

Epidermoid of the Cerebellopontine Angle that Caused Sudden Hearing Loss: Report of a Medically Treated Case

Pontoserebellar Köşenin Ani İşitme Kaybına Neden olan Epidermoidi: İlaçla Tedavi Edilen Bir Olgu Sunumu

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ABSTRACT Epidermoid (cholesteatoma) is a rare mass of the cerebello-pontine angle (CPA). We here with present a case of CPA epidermoid that caused sudden sensorineural hearing loss (SSNHL). A 39-year old woman was presented with the complaints of right hearing loss, tinnitus and dizziness. She had been followed up for CPA epidermoid until the development of hearing loss. Her audiological tests revealed profound sensorineural hearing loss in the right ear, without vestibular findings. Magnetic resonance imaging (MRI) showed a mass of 14x14 mm with irregular borders that filled the right CPA and invaded 7-8th cranial nerves. The mass was interpreted as "epidermoid". Her hearing returned to normal with oral steroid treatment by the 8th day of the treatment. This patient was the second one with SSNHL associated with CPA epidermoid in literature, and the first one successfully treated with steroids. Through this case, we also reviewed CPA masses, and specifically epidermoids with accompanying SSNHL.

Key Words: Cerebellopontine angle; hearing loss, sudden; cholesteatoma; carcinoma, squamous cell

ÖZET Epidermoid (kolesteatom) ponto-serebellar köşe (PSK)'nin kitleleri arasında ender görülenlerdendir. Burada, uzun yıllar önemli bir semptom vermemesine karşın ani sensorinöral işitme kaybı (ASNİK) nedeni olan bir PSK epidermoidi olgusu sunulmaktadır. Otuzdokuz yaşındaki kadın hasta sağ kulağında ani işitme kaybı, tinnitus ve baş dönmesi yakınmaları ile polikliniğimize başvurmuştur. Hastanın takip edilen sağ PSK epidermoid'i öyküsü vardı. Odyometride hastanın sağ kulağında total'e yakın SNIK saptandı, yatırıldığında vestibüler sistem normal bulundu. Manyetik rezonans görüntüleme (MRG)'de PSK'yi tamamen dolduran, 14 mm çapında düzensiz sınırlı, 7 ve 8 nci kafa çiftlerini invaze eden kitle epidermoid olarak yorumlandı. Steroid tedavisi ile hastanın işitmesi 8. günde normal sınırlara geri döndü. Bu hasta, literatürde bildirilen, ani işitme kaybı yapmış 2. ve ilaç tedavi ile işitme kaybı düzelen tek PSK epidermoid'i olgusudur. Bu çalışmada, sunulan olgu aracılığı ile genelde PSK kitleleri, özelde ise epidermoid ile ASNİK ilişkisi gözden geçirilmektedir.

Anahtar Kelimeler: Serebellopontin köşe; işitme kaybı, ani; kolestatom; karsinom, skuamöz hücre

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Cerebello-pontine angle is a subarachnoid space filled with cerebrospinal fluid (CSF), bounded by pons, cerebellum and petrous part of the temporal bone. It contains VIIth, VIIIth and Vth cranial nerves (CNs) in its upper aspect and IXth, Xth, XIth cranial nerves caudally. Various space-occupying lesions found in the CPA have been described of which vestibular schwannomas (VSs) account for 75-80%, followed in frequency by meningiomas and epidermoids (cholesteatomas) with 10-15% and 5-9% respectively^{1,2} Wide variety of rare lesions representing small portion of the CPA masses include paraganglioma, chordoma, chondroma, cholesterol

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granuloma, arachnoid cysts, vascular lesions, neuromas of the other nerves and metastatic tumours.¹

Intracranial epidermoids are believed to originate from foetal ectodermal inclusions and 40-50% of them found in the CPA³ Cerebello-pontine angle lesions are reported to be responsible for around 2% of sudden sensorineural hearing losses (SSNHLs), great majority of which are caused by VSs.³ Herewith, a patient with CPA epidermoid presenting with SSNHL was reported. We have been able to find only one other case with SSNHL and CPA epidermoid in the literature.⁴

CASE REPORT

A thirty-nine-year old female patient was admitted to our department with the complaints of dizziness, tinnitus and suddenly developed hearing loss on the right ear. The patient had a history of twitches in the right eyelid and facial pain on the same side for which her ophthalmologist had referred her to neurosurgery department 13 years ago when her symptoms first emerged. Magnetic resonance imaging (MRI) taken then revealed a mass within the CPA and it was interpreted as “epidermoid”. On the annual follow-up MRI scans, no enlargement of the mass has been detected and the patient has been having occasional and slight facial neuralgia and dizzy spells since then.

