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Congenital Cholesteatoma of Petrous Apex – case report and review of literature

Perlak wrodzony szczytu piramidy kości skroniowej – opis przypadku oraz przegląd literatury

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Article history: Received: 26.02.2017 Accepted: 12.03.2017 Published: 30.03.2017

ABSTRACT:

Objective: Congenital cholesteatomas of the petrous apex are rare lesions that develop insidiously, which delays diagnosis and requires a high index of suspicion. A case report of supralabyrinthine congenital petrous bone cholesteatoma and review of recent literature are presented.

Methods: A 27-year-old woman presented with progressive facial palsy. Otomicroscopy did not reveal any abnormalities, and hearing was normal. Although there were no complaints of vertigo or instability, vestibular examinations indicated a non-compensated peripheral vestibular lesion. Radiological examinations revealed a lesion in the petrous apex and epitympanum that had features of a cholesteatoma.

Results: To preserve normal hearing, the middle fossa approach was chosen for surgery. A damaged part of the facial nerve was reconstructed with cable graft.

Conclusion: Advances in radiological imaging facilitate surgical planning, and improvements in the techniques of lateral skull base surgery enable safe and radical removal of petrous bone cholesteatomas with minimal morbidity

KEYWORDS: congenital petrosal cholesteatoma, facial nerve palsy, facial nerve reconstruction, middle fossa approach

STRESZCZENIE:

Wstęp: Wrodzony perlak szczytu piramidy kości skroniowej jest rzadkim schorzeniem, które rozwija się skrycie, tak więc jego rozpoznanie wymaga dużej dociekliwości. Przedstawiamy opis przypadku wrodzonego perlaka nadbłędnikowego szczytu piramidy kości skroniowej oraz przegląd literatury.

Metodyka: Pacjentka, lat 27, zgłosiła się z powodu postępującego niedowładu nerwu twarzowego. W badaniu otoskopowym stwierdzono normalną błonę bębenkową, słuch był również prawidłowy. Pomimo braku zawrotów głowy i zaburzeń równowagi w wywiadzie, badania wykazały niewyrównane obwodowe uszkodzenie układu przedsionkowego. Badania radiologiczne uwidoczniły zmianę o charakterze perlaka w szczycie piramidy kości skroniowej i epitympanum.

Wyniki: W celu zachowania słuchu do usunięcia perlaka wybrano dojście przez środkowy dół czaszki. Zniszczony fragment nerwu twarzowego został zrekonstruowany autologicznym przeszczepem nerwu usznego wielkiego.

Wnioski: Nowoczesne techniki obrazowania umożliwiają szczegółowe zaplanowanie zabiegu, jak również rozwój chirurgii podstawy czaszki, a także pozwalają na bezpieczne i radykalne usunięcie perlaków szczytu piramidy.

SŁOWA KLUCZOWE: wrodzony perlak szczytu piramidy, porażenie nerwu twarzowego, rekonstrukcja nerwu twarzowego, dostęp przez środkowy dół czaszki

 DOI: 10.5604/01.3001.0009.7991

INTRODUCTION

Petrous apex cholesteatomas, localized medial to the otic capsule, are rare, comprising 4% to 9% of all petrous apex lesions [7,11,14]. They account for only 3% of all cholesteatomas [20]. Classically, they are classified as acquired or congenital, but the etiology may be difficult to establish in advanced cases. The majority of them are acquired, originate from diseases in the middle ear and mastoid, and erode medially around or through the labyrinth. Congenital cholesteatoma, often referred to as epidermoid, is suggested to arise from retained epithelial cell rests – epidermoid formation [12,15]. Cannoni et al. [3] and Fish [5] share an opinion that intra-petrosal congenital cholesteatomas always originate at the level of the ganglion of the facial nerve. Omran et al. [17], in a case series, reported that only 15% of petrous apex cholesteatomas were considered congenital. The majority of congenital cholesteatomas arise in the upper-anterior part of the middle ear behind an intact tympanic membrane [10]. Among 63 cases of congenital cholesteatomas presented by Kojima et al. [9], only two were localized in the petrous apex. These data confirm that congenital cholesteatomas of the petrous apex are uncommon.

