Petrous apex arachnoid cyst: a case report and review of the literature

Cisti aracnoide dell'apice petroso: descrizione di un caso e revisione della letteratura

V. ACHILLI, G. DANESI[1], L. CAVERNI[2], M. RICHICHI
Department of Otolaryngology,"Fatebenefratelli e Oftalmico" Hospital, Milan; [1] Department of Otolaryngology, Ospedali Riuniti, Bergamo; [2] Department of Diagnostic and Interventional Radiology "S. Paolo" Hospital, Milan, Italy

Key words

Temporal bone • Petrous apex tumors • Arachnoid cyst

Parole chiave

Osso temporale • Tumori dell'apice petroso • Cisti aracnoidea

Summary

Cholesterol granuloma and cholesteatoma are the two most common destructive lesions of the petrous apex. Arachnoid cyst is much less common. These three expansile lesions are often indistinguishable on clinical grounds. Accurate pre-operative radiological diagnosis on computed tomography scan and magnetic resonance imaging is important in order to plan the appropriate treatment. Pre-operative radiological differential diagnosis between primary cholesteatoma of petrous apex and a intrapetrous arachnoid cyst remains a significant problem. The following aspects need evaluation for recognition of intrapetrous arachnoid cysts: 1) an awareness of their existence, 2) homogeneous signal on T1 and T2 weighted images, closely resembling cerebro-spinal fluid signal, 3) special heavily weighted T2 images on magnetic resonance imaging: fluid-attenuated inversion recovery imaging, 4) careful correlation of clinical-radiological data. Symptomatic arachnoid cysts are best treated with conservative drainage surgery through middle cranial fossa. A case of a petrous apex arachnoid cyst is reported which has been radiologically mistaken for a primary cholesteatoma and operated through an infratemporal fossa approach type B. The patient (40-year-old female) came to our attention with right trigeminal pain which had been present for one year and dizziness. Neurotologist and skull-base surgeons should include arachnoid cyst as a rare possibility in the evaluation and treatment of petrous apex cystic lesions.

Riassunto

Il granuloma colesterinico e il colesteatoma sono le due lesioni espansive dell'apice petroso di più frequente riscontro. La cisti aracnoidea intrapetrosa è una patologia molto rara. Questi tre tipi di lesioni sono spesso indistinguibili sul piano clinico. Una precisa e accurata diagnosi differenziale preoperatoria delle lesioni dell'apice petroso mediante TC ad alta risoluzione e RM con Gadolinio non è sempre facile ma è di fondamentale importanza per una corretta pianificazione del trattamento. Particolarmente problematica può essere la diagnosi differenziale fra colesteatoma congenito e cisti aracnoide dell'apice petroso. Per una corretta diagnosi di cisti aracnoide intrapetrosa vanno considerati attentamente alcuni fattori: 1) conoscenza della esistenza di tale patologia, 2) omogeneità del segnale di RM nelle sequenze T1 e T2, pressoché identico al segnale del liquor, 3) valutazione della lesione con speciali sequenze T2 pesate (FLAIR), 4) attenta correlazione dei dati clinici e strumentali. La terapia di elezione delle cisti aracnoidee sintomatiche è secondo molti Autori il semplice drenaggio chirurgico per fossa cranica media, associato a rimozione parziale o totale della capsula aracnoidea. Viene qui riportato un caso di cisti aracnoide dell'apice petroso in una giovane donna di 40 anni presentatasi alla nostra osservazione con una nevralgia trigeminale da circa 1 anno, associata a sensazione di ovattamento auricolare destro e disturbi soggettivi dell'equilibrio da tre mesi. TC e RMI evidenziavano una lesione espansiva dell'apice petroso destro le cui caratteristiche morfologiche e di segnale venivano interpretate come colesteatoma congenito. L'approccio chirurgico per via infratemporale tipo B evidenziava una voluminosa cisti a contenuto liquorale dell'apice della rocca. Nella valutazione delle molteplici lesioni dell'apice petroso deve essere inclusa la cisti aracnoidea per gli importanti risvolti chirurgici e prognostici. L'osservazione di questo caso ha fornito l'occasione per una revisione della letteratura di questa rara patologia.

