

Meningocele of petrous apex: A case report, literature review of differential diagnosis and its radiological findings

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ABSTRACT

Meningoceles consist of meninges and CSF that have herniated through a skull defect, usually located in the midline. They are seen more commonly in children and very rarely in adults. We report a case of a 65-year-old patient,

who was diagnosed with an osteolytic lesion in the right petrous apex, after computed tomography of the head was performed. A magnetic resonance imaging scan of the brain showed a cystic CSF collection through the defect. Below we present our experience of a meningocele of petrous apex, and by the way we provide a brief review of its differential diagnosis, as well as its relevant imaging findings.

Key Words: Meningocele; Petrous apex lesions; Differential diagnosis; Imaging findings

INTRODUCTION

Meningoceles are protrusions of the meninges through points of weakness, either in spinal cord or intracranial. They are usually connected with NTDs (neural tube defects) and spina bifida in children. Intracranial meningoceles on the other side seem to be rare lesions, which depending on their size and location could provoke various clinical manifestations. Mainly they present with CSF leak and meningitis, headaches or more rarely seizures. Their differential diagnosis includes a wide variety of lesions, and on that purpose radiological findings seem to play the essential role. Below we present our case report and we provide a brief review of the corresponding differential diagnosis and its imaging findings.

CASE REPORT

A 65-year-old female patient presented in our clinics after a cranial CT-scan, which showed an osteolytic lesion located in the right petrous apex. The patient was complaining about repeated falls during the previous few months. Furthermore she had been suffering from occasional headaches during the previous 15 years (1). Intact neurological status. The cranial CT-scan showed a lytic lesion in the skull, as an extensive nonaggressive erosion of right petrous apex. Given these findings, we performed an MRI examination of the brain, which showed a lesion with high signal intensity on T2-weighted magnetic resonance imaging (MRI), which is in contact with Meckel's cave. A radiologic diagnosis of a meningocele was made. Furthermore an adjuvant to the ICA (internal carotid artery) was identified (2-5). The patient underwent a transnasal / transphenoidal operation. The meningocele was reduced and the defect was plugged with an autologous abdominal fat graft and fibrin glue. Transnasal-transphenoidal approach is a safe, effective, minimally invasive and well tolerated approach for these lesions, allowing to resect the herniated tissue and repair the dural and bone defect (6,7). Postoperative imaging revealed fat filling the petrous apex up to the osseous defect and mild pneumatocephalus. A small amount of tissue was sent to pathology for intraoperative diagnosis (8). The histological diagnosis was bone and connective tissue; no adjuvant therapy was administered.

DISCUSSION

Meningoceles are classified in this group of malformations of craniofacial skeleton, which simply represent defects in scalp, bone, meninges or brain (9-11). They consist a subtype of encephaloceles, as their calvarian defect is associated only with herniation of leptomeninges and cerebrospinal fluid (CSF). In literature series, its shown variable incidences of different types of cephaloceles among patients from different geographic locations,

race and sex, as well as different associations with neural tube defects. Meningoceles are most commonly congenital in origin and have an estimated incidence of one in every 35,000 live births. Spontaneous meningoceles or meningoencephaloceles account for only 8.6% of all meningoencephaloceles (12). The relative incidences of cranial meningoceles versus meningoencephaloceles vary widely, partly as a function of the type of cephalocele. In literature series, Simpson (13,14) found that 24% of reported cephaloceles were cranial meningoceles and 76% meningoencephaloceles.

Etiology

Meningoceles and therefore encephaloceles can be divided according to etiology into:

1. Congenital (the most common)
2. Secondary (traumatic, postsurgical, neoplastic, inflammatory- history of chronic otitis media, or even with idiopathic intracranial hypertension syndrome)
3. Spontaneous (rare)

They may present either as a single isolated congenital anomaly or smaller number of cephaloceles as variable parts of known syndromes (1). Encephaloceles-meningoceles are most commonly associated with Meckel's syndrome and amniotic band syndrome (11). Congenital encephaloceles-meningoceles are divided into three major types: cranial vault, frontoethmoidal, and basal. Secondary encephaloceles-meningoceles are acquired lesions that are most commonly the result of trauma or postsurgical defects. According Bialer et al. meningoceles are significantly more common with patients with idiopathic intracranial hypertension (IIH) (12).