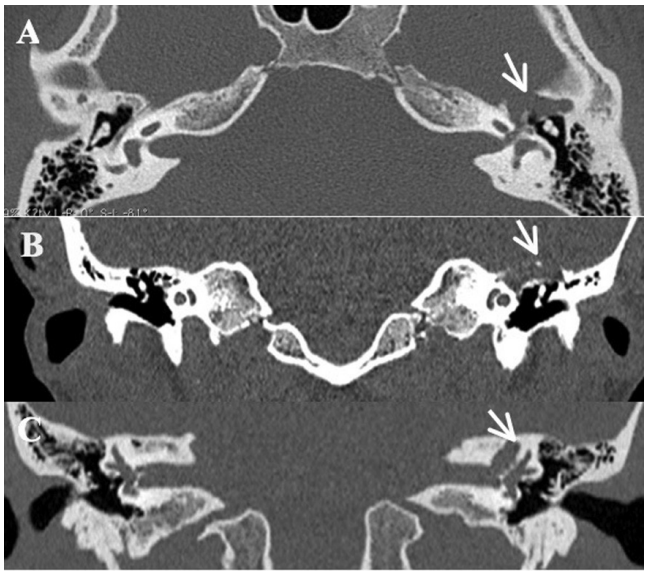

Fig. 1. A – axial, B and C - coronal CT scans demonstrate a soft tissue mass in the left epitympanum above the head of the malleus, extending over the cochlea, destroying tegmen tympani and the upper wall of the internal acoustic meatus.

The most common presenting symptom of petrous apex cholesteatomas is hearing loss, which is observed in up to 96% of cases [1,2,13,16,17]. The second most common symptom is facial palsy, occurring in more than half of patients [2,13,16,17, 22]. Other symptoms like dizziness, instability, or trigeminal neuralgia occur less frequently [1,2,16,17]. Discharge from the ear is observed typically in acquired cholesteatomas [1,2,16,17]. Notably, as congenital cholesteatomas are slow-growing lesions that usually lack inflammatory reaction, they may remain asymptomatic for a long period of time [18,19].

Since symptoms are not specific, the diagnosis and the extent of the disease have to be established based on radiographic studies. The development of modern imaging techniques has created new possibilities in differentiating the lesions of the temporal bone. The radiographic characteristics of some petrous apex lesions according to Isaacson et al. [7,8] are presented in the Table I. CT (computed tomography) enables a detailed evaluation of osseous structures and patterns of bone erosion, whereas MRI (magnetic resonance imaging) helps to significantly narrow down the differential diagnosis of petrous apex lesions [7,8]. Cholesteatomas appear dark on T1- and bright on T2-weighted images, and do not enhance with intravenous contrast. Diffusion weighted images (DWI) show a markedly increased signal intensity, consistent with reduced diffusion [4]. However, there are some limitations with standard clinical DWI sequences, such as low resolution and production of relatively thick image sections [6]. Newer techniques of DWI, with thinner section acquisition and decreased susceptibility to artifacts, allow detection of even small lesions [23].

Sanna et al. [20,22] introduced an anatomic classification system of petrous bone cholesteatomas that includes five categories: supralabyrinthine, infralabyrinthine, massive labyrinthine, infralabyrinthine-apical, and apical. A few years later, Moffat and Smith [16] added two more categories - supralabyrinthine apical and massive labyrinthine-apical. Sanna's classification is, however, most commonly used in the literature. There are few large case series of patients with petrous bone cholesteatomas [1,13,16,17, 22]. The rate of incidence of various categories of apical cholesteatomas is presented in Table II.

