal auditory canal IAC, to reach the porus acusticus. The vestibular nerve arises from bipolar neurons in the vestibular ganglion at the IAC fundus. Its fibers confluence to form a superior and inferior nerve, that course medially to reach the porus acusticus, and join, with the cochlear nerve, to form the eighth cranial nerve.

The vestibular schwannoma (VS) is a relatively frequent intracranial tumor, and is by far the most common tumor of the CP angle [1,2,3]. In fact, and according to studies [6,7], VS develop from the vestibulocochlear nerve much more often than from any other nerve. Additionally, the involvement of vestibular division is way more important than the cochlear.

The vestibular schwannoma begins deep in the CAI, grows slowly initially, and remains intracanal, then goes through the porus, to grow in the APC.

The age is typically between 30 and 50 years of age, and the lesions are sporadic, except for patients with neurofibromatosis type II, among which we find an earlier presentation and typically bilateral vestibular schwannomas as a nearly pathognomonic sign, sometimes associated with meningiomas and ependymomas. There is no sex predilection [5].

Symptoms are usually slowly progressive and result from impingement of surrounding structures and from direct involvement of the nerve [8]. Usually the patient experiences unilateral hearing abnormalities, tinnitus, disequilibrium, headache, and vomiting [7]. Facial nerve manifestations (weakness, twitching) are relatively rare in case of a small tumor. On the other hand, larger tumors can be responsible for deficits involving cranial nerves, cerebellar manifestations, and symptoms related to hydrocephalus [10].

It is not uncommon for cystic areas to develop within schwannomas [8]. In fact, cystic schwannomas are estimated to represent between 6 and 48% of all vestibular schwannomas combined [2].

The cystic component is essentially due to degenerative changes caused by tumor growth with central necrosis, repeated intratumoral hemorrhage, confluence of mucinous cavities, or most usually coalescence of microcysts [11,12].

They differ from solid schwannomas essentially by their rapid growth [2], their aggressiveness, shorter asymptomatic periods, and acute evolution of manifestations (due to compression of surrounding structures). They also have more frequent involvement of the facial nerve [3], unpredictable and low surgery outcomes (such as incomplete resection, unfavorable facial nerve function, and more postoperative hemorrhage [3]), and high morbidity.

Therefore, cystic vestibular schwannomas are severe because of their lethal potential. Their rapid expansion can cause, like in our case, a brain stream compression and hydrocephalus.

MRI is considered to be the best investigation tool for cerebellopontine angle masses, because of its high contrast resolution, and is far superior to CT scan in analyzing the posterior fossa. The aim of imaging is to determine the tumor location, its shape and margins, extent (internal auditory meatus IAM, brain stream, and cerebellar hemispheres), mass effect, and adjacent bone reaction [3].

The imaging features include smooth margins, possible bone remodeling without infiltration or lysis, and sometimes, like in our case, fluid-fluid levels.

The fluid-filled component of the tumor is typically hyperintense on T2 and FLAIR sequences, with sometimes an enhanced cystic wall after gadolinium injection, whereas the solid portions are normally isointense or mildly hypointense relative to the pons on T1-weighted images, and enhanced intensely after intravenous administration of gadolinium. Hemorrhage is rare and can sometimes be seen as a T1 hyperintensity or hypointensity on T2* weighted sequences.

Cysts vary in size and number, but still have the same MRI characteristics.

Other lesions can be developed in the CPA such as lipomas (T1 and T2 hyperintensity, no enhancement, and low signal on fat saturated sequences), epidermoid cysts (usually T1 and T2 isointense to CSF, higher signal compared to CSF around the periphery of the lesion, thin enhancement around the periphery, very bright on DWI), dermoid cysts (typically T1 hyperintense due to lipid components, variable signal on T2, no enhancement on T1 C+) , chondromatous tumors, paragangliomas, and arachnoid cysts (isointense compared to CSF on all sequences), but the major differential diagnosis of solid vestibular schwannoma is the meningioma, and differentiating the two lesions can sometimes be difficult. However, the ability to center the lesion over the IAC is in favor of vestibular schwannoma.