Introduction

Lesions of the petrous apex represent a challenging diagnostic and therapeutic problem even for the most experienced neurotologists on account of their rare occurrence, aspecifity of symptoms, different treatment options and possible confusion with non-pathologic anatomical variants such as asymmetric pneumatization of the petrous apex. High resolution computed tomography scan (HRCT) and Magnetic

Resonance Imaging (MRI) techniques have revolutionized the diagnosis and management of these lesions. The development of HRCT and MRI has paralleled the development of skull base approaches to the petrous apex. Cholesterol granuloma and primary or acquired cholesteatoma are the two most common destructive lesions occurring in the petrous apex [1] (Table I). Arachnoid cysts (AC) which are much less common, accounting for < 1% of petrous apex lesions, are rarely described in the literature and comprise 1% of all atraumatic intracranial mass lesions [2]. Most AC occur in the middle cranial fossa (50-60%). Other commonly involved sites are the frontal convexity, the quadrigeminal, suprasellar and posterior cranial fossa cysterns [3]. In the cerebello-pontine angle they are frequently associated with vestibular schwannomas. The most accurate description of the development of AC is that of Starkman et al. [4]. Duplication of the arachnoid membranes, as a result of developmental aberration in the flow of cerebrospinal fluid (CSF), may be the primary aetiology. The lining is usually composed of flattened arachnoid cells. Ependymal cells are occasionally found but are not considered to play a role in the pathogenetic mechanism [5]. The most plausible explanation for development of intrapetrous arachnoid cysts seems to be the pulsatile pressure of CSF causing protrusions of the arachnoid granulations through weak areas in the overlying middle cranial fossa dura [6]. Arachnoid cysts associated with petrous apex cephalocele has been described [7]. Intrapetrous AC are rare and pauci-symptomatic lesions. Recent advances in imaging technology have greatly improved both diagnostic efficacy and treatment planning. HRCT and MRI with paramagnetic contrast enhancement (Gadolinium) can reliably distinguish between the various primary and secondary lesions of the petrous apex in the majority of cases (Table II). In the differentiation of petrous apex cystic lesions, the possibility of confusion may occur between congenital cholesteatomas and arachnoid cysts since both are smoothly marginated expansile lesions without contrast enhancement, hypointense on T1-weighted and hyperintense on T2-weighted MRI images [1-6].

The present report deals with personal experience in a rare case of petrous apex arachnoid cyst, highlighting the importance of making an accurate pre-operative diagnosis and discussing the potential for its confusion with the more common cholesteatoma.

Table I. Prevalence of Petrous Apex lesions (66 cases)*.

Lesions	%
Cystic lesions	
Cholesterol granuloma	60
Cholesteatoma	9
Mucocele	3
Solid lesions	
Chondroma	3
Giant-cell tumour	3
Cavernous haemangioma	2
Chondrosarcoma	6
Metastatic carcinoma	2
Dermoid cyst	2
Xanthoma	2
Radiographic anomalies	
Retained secretions	6
Asymmetric pneumatization	3

* Modified from Muckle RP, et al. 1998 [14].

Case report

A 40-year-old female came to our Department with a 1-year history of right trigeminal neuralgia, mid-face paresthesia and sensation of fullness in the right ear which had been present for 1 month. She also had a 3-month history of dizziness. Otoscopy was bilaterally normal. There was no evidence of spontaneous or evoked nystagmus. A mild hypoesthesia of the 1st and 2nd branch of the Vth cranial nerve at the right

Table II. Petrous apex lesions: Characteristics on MRI.

Lesion	T1	T2	T1-Gadolinium
Cholesteatoma	Hypo	Hyper	No enhancement
Cholesterol granuloma	Hyper	Markedly hyper	No enhancement
Arachnoid cyst	Hypo	Hyper	No enhancement
Bone marrow asymmetry	Hyper	Hypo	No enhancement
Internal carotid art. aneurysm	Hypo	Mixed	Rim enhancement *but flow-voids artifacts*
Petrous apicitis	Hypo	Hyper	Rim enhancement
Neoplasm	Hypo	Hyper	Enhancing

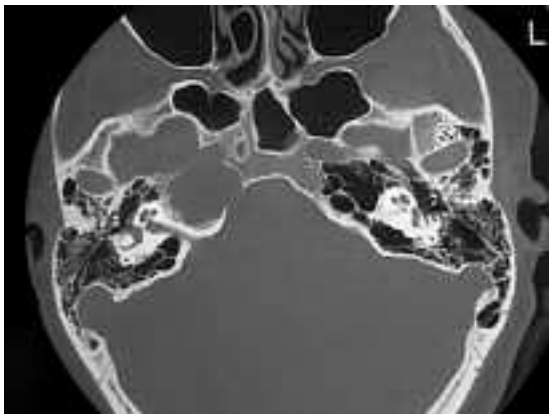

Fig. 1. Axial CT scan with bone details shows hypodense smoothly-marginated lesion of right petrous apex.