Complete surgical removal remains the treatment of choice for petrous apex cholesteatomas. However, this surgery is challenging and requires detailed planning to maximally reduce morbidity. Several surgical approaches to manage petrous apex cholesteatomas have been described - transcochlear, translabyrinthine, transtemporal supralabyrinthine, infracochlear, subtotal or radical petrosectomy, retrolabyrinthine, middle cranial fossa, and transsphenoidal [1,7,16,20,22] . The choice of surgical approach depends on the extent and location of the lesion, hearing impairment, facial nerve function, and involvement of vital structures such as the sigmoid sinus or carotid artery [7,16].

CASE REPORT

A 27-year old woman presented with a 9-month history of progressive left facial paresis. There were no complaints of hearing loss or vertigo. The patient denied any symptoms of otitis media. Initially, a diagnosis of Bell's palsy was made, and the patient was treated with steroid therapy and facial rehabilitation but without improvement. As the function of the facial nerve became progressively worse, CT and MRI of the skull base were performed and revealed a lesion in the left petrous bone. The patient was referred to our Department with a presumptive diagnosis of petrous bone cholesteatoma.

On admission, left facial nerve palsy was observed and graded as IV/V according to the House-Brackmann scale. Otomicroscopy revealed intact tympanic membranes in both ears. Pure tone audiogram and speech audiometry showed normal hearing in both ears. ABR did not show any abnormality.

Videonystagmography (VNG) indicated a non-compensated peripheral vestibular lesion. In VNG, spontaneous nystagmus with a dominant vertical component and positional nystagmus directed to the right were observed. Saccade and oculomotor tests were normal. There was unilateral defective tracking (R>L). Weakness of the left ear in the caloric test was observed. Sensory organization test (SOT) showed signs of left vestibular dysfunction.

Computed tomography of the temporal bone revealed a soft tissue lesion within the left petrous apex of the temporal bone, extending over the cochlea to the left epitympanum above the head of the malleus (Fig.1). Imaging showed bone erosion with smooth margins within the petrous apex, eroded wall of the internal auditory canal, tegmen tympani, and above the geniculate ganglion. Subsequent MRI confirmed the presence of the soft tissue mass which was hypointense on T1- and hyperintense on T2-weighted images and extended along the pyramid of the temporal bone (Fig.2). Diffusion-weighted imaging revealed a hyperintense lesion in that area.

The patient underwent surgery; due to localization of the lesion and preserved normal hearing in the affected ear, the middle fossa approach was chosen. Once the skin and subcutaneous tissues were dissected, the temporalis muscle was sectioned, elevated and secured with retractors. The zygoma and the osseous external auditory canal provided landmarks for craniotomy bone flap. A 3x3cm craniotomy bone flap (centered 1.5 cm anteriorly to the root of the zygoma) was raised up. The dura mater was elevated along the floor of the middle fossa. Cholesteatoma matrix was shown immediately after elevation of the dura. The greater superficial petrosal nerve, arcuate eminence, petrous ridge, and middle meningeal artery were localized. In the region of the arcuate eminence, the superior semicircular canal was 'blue-lined'. Tegmen tympani was destroyed by the cholesteatoma, which extended from the anterior epitympanum medially towards the internal auditory canal along the facial nerve. The upper wall of the internal auditory meatus was destroyed. There was no ossicular chain, bony labyrinth, or carotid artery involvement. The cholesteatoma was removed completely. The facial nerve was almost completely destroyed proximal to the geniculate ganglion. The damaged part was resected and managed with cable graft using the great auricular nerve.

