

Primary Petrous Apex Cholesteatoma: A Case Report

Primer Petroz Apeks Kolesteatomu: Olgu Sunumu

ABSTRACT

While primary cholesteatoma is rare, its presentation with only double vision merits extra attention. A patient with primary cholesteatoma of the petrous apex where the only symptom was double vision is presented. Due to extreme medial localization there was absence of the common symptoms of hearing loss and facial paralysis. The patient was operated by the middle fossa intracranial approach and did well after surgery with no additional neurological deficit and no CSF (cerebrospinal fluid) fistulas. Extremely medially located primary petrous apex cholesteatoma are rare, can present with uncommon symptoms and can be successfully treated by the temporal intradural approach.

KEY WORDS: Cholesteatoma, double vision, surgical technique.

ÖZ

Petroz apex Primer kolesteatomu ve sadece çift görme şikayeti olan hasta sunulmuştur. Hasta orta fossa intrakranial yaklaşımı ile opere edildi ve cerrahi sonrası ek nörodefisit ya da beyin omurilik sıvısı fistülü olmadı. Primer petroz apex kolesteatomlarının ekstrem medial yerleşimi nadirdir. Kendilerini nadir semptomlar ile gösterebilir. Temporal intradural yaklaşımı ile başarılı bir şekilde tedavi edilebilir.

ANAHTAR SÖZCÜKLER: Kolesteatoma, çift görme, cerrahi teknik.

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INTRODUCTION

Primary congenital cholesteatoma of the petrous bone is rare. Although it is less common than the acquired lesion itself, it still possesses the same challenging problems in accurate diagnosis and safe total surgical removal. Unlike the acquired lesions, primary cholesteatomas can be found beyond the tympanomastoid space and may occur in various intracranial locations besides the petrous apex (5, 10).

The authors report a case of primary cholesteatoma of the petrous apex and discuss the relevant criteria for their accurate diagnosis and safe surgical removal.

CASE REPORT

A twenty-eight-year-old male patient was admitted to our department with complaint of double vision of ten days duration. He was initially seen by an ophthalmologist for his double vision and was referred to an ENT specialist. His otological examination and ototympanogram were normal and he was referred to our department. Except for a doubtful limitation of lateral gaze in the left eye, his neurological examination was within normal limits.

A cranial MRI study of the skull base showed a mass lesion of 5x10x20 mm at the left anterior petrous apex, medial to the temporal lobe that extended to the internal carotid artery. The lobulated mass was hypointense in T1 and hyperintense in T2 series, and showed no increased signal intensity in postcontrast series (Figure 1A).

A CT scan showed an expansile lesion with smooth margins that did not enhance following administration of IV contrast. There were no signs of chronic mastoiditis or bone destruction at the petrous apex. The pneumatization of the contralateral petrous bone was normal (Figure 1B).

A left temporal craniotomy was performed and a cholesteatoma located in close proximity to the left internal carotid artery was totally removed with an intradural middle fossa approach. The opening of the dura at the base was sutured, reinforced with fascia and tissue glue and the dural opening at the temporal convexity was ultimately sutured in a watertight manner.

The patient had an uneventful recovery. The pathology result came back as cholesteatoma. On his 2nd month control, the neurological examination was within normal limits with no symptoms of rhinorrhea and a control MRI showed no residual lesion (Figure 2A, 2B).

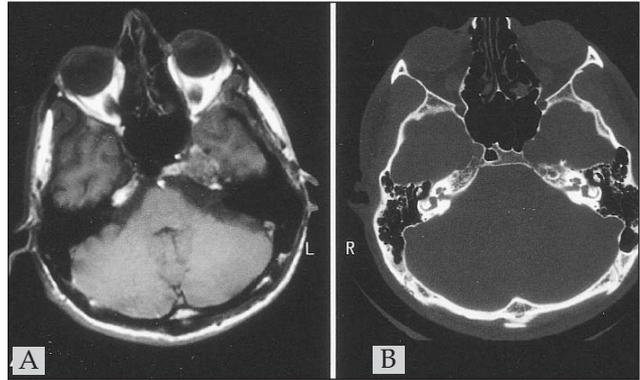


Figure 1 A: Preoperative T1 weighted MR image of the patient.
B: CT images of the patient showing normal pneumatization of the contralateral petrous bone.

