ORIGINAL RESEARCH PAPER

Otorhinolaryngology

A RARE CASE REPORT OF ARACHNOID CYST OF THE POSTERIOR FOSSA PRESENTING WITH UNILATERAL SENSORINEURAL HEARING LOSS

KEY WORDS: Arachnoid cyst, hearing loss, cerebellopontine angle.

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ABSTRACT

Arachnoid cyst are rare intracranial tumors accounting for 1% of all brain lesions. Symptomatic arachnoid cyst of CP angle are even a rarer occurrence.CPA arachnoid cyst have a female preponderance and are progressive [1]. Arachnoid cysts of the posterior fossa may produce vague symptoms typical of a tumor, such as headache, dizziness, tinnitus, progressive sensorineural hearing loss, and neurovascular deficits. Management of these lesions is controversial; if the arachnoid cyst is symptomatic, surgical treatment is known to alleviate symptoms in 95% of the patients.The recurrence rate is approximately 10%, which warrantes regular follow-up in post-operative cases. Here, we report a case of a 19-year-old female presenting with right-sided hearing loss due to right cerebellomedullary cistern arachnoid cyst.

INTRODUCTION

Arachnoid cysts are benign pouchlike CSF-filled intraarachnoid masses. It can be primary, in which case it develops due to the splitting of the arachnoid membranes and the resulting collection of CSF. It can be secondary to infection, trauma, intracranial hemorrhage, or following surgery[7].

Arachnoid cysts are mostly non-syndromic but sometimes may be associated with syndromes acrocallosal, Aicardi, and Pallister-Hall syndromes[2]. They represent only 1% of all intracranial lesions and are most commonly found in the middle cranial fossa. Arachnoid cysts are most commonly observed in the Sylvian fissure, followed by the cerebellopontine angle (CPA) with approximately 10% frequency [3]. Signs and symptoms of arachnoid cysts vary depending on their anatomical location and size. Supratentorial cysts may cause untreatable headaches, difficulties in concentration, temporal lobe epilepsy, and, in pediatric patients, bulging of the parietal bone. Posterior fossa arachnoid cysts may manifest with hypoacusis, tinnitus, dysmetria, ataxia, sometimes vomiting and papilloedema from raised Intracranial tension, from fourth ventricle obstruction[4,5,8].

Case

A 19-year-old female patient came to our hospital with complaints of right-sided hearing loss, which was gradually progressive for one year. She had no history of Dizziness or Headache. There were no neurological symptoms. There was no history of infections or trauma in the recent past. On examination bilateral Tympanic membrane was intact, and neurological examination was normal. There was no nystagmus or cerebellar signs.

Audiometry evaluation showed profound hearing loss in the right ear of 85 dB (with masking >113.3 dB Loss). ABR of the right ear showed no peaks. DPOAE was found to be absent in the right ear. Due to the asymmetric nature of hearing loss, MRI Brain with Gadolinium contrast was ordered which showed an Arachnoid cyst at the right cerebello medullary cistern measuring 2.2x1.5x1.9 cm causing compression and displacement of the cisternal part of the right vestibulo cochlear nerve, cisternal segment of the right superior cerebellar artery superiorly and mild mass effect on the medulla and the lesion showed no post-contrast enhancement.

A neurosurgery consult was obtained, and the patient was advised to wait and watch with a review MRI after three months.

DISCUSSION

Symptomatic Arachnoid cyst of cerebellopontine angle is rare[1]. Arachnoid cysts are mostly asymptomatic, if symptomatic they usually present with vague symptoms, headache, dizziness, or generalized unsteadiness[4]. CP angle Arachnoid cysts have a female preponderance[1]. Clear otoneurological symptoms of uni- and bilateral hearing loss, giddiness, and tinnitus have also been reported. In this patient, after clinical examination and preliminary audiovestibular evaluation, retrocochlear pathology was observed.MRI Brain with contrast was advised. MRI is adequate to reveal the presence of arachnoid cysts and for evaluation of the compression of cysts on neural structures if present[10].