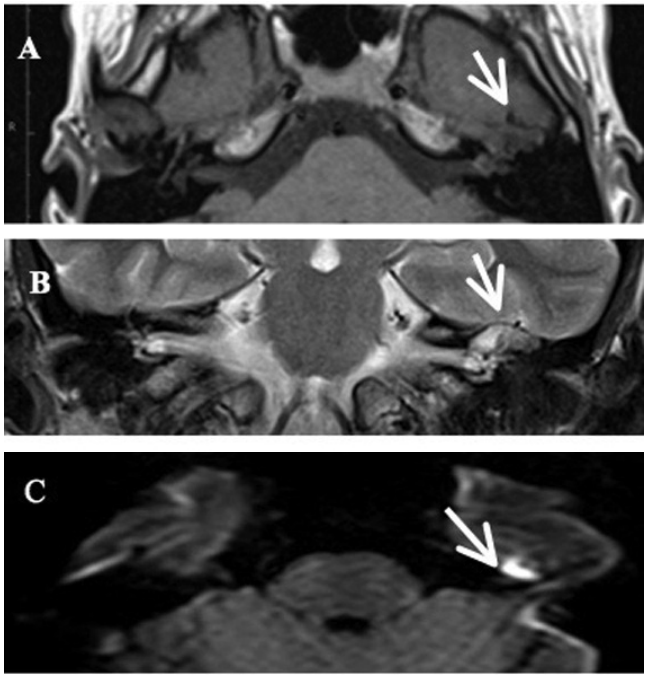

Fig. 2. MRI. A. T1-weighted images. B. T2-weighted images. C. DWI. The soft tissue mass was hypointense on T1- and hyperintense on T2-weighted images and extended along the pyramid of the temporal bone. Diffusion-weighted imaging revealed a hyperintense lesion in that area.

The postoperative period was uneventful. No instability or vertigo were observed after surgery. Hearing was preserved.

DISCUSSION

Since there was no connection between the cholesteatoma and the intact tympanic membrane, and the patient denied any previous history of middle ear inflammation or trauma, we consider this cholesteatoma to be congenital. Localization of the lesion is consistent with supralabyrinthine cholesteatoma, according to Sanna's classification and supralabyrinthine-api-

Tab. I. Differential diagnosis and radiographic characteristics of selected petrous apex lesions. Adapted from Isaacson B. et al. [8].

LESION	MRI			CT	OTHER
	T1 GAD(-)	T1 GAD(+)	T2		
Cholesteatoma	Hypointense	No enhancement	Hyperintense	Smooth erosion	Restricted diffusion on DWI
Cholesterol granuloma	Hyperintense	No enhancement	Hyperintense	Smooth erosion	
Petrous apicitis	Hypointense	Rim enhancement	Hyperintense	Destroyed septae	No restricted diffusion on DWI
Effusion	Iso- to hypointense	Slight enhancement	Hyperintense	Intact septation	Hyperintense on FLAIR
Schwannoma	Isointense	Enhancement	Hyper- or hypointense	May show dilatation of IAC	Centered over porus acusticus

Tab. II. Petrous bone cholesteatoma – large case series

AUTHOR	NO. OF CASES	SUPRA LABYRINTHINE	MASSIVE LABYRINTHINE	INFRA LABYRINTHINE	INFRA LABYRINTHINE APICAL	APICAL
Sanna 1993	54	24 (45%)	13 (24%)	12 (22%)	5 (9%)	0
Omran 2006	93	41 (44%)	42 (45%)	7 (8%)	3 (3%)	0
Magliulo 2007	52	19 (36%)	12 (23%)	15 (29%)	4 (8%)	2 (4%)
Moffat 2008	43	21 (21%)	27 (63%)	1 (2%)	3 (7%)	3 (7%)
Aubry 2010	28	2 (7%)	10 (36%)	6 (21%)	4 (14%)	6 (21%)

Tab. III. Surgical approaches for supralabyrinthine cholesteatomas - review of literature.