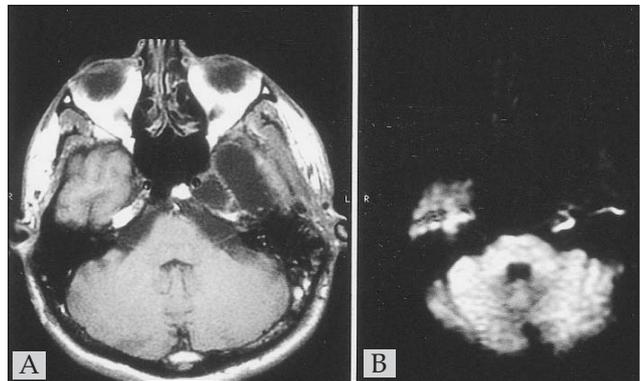


Figure 2 A: Postoperative T1 weighted MR image of the patient.
B: Postoperative diffusion MR images confirming the total excision.

DISCUSSION

The petrous apex is the part of the temporal bone that lies between the inner ear and clivus. It may be divided into two compartments by the internal auditory canal. It is the larger anterior portion, which lies medial to the cochlea, that is most frequently affected by the disease process (14). The horizontal segment of the intrapetrous carotid artery is a surgically important structure that is contained in this segment (4).

Various lesions of the petrous apex are listed in Table I. Cholesteatomas comprise an important group among these pathologies. Cholesteatomas of the petrous apex have two different origins; congenital and acquired. Congenital cholesteatomas are believed to arise from squamous rests (1, 12). Close association of the tympanic ring and internal auditory canal in fetal life may make migration of external canal ectoderm possible (2). In this context they are analogous to intracranial epidermoids and may occur, besides their usual locations of petrous apex, external canal and tympanomastoid region, in numerous other intracranial regions, including the cerebellopontine angle (10).

Acquired cholesteatoma is caused by hypoventilation of the middle ear with resultant retraction of the tympanic membrane. Although epitympanic cholesteatoma is a common disease, deep penetration into the petrous apex is rare. This may occur through a variety of routes, the most common of which are supralabyrinthine (via the anterior epitympanic space) or subcochlear (3). Facial nerve dysfunction is therefore frequent in acquired deep temporal cholesteatomas.

There are two main differences between congenital and acquired petrous apex cholesteatomas. In the acquired form, the squamous lined pocket communicates with the body surface via the external auditory canal and is therefore bacterially colonized and can be frankly infected (18). In contrast, congenital cholesteatomas lie

deeply buried within the temporal bone and are sterile. Secondly, pneumatization of the mastoid is reduced in the acquired form due to the effect of long standing chronic otitis media, while pneumatization of the mastoid is normal in the congenital form.

Clinically primary cholesteatomas present themselves mostly with hearing loss and facial paralysis (3). In some series, 83-94% presented with hearing loss and 20-50% presented with facial paralysis (15, 17). These are the most common symptoms of both the primary and acquired forms and cannot be a criteria for differentiation (17). Congenital cholesteatomas present with more variable and milder symptoms. An absence of infection is an important criterion for primary cholesteatomas besides radiological differences.

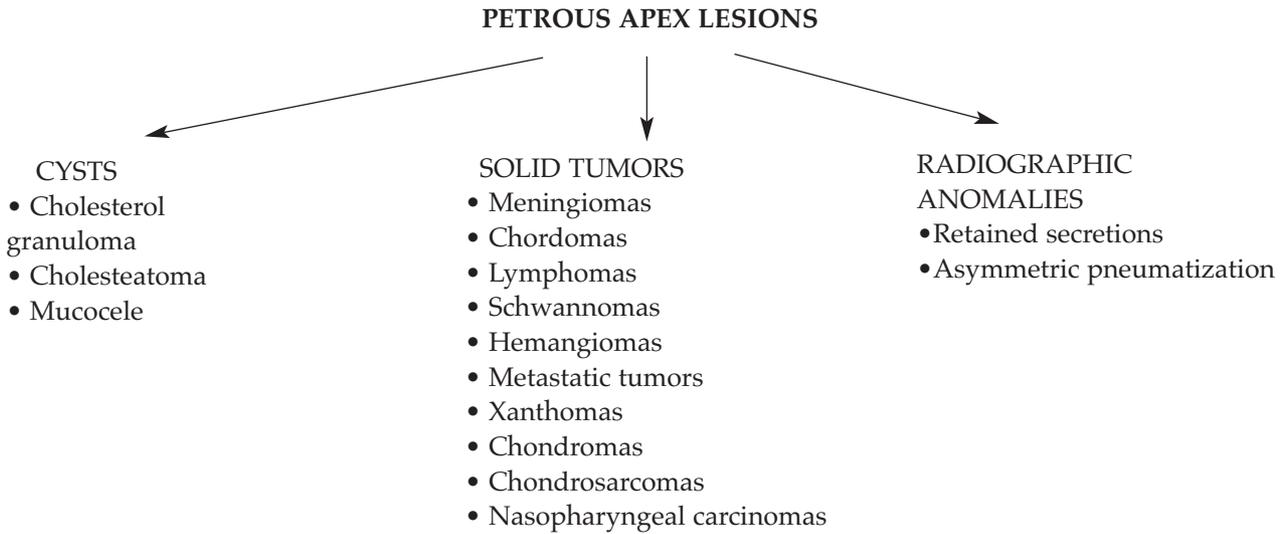
Improved diagnostic imaging modalities had a profound effect on the diagnosis and differential diagnosis of petrous apex lesions (Table II). On CT scan, cholesteatoma of the petrous apex appears as an expansile lesion with smooth margins that does not enhance following iv contrast administration (16). Although variable, it may be hypodense to adjacent brain tissue (8).