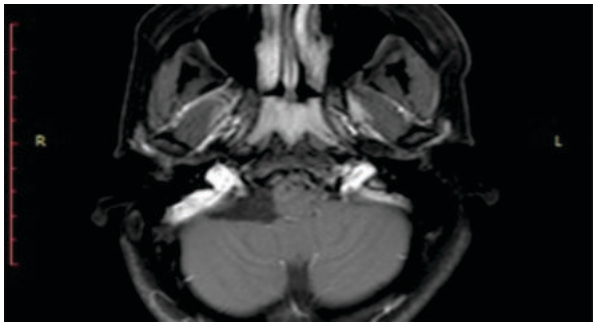

Figure 1: a)Axial T1-weighted MRI shows an arachnoid cyst with signal intensity similar to that of CSF.

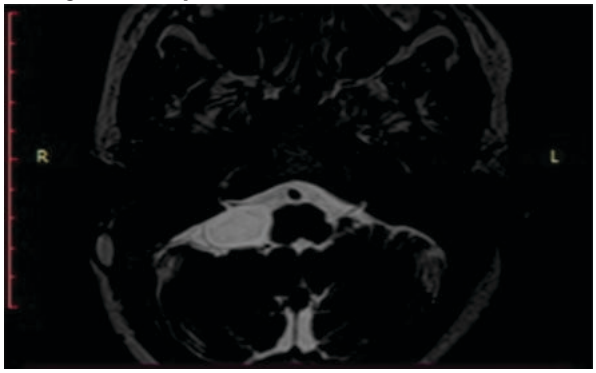

Figure 2: (b) Axial T2-weighted MRI shows the cyst displacing the vascular structures of the CPA.

In our patient, an arachnoid cyst located in the cerebellar convexity led to compression of the homolateral CPA. Pappas and Brackmann et al [6] reported cases of arachnoid cysts of the posterior fossa whose first clinical symptom was retrocochlear hearing loss without any other symptoms of cerebellar dysfunction. Posterior fossa arachnoid cysts presenting with sudden hearing loss were reported by Cadoni et al. (2006). The absence of DPOAE on the right side denotes that the cochlear impairment was also involved in the genesis of the hearing loss, which may be a consequence of cochlear blood supply interruption[10,11].

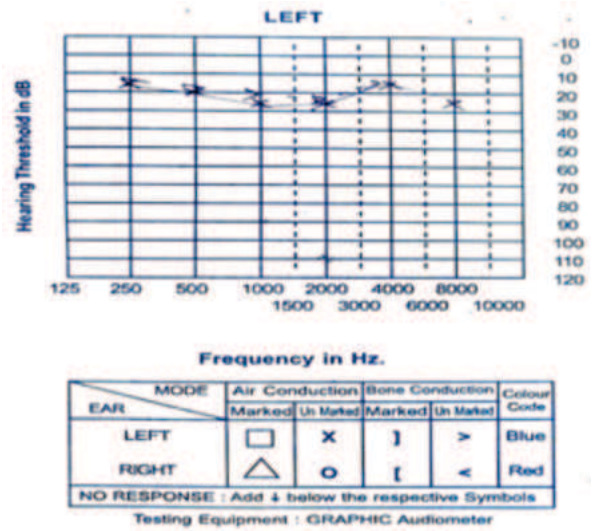

Figure 3: Pure tone audiometry showing Left minimal Hearing loss.

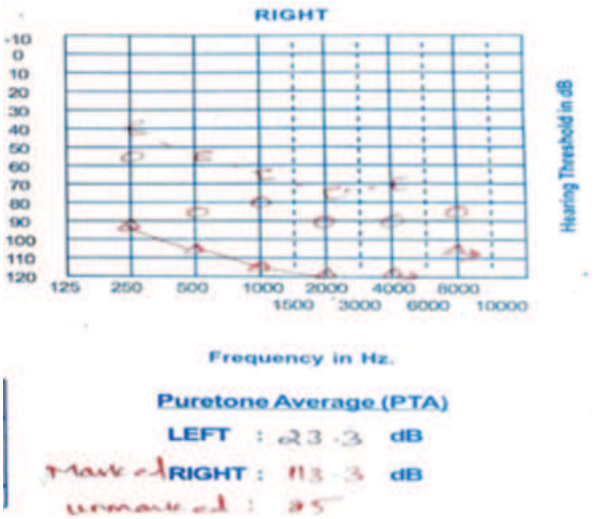

Figure 4: Pure tone audiometry showing Right profound Sensorineural hearing loss.