SURGICAL APPROACH	SANNA 1993	OMRAN 2006	MAGLIULO 2007	MOFFAT 2008	AUBRY 2010	TOTAL
Middle cranial fossa	10	1			2	13
Modified transcochlear	9	8				17
Enlarged translabyrinthine		15				15
Transtemporal supralabyrinthine			5	9 (1+MF)		14
Transotic		8	3			11
Subtotal petrosectomy		7	11			18
Radical petromastoidectomy	5	2				7
Total	24	41	19	9	2	

cal, according to Moffat's classification. Sanna et al. [20,22] describe supralabyrinthine cholesteatomas as characteristically congenital or resulting from deep ingrowth of an acquired epitympanic cholesteatoma. It is centered in the region of the geniculate ganglion, involving the anterior epitympanum and extending medially towards the internal auditory canal, like in the presented case [17,20,22]. Supralabyrinthine cholesteatomas comprise between 7% and 45% of all petrous bone cholesteatomas (Table II). In the petrous apex, cholesteatoma commonly cause facial nerve palsy, which is observed in 58% of cases [21]. If preoperative facial palsy is present, the nerve may be compressed but anatomically intact, substituted by fibrous tissue, or completely interrupted [17,21]. In the first instance, decompression is the treatment of choice [11,17,21,22]. In the latter two cases, nerve re-routing and end-to-end anastomosis or cable graft anastomosis of the proximal portion of the nerve is still available and is recommended; this was done in the presented case [2,7,13,17,21,22]. This recommendation comes from the opinions that preoperative facial nerve palsy carries a poor prognosis. Grade III or greater palsies rarely recover after simple decompression and should be managed more aggressively. In contrast, when preoperative facial function is normal, the prognosis for its maintenance is nowadays excellent [1,2,13]. An early diagnosis due to improved imaging allows removal of cholesteatomas at earlier stages, which is the best way to maintain optimal facial function [13].

Facial palsy was the only symptom in our patient. Otoscopic examination revealed a normal tympanic membrane. Lack of otoscopic abnormalities was observed in 13 out of 52 patients with petrous bone cholesteatomas described by Magliulo [13]. The patient had no complaints of vertigo or imbalance in spite of a non-compensated peripheral vestibular lesion diagnosed on VNG, possibly because of a slow growth of the lesion and simultaneous compensation. The abnormalities in VNG and SOT were most likely caused by compression of the superior vestibular nerve by the cholesteatoma that invaded the internal acoustic meatus.

In the presented case, the diagnosis was made on the basis of radiological examinations. CT showed bone erosion with smooth margins, suggesting cholesteatoma, and MRI enabled more precise diagnosis by showing features of a cholesteatoma.

Various surgical approaches are used for removal of supralabyrinthine cholesteatomas, depending on etiology, size of cholesteatoma, and hearing status (Table III). In the past years, petrous bone cholesteatomas were managed with permanent fistulization through trans-mastoid and trans-labyrinthine approaches, which resulted in a very large, poorly drainable postoperative cavity [20]. The evolution of the concept of obliteration techniques solved some of the problems of open cavities but did not allow for hearing preservation. The middle fossa approach gives the possibility to remove the lesion and maintain the function of middle and inner ear. However, this approach has limitations and, according to Sanna's [22] suggestion, should be used only for small supralabyrinthine cholesteatomas without posterior and anterior extensions. Since there was no posterior extension of the cholesteatoma, the middle fossa approach permitted total removal of the cholesteatoma with preservation of hearing.

CONCLUSION

The advances in radiological imaging facilitate surgical planning, and improvements in the techniques of lateral skull base surgery enable safe and radical removal of petrous bone cholesteatomas with minimal morbidity.

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Word count: 1700 Tables: 3 Figures: 2 References: 23

Access the article online: DOI: 10.5604/01.3001.0009.7991

Table of content: http://otorhinolaryngologypl.com/resources/html/articlesList?issueId=9790

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Competing interests: The authors declare that they have no competing interests.

Cite this article as: Imie X.,: Congenital Cholesteatoma of Petrous Apex – case report and review of literature; Pol Otorhino Rev 2017; 6(1): 62-67