Clinical signs

The petrous apex is a 4-sided-pyramid with its apex anteromedial and its base posterolateral. It is wedged in at the base of skull, anteromedial to the inner ear, between the the posterior border of the great wing of the sphenoid bone and the basilar part of occipital bone. The lesions of petrous apex can affect the surrounding anatomical structures (such as ICA, trigeminal ganglion, sixth cranial nerve, jugular bulb, inferior and superior petrosal sinuses). That's the reason why these lesions can be presented with various symptoms and clinical signs, which are dependent on the nature of pathologic process, size and location. Muckle and colleagues (3) reported that the most common presenting symptoms were hearing loss, dizziness, headaches, and tinnitus. Decreased cranial nerve V function was present in 22%. Other symptoms can

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be sixth cranial nerve palsy and diplopia. Symptoms often present months or years before diagnosis, and incidental discovery is not uncommon.

Diagnosis

Diagnosis and management of petrous apex lesion present unique challenges secondary to their centralized location and their anatomical adjacencies. Imaging studies (CT-Scan, MRI) are the primary method used to diagnose these lesions. Subsequently we report a differential diagnosis of petrous apex lesions, as well as their

characteristics on imaging findings.

Differential diagnosis

Generally petrous apex lesions can be divided into 4 categories:

1. Inflammatory / Cystic
2. Cerebrospinal fluid / Cephaloceles
3. Neoplasms (such as Chordosarcoma, Chordoma, meningioma, metastasis)
4. Other rare lesions (Schwannomas, Paragangliomas)

More specially and regarding cephaloceles-meningoceles we have to take into consideration the non-neoplastic lesions. They are representing an amount of more than 90% of all petrous apex lesions. Depending on pathophysiology, the majority of nonneoplastic petrous apex lesions are inflammatory complications of air cell disease. Pneumatization of petrous apex occurs when epithelium-lined air cells develop as medial communication from the mastoid air cells. The air cells of the petrous apex are susceptible to similar pathologic processes that occur in the mastoid segment including obstruction, opacification, inflammation, and infection.

We report a review of differential diagnosis of these lesions (Table 1).

TABLE 1

Differential diagnosis of these lesions

Petrous apex lesion Inflammatory/Cystic	Characteristics/Pathophysiology
Cholesterin granulom/ Xanthogranulom	Cholesterin crystal, Foam cells. Anaerobic catabolism of blood and blood products.
Mucocele	Obstructed mucosalized air space.
Cholosteatoma/Epidermoid	Epithelial cell remnants/multipotential embryonic cells.
Effusion or retained	Infection of middle ear or mastoid -> trapping of residual fluid in the petrous apex.
Cerebrospinal fluid cysts and cephaloceles	
Meningocele/CSF cyst/ arachnoid cyst	Points of weakness, splitting of meninges, skull base.
Cephaloceles/Meckel's cave diverticulum	Protrusion of arachnoid or dura from Meckel's cave.
Abszess/petrous apicis	Spread of infection through preformed air-cell tracts in temporal bone/usual otitis media.
Aneurysma	Petrous carotis interna aneurysma/ posttraumatic, congenital, mycotic.
Asymmetric pneumatization	Asymmetrie of air cells (5-10 % of all individuals). 10-30 % air cells in temporal bones.

Imaging findings

A low-dose non-contrast CT scan (80 mA & 120 kV) was performed with a Toshiba Aquilion CX CT helical scanner with wide window width (2700 HU) and level (400 HU) using a bone filter kernel. MR imaging was performed on a Siemens Skyra 3-Tesla MR unit with standard T1-weighted (spin echo; TR 600 ms/TE 20 ms; 5 mm slice thickness), T2-weighted 3d (spin echo; TR 1400 ms/ TE 152 ms; 0,8 mm slice thickness) and T2-weighted inversion recovery (spin-echo; TR 9000 ms/ TE 81 ms; 5 mm slice thickness) sequences; additional diffusion weighted imaging (RESOLVE DWI; TR 6070 ms/ TE 65 ms, b=0 & 1000; 4 mm slice thickness), and a contrast-enhanced T1 weighted 3d (gradient echo [MP RAGE]; TR 2200 ms/ TE 2,45 ms; 1 mm slice thickness) sequence was executed. The CT scan shows a unilateral expansile cyst with a maximum diameter of 21 mm and thinned sclerotic but sharply defined osseous margins in the right petrous apex, the body of the sphenoid bone and the clivus (Figure 1). The lesion erodes the bone to the posterior portion of Meckel's cave, the carotid canal wall and the petrooccipital fissure. The cyst content has got a homogenous fluid-like attenuation (10-20 HU). T1-weighted MR images show a homogenous low signal, non-enhancing cyst content and a thin wall of low signal intensity encircling the cyst, with mild rim enhancement. T2-weighted MR images reveal fluid-like homogeneous hyperintensity. There is no diffusion restriction within the cyst. The cyst neither extends into the cerebellopontine angle nor into the inner or middle ear structures. In the contrast enhanced MR images and the 3d T2-weighted images the cyst appears related with Meckel's cave (Figure 2a-d).