CPA masses may arise from Cerebellopontine cistern or CPA structures(arachnoid cyst, non-acoustic schwannoma, aneurysm, melanoma), congenital cysts (epidermoid cyst, dermoid cyst, lipoma), tumor invading into CPA(cholesterol granuloma, chondromatous tumors, chordoma, endolymphatic sac tumor, hemangioblastoma,ependymoma, medulloblastoma, pituitary adenoma, glioma, choroid plexus papilloma, lymphoma, dysembryoplastic neuroepithelial tumor, paraganglioma [14].

A review of the literature showed that many patients with arachnoid cyst of the posterior cranial fossa presented with hearing loss as the chief presenting symptom. Jayarao et al. (2009) Presented a case of a young girl with right-sided sensorineural hearing loss and tinnitus secondary to a 12- - mm, non-enhancing, right cerebellopontomedullary cystic lesion that was hyperintense on T2-weighted MRI images. Ottaviani et al. (2002) reported a patient with an arachnoid cyst of the posterior cranial fossa with sensorineural hearing loss and tinnitus.MRI of this 21-year-old patient showed an arachnoid cyst in the left cerebellar convexity that produced pressure on cerebellar hemisphere forward and caused CPA compression.

AC are mostly asymptomatic in the majority of cases and do not grow[1]. Conservative management with serial MRI imaging will identify those cases with gradual cyst enlargement that may need surgical treatment [9]. Surgical indication and technique should be based on the following criteria: (1) type of AC (communicating or noncommunicating cyst), (2) Rate of growth of the cyst, (3) compression or displacement of the adjacent neurovascular structures, and most importantly (4) patient's symptoms and signs, and especially ongoing clinical worsening [13]. Different modes of surgical available are Cyst fenestration, stereotactic puncture, endoscopic cyst fenestration, cystoperitoneal shunt, cyst marsupialization into the subarachnoid space, and complete resection or partial resection of the cyst wall. Aspiration techniques like stereotactic puncture or endoscopic puncture are commonly employed. Insufficient fenestration or Reclosure of the cyst wall resulting in inadequate drainage into the subarachnoid space may lead to recurrence and regrowth. Cystoperitoneal shunting also carries a considerable rate of obstruction and/or recurrence. Endoscopic decompression of the arachnoid cysts of the posterior fossa has been successfully employed and consists of an endoscopic cystocisternostomy. This is a minimally invasive technique that has been reported to be safe as well as effective and, in the case of failure, more aggressive surgery is undertaken. Vestibular symptoms may resolve with Surgical management, but auditory deficits are less likely to recover after surgery[9].

Patients with symptomatic posterior fossa arachnoid cyst may benefit from surgery. Open surgery with radical resection of the entire cyst wall of the AC located in the posterior fossa is associated with a low rate of recurrence and also prevents injury to adjacent neurovascular structures. Suboccipital craniotomy provides a good and safe exposure of the cyst and surrounding neurovascular structures. Some authors recommend the placement of a cystoperitoneal shunt in patients with hydrocephalus[13].

As a high failure rate has been reported with microsurgical procedures and with minimally invasive neuroendoscopic decompression, long-term clinical follow-up is warranted. If the patient develops clinical symptoms and/or otoneurological symptoms the patient should undergo an MRI. In the event of the MRI finding a new cyst, a more aggressive therapeutical option should considered[11].

CONCLUSION

Patients presenting with unilateral sensorineural hearing loss may rarely have an associated arachnoid cyst of the posterior fossa. So a high index of suspicion is needed in especially young patients presenting with hearing loss to identify these types of uncommon CPA masses.