Her pure tone audiometric test results were Ac: 93, Bc: 60 dBHL on the right ear; Ac: 12, Bc: 12 dBHL in the left ear (Figure 1). She displayed neither spontaneous nystagmus nor any definitive abnormality in the balance tests. On the MRI scan, there was a heterogeneous and hypodense mass within the CPA at the dimensions of 14x14 mm with serrated borders. The mass appeared to fill the CPA encasing the VII-VIIIth CN complex without any sign of pressure to the pons and cerebellum (Figure 2a). We were not able to compare the latest and earlier MRI scans in terms of the mass's relation with the nerves since the pervious images did not show cranial nerves clearly. However, its size was largely the same. MRI with contrast revealed no enhancement of the mass (Figure 2b). Diffusion weighted image (DWI) showed diffusion

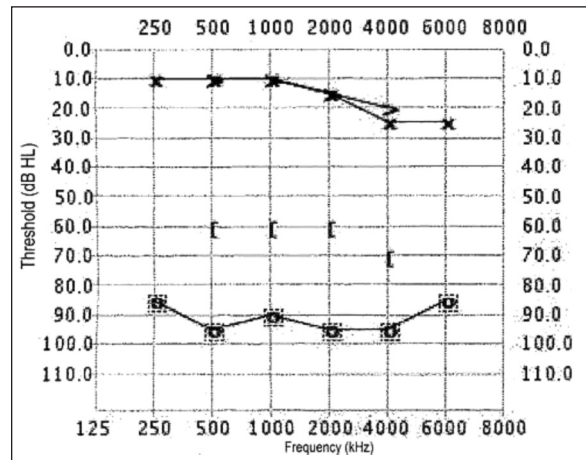


FIGURE 1: Pre-treatment pure-tone audiogram of the patient with CPA epidermoid with SSNHL.

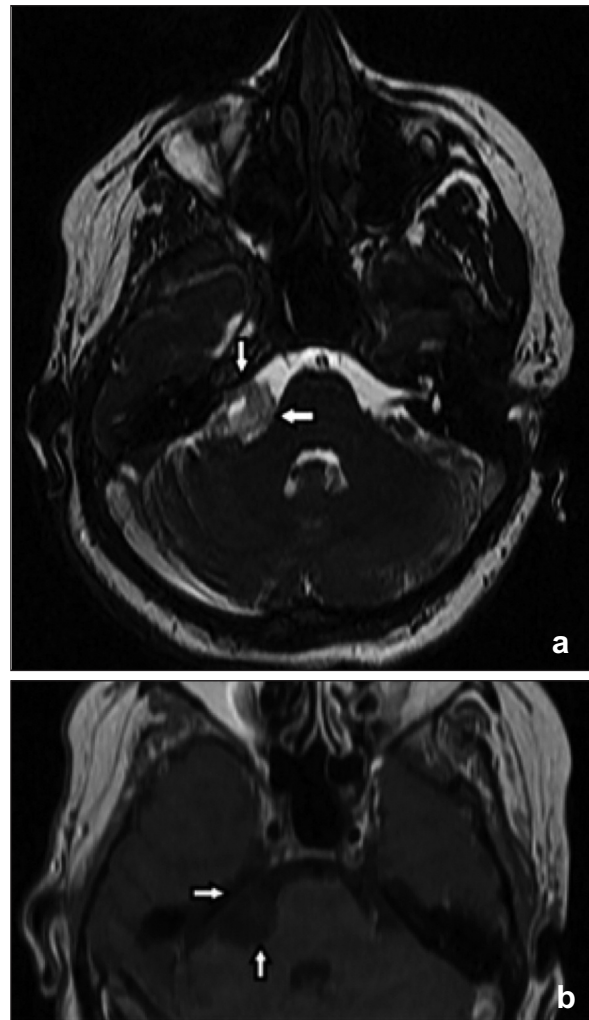


FIGURE 2: a) non-contrasted 3D FIESTA sequence b) with i.v. Gadolinium T1W sequence (Emaray, Biem İlaç San. Tic. A.Ş. Ankara/Turkey) enhanced T1-weighted temporal bone MR images show: a) non-enhancement with contrast material, b) hypochoic irregular PCA mass at the dimensions of 14x14 mm that invades 7-8th nerve complex.