In our case, and because the tumor was mainly cystic, the first differential diagnosis was the arachnoid cyst, but the small solid contingent developed in the porus, and the persistence of the hypersignal on the cystic component, made us rule out the arachnoid cyst hypothesis.

Treatment options for cystic schwannomas include observation with a wait-and-see approach, surgery, radiosurgery, or a combination of these last two options, and we highlight the importance that should be given to preserve the facial nerve. Certain criteria determine the treatment modality, such as tumor size, growth rate, hypervascularity, location, and hearing level [3].

4. Conclusion

We reported a case of a patient with a stage 4 cystic vestibular schwannoma, which is an uncommon form of neuroma.

Clinical features include mainly hearing loss, dizziness, and tinnitus.

MRI is definitely the best way to explore the posterior fossa, especially the cerebellopontine angle, and helps us differentiate between all numerous tumors than can develop in this space.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study. Yes

References

[1] Gilbert Dechambenoit, Tumeurs cérébrales. Manuel de neurochirurgie. Sauramps

[2] E. Piccirillo, M. R. Wiet, S. Flanagan, F. Dispenza, A. Giannuzzi, F. Mancini, M. Sanna Cystic vestibular schwannoma : Classification, management, and facial nerve outcomes. Otology & Neurotology. 30:826-834 © 2009, Otology ² Neurotology, Inc.

[3] J. H. Han, K. H. Baek, Y. W. Lee, Y. K. Hur, H. J. Kim, I. S. Moon. Comparison of clinical characteristics and surgical outcomes of cystic and solid vestibular schwannomas.Department of Otorhinolaryngology, Yonsei University College of Medicine, Seoul; and Department of Otorhinolaryngology - Head and Neck Surgery, Inha University College of Medicine, Incheon, Korea Otology & Neurotology. 39: e381-286 © 2018, Otology & Neurotology, Inc.

[4] K. Sing, M. P. Sing, C.L. Thukral, K. Rao, K. Singh, A. Singh Role of magnetic resonance imaging in evaluation of cerebellopontine angle schwannomas Association of Otolaryngologists of India 2014. Indian J Otolaryngol Head Neck Surg (Jan-Mar 2015) 67(1):21-27; DOI 10.1007/s12070-014-0736-0

[5] Anne G. Osborn, G. L. Hedlund, K. L. Salzman Osborn's Brain: Imaging, pathology, and anatomy. Second edition: Chapter 23: Cranial nerves and nerve sheath tumors Universsity of Utah School of Medicine, Salt Lake City, Utah.

[6] J.D Clemis, W.J. Ballad, P.J. Baggot, S.T. Lyon Relative frequency of inferior vestibular schwannoma. Arch Otolaryngol Head Neck Surg 112(2): 190-194

[7] Nerve origin of the acoustic neuroma. A. Komatsuzaki, A. Tsunoda J. Laryngol Otol 115(5): 376-379.

[8] P. Catalano, E. Fang-Hui, P. M. Som Fluid-fluid levels in benign neurogenic tumors. AJNR 18:385–387, 0195-6108/97/1802–0385 © American Society of Neuroradiology

[9] R. Brow. Pre and postoperative management of the acoustic tumor patient. Management, vol 2. University Park Press, Baltimore.

[10] R.G Hart, D.P. Gardner, J. Howieson Acoustic tumors: atypical features and recent diagnostic tests.Neurology 33(2); 211-221.

[11] S. Charabi, L. Klinken, M. Tos, et al. Histopathology and growth pattern of cystic acoustic neuromas.

[12] C.K. Park, D.C Kim, S.H. Park, et al. Microhemorrhage, a possible mechanism for cyst formation in vestibular schwannomas.

[13] S. Wandong, L. Meng, L. Xingang, L. Yuguang, Z. Shugan, W. Lei, W. ChengyuanCystic acoustic neuromaDepartment of Neurosurgery, Qilu Hospital of Shandong University, Jinan, PR China.Journal of clinical neuroscience 12(3), 253-255. 0967-5868/$ © Elsevier Ltd. DOI: 10.1016/j.ocn.2004.03.040