REFERENCES

1. Nisson, P. L., Quintero-Consuegra, M. D., & Lekovic, G. P. (2024). Arachnoid cyst of the Cerebellopontine Angle: a Systematic Literature review. World Neurosurgery, 182, e675–e691. https://doi.org/10.1016/j.wneu.2023.12.018.
2. Gaillard F, Knipe H, Altadill A, et al. Arachnoid cyst. Reference article, Radiopaedia.org (Accessed on 09 Jul 2024) https://doi.org/10.53347/rID-929.
3. Ucar, Tanju & Akyuz, Mahmut & Kazan, Saim & Tuncer, Recai. (2000). Bilateral Cerebellopontine Angle Arachnoid Cysts: Case Report. Neurosurgery. 47. 966-8. 10.1097/00006123-200010000-00034.
4. Helland CA, Wester K A population-based study of intracranial arachnoid cysts: clinical and neuroimaging outcomes following surgical cyst decompression in adults Journal of Neurology, Neurosurgery & Psychiatry 2007;78:1129-1135.
5. Buongiorno, G., & Ricca, G. (2003). Supratentorial arachnoid cyst mimicking a

Ménière's disease attack. The Journal of laryngology and otology, 117(9), 728–730. https://doi.org/10.1258/002221503322334602.
6. Pappas, D. G., & Brackmann, D. E. (1981). Arachnoid cysts of the posterior fossa. Otolaryngology--head and neck surgery : official journal of American Academy of Otolaryngology-Head and Neck Surgery, 89(2), 328–332. https://doi.org/10.1177/019459988108900236.
7. Mustansir F, Bashir S, Darbar A (April 10, 2018) Management of Arachnoid Cysts: A Comprehensive Review . Cureus 10(4): e2458. DOI 10.7759/cureus.2458.
8. Jayarao, M., Devaiah, A. K., & Chin, L. S. (2009). Recovery of sensorineural hearing loss following operative management of a posterior fossa arachnoid cyst. Journal of Neurosurgery. Pediatrics, 4(2), 121–124. https://doi.org/10.3171/2009.3.peds08416.
9. Cadoni, G., Agostino, S., Volante, M., & Scipione, M. S. (2006). Sudden cochlear hearing loss as presenting symptom of arachnoid cyst of the posterior fossa. PubMed, 26(2), 115–117. https://pubmed.ncbi.nlm.nih.gov/16886854
10. O'reilly, R. C., & Hallinan, E. K. (2003). Posterior fossa arachnoid cysts can mimic Meniere's disease. American journal of otolaryngology, 24(6), 420–425. https://doi.org/10.1016/s0196-0709(03)00097-8.
11. Ottaviani, F., Neglia, C. B., Scotti, A., & Capaccio, P. (2002). Arachnoid cyst of the cranial posterior fossa causing sensorineural hearing loss and tinnitus: a case report. European archives of oto-rhino-laryngology : official journal of the European Federation of Oto-Rhino-Laryngological Societies (EUFOS) : affiliated with the German Society for Oto-Rhino-Laryngology - Head and Neck Surgery, 259(6), 306–308. https://doi.org/10.1007/s00405-002-0461-2
12. Thinakara-Rajan, T., Janjua, A., & Srinivasan, V. (2006). Posterior fossa arachnoid cyst presenting with isolated sensorineural hearing loss. The Journal of laryngology and otology, 120(11), 979–982. https://doi.org/10.1017/S0022215106002337
13. Samii, M., Carvalho, G. A., Schuhmann, M. U., & Matthies, C. (1999). Arachnoid cysts of the posterior fossa. Surgical Neurology, 51(4), 376–382. https://doi.org/10.1016/s0090-3019(98)00095-0
14. Bonneville, F., Sarrazin, J., Marsot-Dupuch, K., Iffenecker, C., Cordoliani, Y., Doyon, D., & Bonneville, J. (2001). Unusual lesions of the cerebellopontine angle: a segmental approach. Radiographics, 21(2), 419–438. https://doi.org/10.1148/radiographics.21.2.g01mr13419