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Cerebellopontine Angle Cholesteatoma: Case Report and Literaure Review

Serebellopontin Köşe Kolesteatomu: Olgu Sunumu ve Literatür Taraması

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ABSTRACT

Acoustic neuromas are the most common tumors in cerebellopontine angle followed by meningioma, arachnoid cyst, facial neuromas and glomus jugulare tumours. Cholesteatoma is a lesion formed of a mass of stratified keratinising squamous epithelium which is seen usually in the middle ear or mastoid cavity. In this study, we present a 35 years old patient with a complaint of headache caused by a cholesteatoma in cerebellopontine angle. The patient is considered to have a tumor in cerebellopontine angle according to radiologic imaging but the true diagnosis could be made intraoperatively. Clinical presentation, differential diagnosis and treatment modalities are discussed with the literature on the subject.

Keywords
Cerebellopontine angle; skull base; cholesteatoma; diagnosis

ÖZET

Serebellopontin köşede en sık görülen patoloji akustik nörinomlardır. Menenjiom, araknoid kist, fasiyal nörinom ve glomus tümörleri bu bölgede görülebilen diğer lezyonlardır. Kolesteatom, genellikle orta kulak ve mastoidde görülen ve çok katlı yassı epitelin oluşturduğu keratin epitel birikintileridir. Bu olgu sunumunda 35 yaşında baş ağrısı şikayetiyle kliniğimize başvuran ve serebellopontin köşede kolesteatom tanısı konulan bir hasta sunulmuştur. Öncelikle köşe tümörü olarak değerlendirilen hastada doğru tanıya ancak ameliyat sırasında ulaşılabilmiştir. Ayırıcı tanı, klinik bulgular ve tedavi seçenekleri literatür bilgileri eşliğinde tartışılmıştır.

Anehtar Sözcükler
Serebellopontin köşe; kafa tabanı; kolestatom; tanı

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INTRODUCTION

Cerebellopontine angle (CPA) lesions are predominantly skull base tumors affecting the posterior fossa.[1] Studies showed that acoustic neuroma is by far the most common tumor in this region followed by menengioma, cholesteatoma and glomus jugulare tumor.[2] CPA cholesteatoma consists of stratified squamous epithelium surrounded by desquamous keratin developed from the epithelial residuals in this area. These lesions are most probably congenital but growing up to large dimensions without any symptoms make CPA cholesteatomas silent until 2-4. decades unless the patient does not have a cranial imaging study for another reason.[3] The lesions are frequently adjacent to the brainstem and may compress the facial or vestibulocochlear nerve. In such cases the incidence of facial paralysis is much more than schwannomas.[2] Computerized tomography (CT) and magnetic resonance imaging (MRI) are used for diffential diagnosis in CPA lesions.[4] Primary cholesteatomas have a lower dansity than brain tissue in CT. They are hypointense in T1 sections and iso/hyperintense in T2 sections in MRI. The most common confusing lesion in the diffrential diagnosis of CPA cholesteatomas in imaging techniques are arachnoid cysts. Both lesions are in cerebrospinal fluid dansity but arachnoid cysts have a more smooth surface. Because of their non-specific features, audiologic tests have no importance in the differential diagnosis of CPA cholesteatomas.

CPA cholesteatomas are that kind of lesions which are rare and difficult to diagnose and treat. We present a case of CPA cholesteatoma and discuss the clinical presentation, differential diagnosis and treatment modalities with the literature on the subject.

CASE REPORT

A 35 years old patient admitted to our department with headache and dizziness. He had no previous history of any ear complaints. Otoscopic, neurological and vestibular examinations were all normal. A mixed type mild hearing loss was recorded for the left ear in the audiogram. Tympanogram was normal and transient evoked otoacoustic emissions (TEOAEs) were observed bilaterally. In the gadolinium MRI; a mass with 4x3 cm dimensions, hypointense in T1 sections and hyperintense in T2 sections in the left CPA was detected (Figure 1). The lesion was extending to the mastoid bone. We could not be sure whether it was a cholesteatoma or a tumoral lesion. A decision for operation would be given in either situation. After a standart cortical mastoidectomy cholestetoma sac was encountered via translabyrinthine approach so that the diagnosis was made intraoperatively. Cholesteatoma was extending to the mastoid bone, eroding the lateral and superior semicircular canals. The mobility of the ossicules were slightly limited. The sac was scraped from cerebellum and CPA at the dura mater level (Figure 2). There was no defect on dural layer. The cavity was obliterated with fat tissue. External ear canal was closed without cul de sac procedure. The patient was discharged on the third day without any complications. Postoperative audiograms revealed a severe sensorineural hearing loss. The post-