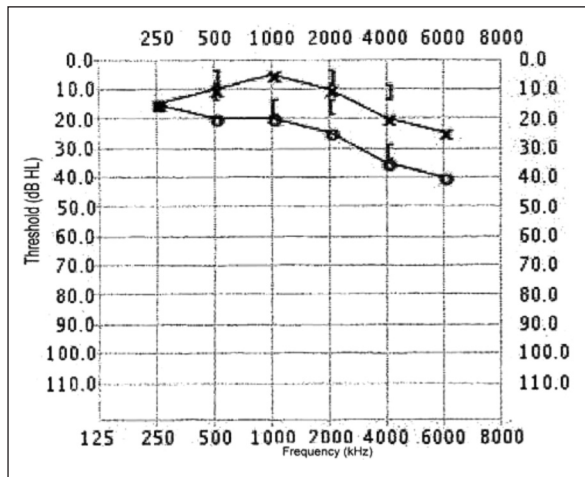


FIGURE 3: Pure tone audiogram of the patient after successful steroid treatment.

restriction in the right CPA as to confirm the diagnosis of epidermoid. The patient responded well to the oral steroid, prednisolone treatment at tapering doses starting with 1mg/kg, with almost complete resolution of the hearing loss (Right Ac: 22, Bc: 13 dBHL) (Figure 3). She opted for non-surgery with regular follow-ups, and 1 year after the occurrence of SSNHL, the hearing level remained stable without additional symptoms and signs.

Written informed consent has been obtained from the patient for the publication of this report.

DISCUSSION

Epidermoids of the CPA are congenital lesions arising from ectodermal squamous epithelial remnants and normally display linear growth pattern. They tend to conform the space they occupy rather than pushing the borders, and surround the nerves crossing the space.^{2,5} They usually give symptoms within the 3rd-5th decades of life, possibly due to their slow growth. Miller et al and Moffat et al reported in their 23 and 15-case series respectively, the average age of presentation as 40.9– 44 years [5,6]. The signs and symptoms are usually related to the involvement of the Vth CN (in 40% of the cases), and VIIIth CN (in 67% of the cases); in the form of hemifacial spasm and trigeminal neuralgia, and tinnitus, progressive sNHL, imbalance-vertigo

respectively. However, sNHL caused by CPA epidermoids are usually milder than seen with the VSs of the same location.^{1,2,6} In our patient, it is very likely that the fasciculation in the eyelids and facial neuralgia she experienced in the past were due to the compression to the Vth CN. Likewise, her dizzy spells could be attributed to the VIIIth CN involvement by the mass. MRI is the imaging of choice for intracranial epidermoids including the ones seen in the CPA, which reveals non-enhancing mass of low intensity with irregular contours. Diffusion weighted images shows diffusion restrictions and relative hyper-intensity comparing to the CSF, which differentiates CPA epidermoids from the arachnoid cysts.⁷ Cerebello-pontine angle epidermoids should also be differentiated from the other non-enhancing CPA masses such as dermoid cysts and lipoma.¹ Treatment of choice for the lesion is complete removal especially for symptomatic patients as it is the case for its middle ear counterpart.⁶ However, surgical removal carries the risks of tumor residua/recurrence and complications such as hearing loss, imbalance, CSF leaks, facial paralysis and meningitis up to 30-40% of the cases.^{5,6} Therefore, potential risks and benefits of the surgery should be weighted carefully on the case bases.

Although the tumour follows a benign course, it tends to recur when the removal is incomplete, and subtotal resection may also lead to malignant transformation.^{5,8} As well as malignant degeneration, intra-lesion haemorrhage would result in enlargement of the mass.⁹ Neither in the previously published case, nor in our patient, these complications seemed to play a role in the resulting SSNHL. In the other published CPA epidermoid case with SSNHL, hearing loss was related to the distortion of the VIIIth CN fibres by the mass, which necessitated surgery.⁴ Normally, involvement of the nerves by the epidermoids has been showed to be in the form of demyelination, inflammatory reactions and later in the course of the pathology nerve ischemia as to potentially cause permanent nerve damage.² Vestibular schwannoma, the most common, and also with the best-studied CPA lesion is reported to cause SSNHL in 13% of the cases. In

some of the CPA vestibular schwannomas with SSNHL patients, either with unknowingly or deliberately given steroid treatment did not yield in stable hearing results and/or complete improvement of the hearing loss.¹⁰

In our patient, excellent response to the steroid treatment suggests inflammation or oedema of the nerve sheath, rather than some morphological alterations. This may be likened to the facial paralysis due to middle ear cholesteatoma, which

is also explained by direct compression and inflammation around the nerve.

This case is important for once more underlining the value of MRI in SSNHL, not only in diagnosing possible cochlear and retrocochlear pathologies, but also for avoiding unnecessary, even potentially harmful treatment modalities such as hyperbaric oxygen treatment. Additionally, it also shows that PCA epidermoids are also capable of causing SSNHL.

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