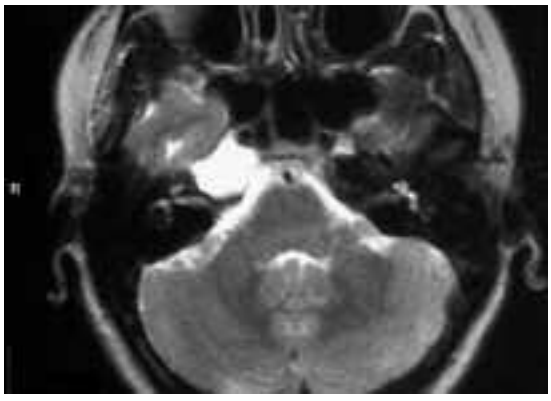

Fig. 2. Axial MRI: on T2-weighted images, mass is hyperintense to brain parenchyma.

side of the face was observed. Pure tone audiometry and speech discrimination were normal in both ears. Brainstem auditory evoked responses (BAER) were normal. Caloric tests showed normal bilateral vestibular function. HRCT scan without contrast enhancement revealed a hypodense lesion at the right petrous apex with smooth bony margins (Fig. 1). MRI with Gadolinium showed a cystic lesion of the right petrous apex, hypointense to brain on T1-weighted spin-echo images, hyperintense on T2-weighted images (Fig. 2), without Gadolinium enhancement except for a very slight rim of enhancement in the peripheral area of the lesion (Fig. 3). Imaging features suggested right congenital petrous apex cholesteatoma. Surgical treatment was, therefore, planned.

An infra-temporal fossa type B approach was performed to gain access to the petrous apex preserving the otic capsule. A subtotal petrosectomy was carried out. The intra-petrous vertical and horizontal carotid artery were identified and skeletonized. Drilling of bone between the cochlear promontory and vertical carotid artery showed a large cavity filled with CSF, instead of cholesteatoma. The cavity occupied the entire petrous apex. Trigeminal ganglion fibres were superiorly displaced by the cystic lesion. No brain herniation into the cavity was observed. The petrous apex cavity seemed to be in communication with the cerebello-pontine cistern through Meckel's cave. The wall of the arachnoid cyst was subtotally removed preserving V3 and the fibres of the gasserian ganglion. The cavity was completely filled with abdominal fat and a blind-sac closure of the external auditory canal was carried out. The post-operative course was uneventful with the trigeminal pain disappearing almost immediately. Total right conductive hearing loss occurred. Facial nerve function was normal. Headache and parestesia on the right side of the face improved, but persisted. CT scan performed on the 3rd post-operative day showed no intra-cranial complication (Fig. 4). The patient has been followed for one year with no recurrence of the cyst being revealed at MRI.

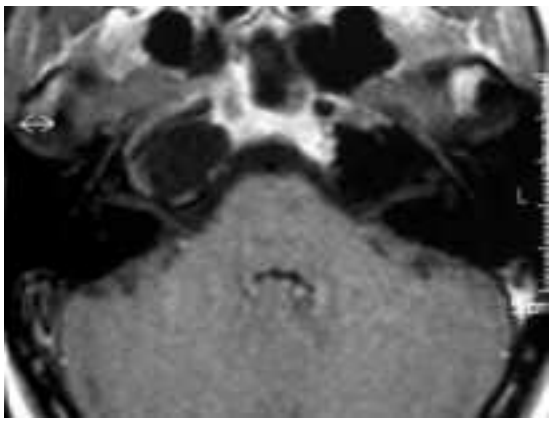

Fig. 3. Axial MRI: on T1-weighted images with GDTPA only a very slight peripheral rim of enhancement is visible (white arrow). Mass does not take up Gadolinium and is hypointense to brain.

Discussion

Arachnoid cysts have been defined as developmental cavities located entirely within the arachnoid membrane, lined with collagen and arachnoid mater cells, containing clear CSF-like fluid and being continuous with the surrounding normal arachnoid mater [8].

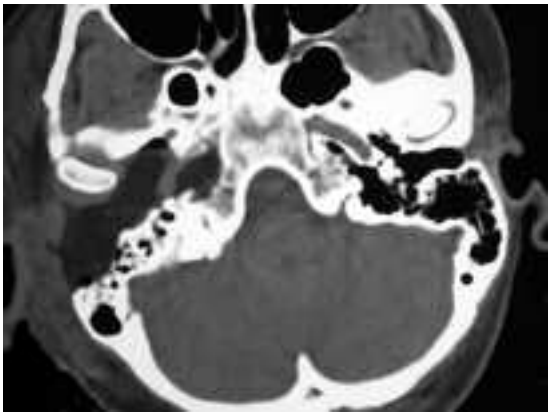

Fig. 4. Post-operative CT scan (day 3) showing extension of right infra-temporal petrosectomy obliterated with abdominal fat.