Congenital cholesteatoma is generally not distinguishable from a cholesterol granuloma on CT. Because cholesterol granuloma typically occurs in patients with extensively pneumatized petrous apices, presence of an extraordinary degree of pneumatization of the contralateral apex strongly suggests a cholesterol granuloma (18).

Table I: Imaging features of petrous apex lesions.

	CT	Contrast CT	T1, MRI	T2, MRI	Gadolinium
Cholesterol Granuloma	Isodense to brain	Rim enhancing (occasionally)	Hyperintense	Hyperintense	Non-enhancing
Cholesteatoma	Isodense to CSF though variable	Non-enhancing	Hypointense	Hyperintense	Non-enhancing
Mucocele	Isodense to CSF	Non-enhancing	Hypointense	Hyperintense	Rim enhancing Mass is non-enhancing
Asymmetric Pneumatization	Hypodense (Normal septae)	Non-enhancing	Hyperintense	Hypointense	Non-enhancing
Chordoma	Hypodense, calcification	Enhancing	Homogeneous	Heterogeneous	Enhancing
Chondroma/ Chondrosarcoma	Popcorn calcification	Enhancing	Homogeneous	Heterogeneous	Enhancing

Table II: Classification of petrous apex lesions.



On the other hand, acquired cholesteatoma may be readily distinguished from the congenital form and from cholesterol granuloma on CT by signs of bone destruction connecting the middle ear with petrous apex and by signs of chronic mastoiditis.

On MRI, congenital and acquired cholesteatomas are usually of low signal intensity on T1-weighted images, high intensity on T2-weighted images, and do not demonstrate enhancement with gadolinium contrast (11, 21). It is the low signal intensity that usually differentiates cholesteatoma from cholesterol granuloma (20, 23).

The surgical aim should be total excision of the tumor with preservation of existing neuronal function and prevention of CSF leakage. The surgical approaches to be adopted vary according to the presenting symptoms and the extent of the disease.

Though a variety of approaches such as the suboccipital, transsphenoidal and transpalatal-transclival are possible the most suitable approaches for complete cholesteatoma removal are the translabyrinthine-transcochlear and middle fossa approaches and a combination in extensive tumors (6, 7, 13).

The translabyrinthine-transcochlear approach may be more suitable in acquired cholesteatomas with hearing loss and facial nerve dysfunction (9). This approach provides good access to the apex but

impairs hearing and requires facial nerve immobilization. More facial nerve skeletonization means more postoperative facial nerve dysfunction.

The subtemporal middle fossa approach offers a high rate of hearing and facial nerve preservation. It is usually performed by an epidural approach, but is more easily done through an intradural approach. The main reason for an extradural approach is preventing dural tears and CSF leakage with an epidurally located pathology. Dural tears are common with the extradural approach and exposing the medial structures results in more lacerations with more bleeding (19, 22). On the other hand, the intradural approach in primary cholesteatomas, as used in our case, provides a more easier and bloodless route to the more medial structures, and offers a better chance of total excision of the tumor under direct microscopic visualization. Since the primary lesion is sterile, direct opening of the dura at the temporal base provides an excellent approach to the tumor. Packing the cavity with fat and suturing of the dura at the temporal base combined with tissue glue provides an unlikely CSF leakage prevention.

Recurrence of tumor following petrous apex surgery occurs most likely at the medial intrapetrous structures, such as the area around the internal carotid artery or the labyrinthine portion of the facial nerve. Total excision may sometimes not be

possible through an extradural approach. Total excision is more often possible by using an intradural approach, which leads to less frequent tumor recurrence.

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