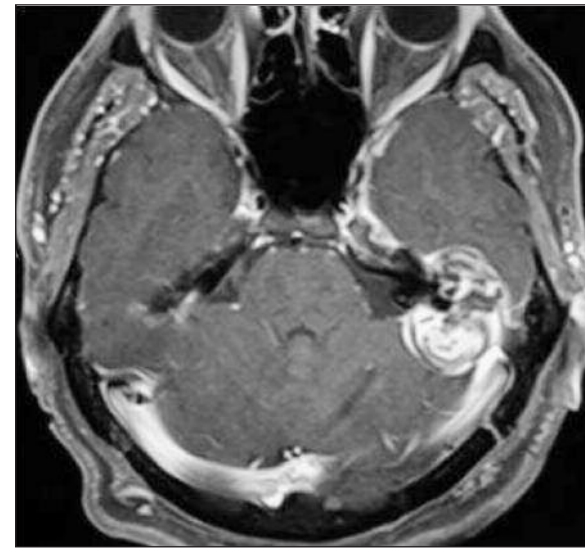

Figure 1. Axial T2 Temporal magnetic resonance imaging.

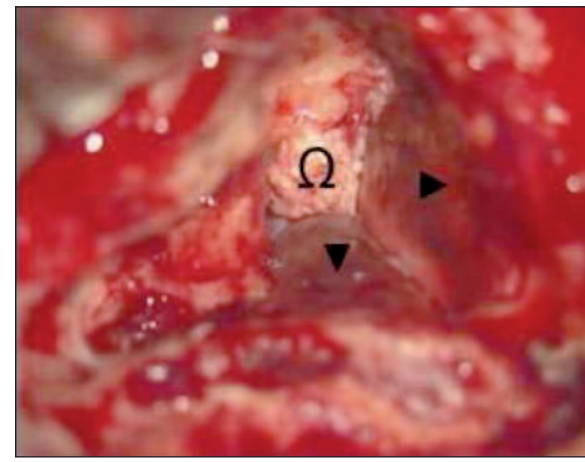

Figure 2. Ω: cholesteatoma; ►: middle fossa dura; ▼: posterior fossa dura.

operative temporal MRI showed no residuel lesion and there was no recurrence in the 3 years follow up period.

DISCUSSION

CPA is a dense area of vital neurological structures. Tumors growing in this region may cause significant dysfunction of cranial nerves and even death if allowed to grow too large. Advances in otolaryngology in the past two decades have changed our viewpoint to CPA lesions. The high quality of MRI and CT have allowed earlier diagnosis of even smaller lesions in this area. According to the literature approximately 80% of CPA tumors are acoustic neuromas.[1] However one must be aware about the unusual tumors in CPA. The most frequent non-acoustic tumors in this region are menengioma, primary cholesteatoma, glomus tumors, facial nerve schwannomas, trigeminal nerve schwannomas, metastatic tumors and giant cell tumors.

Moffat and Ballagh reported that the most common tumor in CPA is acoustic neuroma (80.7%) in their study of 305 patients with CPA tumors. The most frequent tumors in the rest of the patients are menengioma, primary cholesteatoma and Fisch type C glomus jugulare tumors respectively. In the same study facial nerve schwannomas (1%), trigeminal nerve schwannomas (1%), arachnoid cysts (0.7%) and giant cell tumours (0.7%) are mentioned to be the rare tumoral lesions in this area.[2]

Cholesteatomas form up 4.6% of CPA lesions and arise from the inclusion rests of squamous epithelium.[3] They are thought to have congenital origin. These lesions had miscellaneous names in the literature such as primary keratoma or primary epidermoid.[5] They may cause a wide spectrum of symptoms such as hemifacial spasm, progressive facial paralysis, hearing loss or vertigo. The non-specific symptoms made the diagnosis harder sometimes for even experienced clinicians. Beynon et al. reported a case of CPA cholesteatoma presenting with positional vertigo without hearing loss and tinnitus.[6] Cholesteatomas in CPA tend to grow medially, causing earlier compression symptoms to the brainstem than petrous apex cholesteatomas. Although hearing loss is not significant in most of the patients, audiologic evaluation is very important. Quaranta et al. reported that 90% of patients with CPA cholesteatoma had abnormal auditory brainstem response although all of the pure tone audiograms were normal.[7]

Radiological imaging techniques are the best diagnostic tools in making the right diagnosis for CPA lesions. Because of the avascularity CPA cholesteatomas do not have the attitude of the contrast agent. They are seen as hypodense lesions with irregular lobule borders in CT. MRI is the gold standart technique for the diagnosis. Cholesteatomas are hypointense in T1 sections and iso/hyperintense in T2 sections.[8] Arachnoid cyst and cholesterol granuloma should also be considered in the differential diagnosis. Cholesterol granuloma is hyperintense on T1-weighted images and the arachnoid cyst can be distinguished by having a smoother surface. Treatment of CPA cholesteatoma is surgical excision. Translabyrinthine or retrosigmoid approach can be chosen depending on the size, hearing status and localization. Radiological imaging is important for the residual or recurrent lesions in the follow up period.

To conclude differential diagnosis of CPA lesions can be difficult. Meticulous analysing of the radiological studies can be helpful. Treatment is a debate as well. If the clinician decides surgical excision, one must investigate all alternative surgical routes in order to achieve best results.

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