Much controversy still exists regarding their pathogenesis. Congenital AC are thought to develop from splitting or duplication of the arachnoid membrane into two layers, forming an intra-arachnoid cavity filled with CSF. Two different mechanisms may explain cyst enlargement. The *non-communicating* cyst expands through continued CSF production by the arachnoid membrane. The *communicating* cyst expands probably because of a unidirectional valve mechanism that permits CSF to enter the cavity but not to escape from it [9]. The cyst becomes symptomatic when the enlargement causes compression and dysfunction of neural structures or alterations in CSF dynamics. AC could also form secondary to adhesions and loculations of the subarachnoid space after meningitis, haemorrhage or surgery. AC of the petrous apex are extremely rare lesions. Few anecdotal cases have been described in the literature. In 1967, Hall et al. [10] described a 33-year-old male with spontaneous CSF otorrhea. There was no history of infection or trauma. At surgery a dural defect in the middle cranial fossa was found, through which glial tissue protruded into an anomalous opening in the petrous apex. In 1995, Cheung et al. [10] described two cases of petrous apex AC presenting with two different clinical scenarios: a 46-year-old female with trigeminal neuralgia and a 5-year-old girl with headache, right total hearing loss and CSF leak from the nose. Both underwent surgery with presumptive pre-operative diagnosis of congenital cholesteatoma of the petrous tip. In 2002, Batra et al. [2] described a 55-year-old female with a 6-month history of trigeminal symptoms (right-side toothache associated with lower facial paraesthesia and tingling sensations). A CT scan revealed a well-defined, low density, smoothly marginated expansile lesion in the right petrous apex that was interpreted as a cholesteatoma and approached through a sub-temporal intra-dural route. Intra-operatively a large arachnoid cavity filled with CSF fluid was found in the petrous bone and treated with simple surgical drainage into the adjacent basal cistern.

Pre-operative diagnosis of petrous apex lesions continue to present challenging diagnostic and therapeutic problems. In the differentiation of petrous apex lesions, a possibility for confusion may arise when, at CT scan, a low density, smoothly marginated expansile lesion is encountered that is hypointense on T1-weighted and hyperintense on T2-weighted MRI images, with no Gadolinium enhancement. These findings suggest congenital cholesteatoma (epidermoid tumour) or mucocele. The MR signal and CT scan features of arachnoid cyst and cholesteatoma of the petrous apex are similar but can be differentiated by recently introduced, heavily weighted T2 images: fluid-attenuated inversion recovery (FLAIR) and by Diffusion Weighted Imaging (DWI). FLAIR sequences suppress the completely free water signal, such as the CSF of an arachnoid cyst, whereas a primary cholesteatoma would maintain a high signal intensity [11]. Moreover, the Apparent Diffusion Coefficient (ADC) of DWI does not show restricted diffusion in arachnoid cysts, unlike other similar lesions (congenital cholesteatomas). CSF flow studies using flow-sensitive reversed fast imaging with steady state pulse sequences (PSIF) with electrocardiographic gating have recently been used in differentiating between cysts and enlarged CSF spaces [12]. Pre-operative differential diagnosis between AC and cholesteatoma is of great importance since different surgical strategies are used in the treatment of these two lesions. Symptomatic arachnoid cysts of the petrous apex should be treated by surgical drainage through a middle cranial fossa-trans-petrous approach with partial or complete removal of the arachnoid wall. Cholesteatoma requires more extensive surgery through the infra-temporal fossa, trans-cochlear or trans-otic approach, often with removal of the otic capsule and sacrifice of hearing, and, sometimes, transposition or reconstruction of the facial nerve. The report by Tsuruda et al. [13] on 3 surgical proven cases of petrous apex lesions (two cholesteatomas and 1 arachnoid cyst) and two non-surgically proven lesions (arachnoid and ependymal cyst) demonstrates the possibility to reliably differentiate between cystic lesions of the petrous apex through diffusion-weighted MR images. Arachnoid cyst should be included in the differential diagnosis of the rare and heterogeneous group of petrous apex expansile lesions.

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■ Address for correspondence: Dr. V. Achilli, U.O. ORL, Ospedale Fatebenefratelli-Oftalmico, c.so di Porta Nuova 23, 20100 Milano, Italy - Fax +39 02 43995878 - E-mail: vittorio.achilli@fbf.milano.it