Figure 1) Axial bone algorithm CT scan shows a unilateral expansile cyst extending into the clivus and the right petrous apex (asterisk)

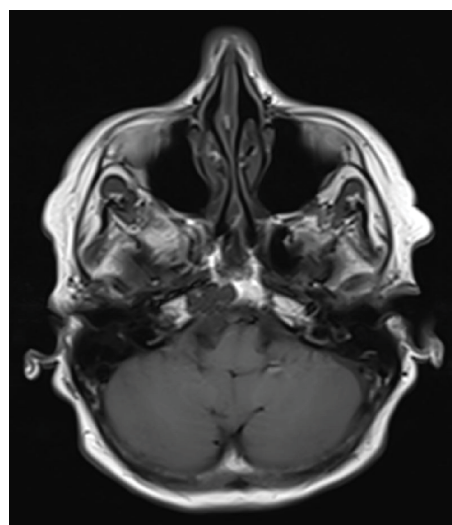


Figure 2a) Axial non-contrast T1-weighted SE (TR 600ms/ TE 10ms/ slice thickness: 5 mm) MR image reveals a homogenous low signal process, like a cyst (arrow)

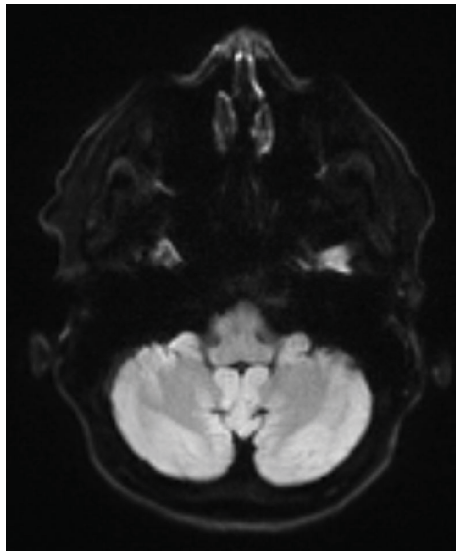


Figure 2b) Axial DWI [diffusion weighted imaging] (TR 6070ms/ TE 65ms/ slice thickness: 4 mm) MR image demonstrates no diffusion restriction within the lesion

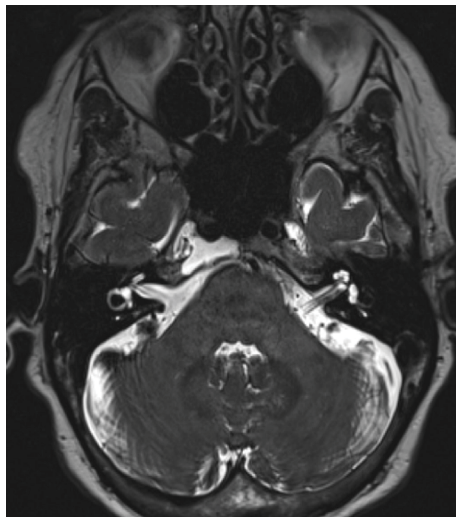


Figure 2c) Axial T2-weighted 3d SE (TR 1400 ms/ TE 152ms/ slice thickness: 0,8 mm) MR image reveals a cyst (asterisk), that appears related with Meckel's cave (arrow)

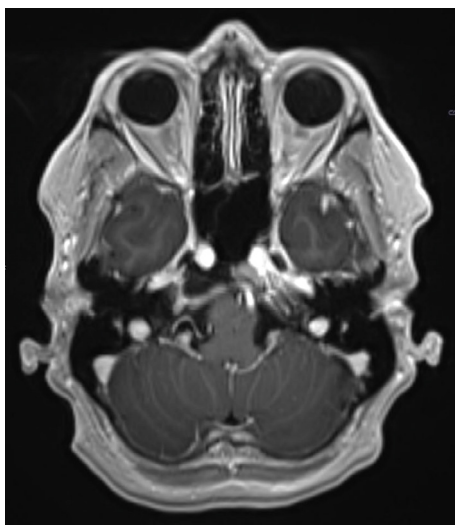


Figure 2d) Axial contrast-enhanced fatsaturated T1-weighted GE (TR 2200ms/ TE 2,45ms; slice thickness: 1 mm) MR image shows a low-signal-intensity cyst with mild rim enhancement (arrow)

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