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Case Report .

Petrous Apex Cephalocele: A Rare Case Report

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ABSTRACT

Cephalocele of the petrous apex is considered a rare condition that affects the petrous apex. Our aim is to report a rare case of a petrous apex cephalocele in a 44-year-old.

We will discuss her clinical presentation, radiographic imaging, pathological findings and literature review. Because the occurrence of such a condition is rare, we believe that reporting this case would add more information to the existing fund of knowledge.

Keywords: Cephalocele, Petrous Apex, Temporal Bone.

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INTRODUCTION

Cephalocele of the petrous apex is a rare condition. It is characterized by cystic expansion and herniation of the arachnoid or dura maters from Meckel's cave. The stimulus for petrous apex cephalocele formation is unclear. A possible mechanism for the formation of a cephalocele is a transient or sustained increase in intracranial pressure (ICP) that is transmitted into Meckel's cave. More details will be discussed later.

CASE PRESENTATION

A 44-year-old Saudi female patient known case of prolactinoma, presented to the Otology and skull base clinic at our center, complaining of postural dizziness. Associated with referred otalgia. No cerebrospinal fluid (CSF) rhinorrhea, hearing loss or tinnitus. The patient was referred from Neurology clinic due to recurrent migraine headache for three years, associated with blurred vision. The headache slightly improved by Botox injection two years ago. Neurologic examination was normal, Videonystagmography (VNG) showed no central or vestibular abnormality. Initial Investigations requested included CT scan, MRI. Imaging studies showed bilateral cystic lesion on the petrous apex (Figure 1,2,3) and a partial empty sella turcica was also noticed (Figure 4). The characteristic findings on CT and MRI suggested a Petrous Apex Cephalocele (PAC). The Patient was advised to follow up every six months then yearly at the clinic and with imaging. The last follow up three years after presentation, MRI showed no progression and the Patient didn't develop any symptoms (Fig 5a-b).

Cephalocele of the petrous apex is a rare condition. It is characterized by cystic expansion and herniation of the arachnoid or dura maters from Meckel's cave.[1-3] Petrous apex is the middle part of the petrous portion of the temporal bone; it is pyramidal in shape and a solid bone that protects the auditory and vestibular apparatus.[2-4] The stimulus for petrous apex cephalocele formation is unclear. A possible mechanism for the formation of a CSF cephalocele is a transient or sustained increase in ICP that is transmitted into Meckel's cave.[3]

Lesions of petrous apex are difficult to diagnose based on clinical findings only. Imaging studies play a significant role in the diagnosis and demonstration of the extension of the lesions through the skull base. The imaging study of choice to diagnose a petrous apex cephalocele is MRI.[2-5]

There are many differential diagnoses for petrous apex lesions which include cholesteatoma, cholesterol granuloma, cephalocele and mucocele. It is important to know that none of the other differentials arise from Meckel's cave.[1]

Most of the time, cases are asymptomatic and discovered incidentally during imaging for other indication. However, it can be symptomatic in some patients.[1,2,4] Because the occurrence of such a condition is rare, it can be easily missed most of the time and misdiagnosed as cholesteatoma which is different in pathophysiology and management.[4]

Signs and symptoms vary from patient to another, including hearing loss, headache, CSF otorrhea, rhinorrhea, tinnitus, diplopia and facial spasm. Factors that determine the severity of the defect include the size, nature of the lesion and the invasion of nearby structures.[4,5]

In regards to our patient, based on her presentation and imaging, cephalocele was suspected, symptomatic treatment showed improvement which led us consider other causes for the headache and vertigo rather than cephalocele.

There are key factors that determine the type of management for cephalocele, either conservative or surgical approach, depending on clinical presentation and anatomic location.[3-4] Operative indications include progressive growth, neural compression, hydrocephalus, seizures, refractory symptoms or CSF otorrhea\rhinorrhea. Typically; for incidentally discovered lesions treatment is not recommended.[1,4] Considering that our patient does not have any significant symptoms that justify surgical intervention and depending on the series of images over the past three years that showed stable lesion (Figure 5a-b). Therefore, conservative management is applicable to such a case. Also, it is important to consider the intricate anatomy of the petrous apex and its relation to critical structures that may expose the patient to complications that can be avoided with conservative approach. with careful assessment and observation for any progression of symptoms like an intractable headache that does not response to medical treatment or if there are new symptoms occur as CSF otorrhea\rhinorrhea that need immediate intervention.

Follow up with series of MRI and CT scans are helpful in detecting lesion invasion to the inner ear or the adjacent neurological structures. Any potential invasion of these structures requires surgical intervention.

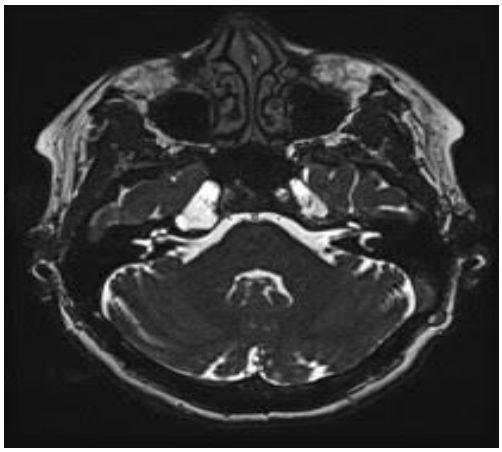


Figure 1: Axial T2 MRI image shows a high signal and bilateral expansion of the petrous apex.

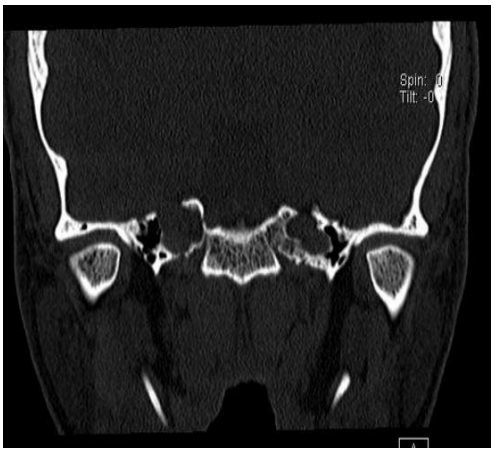


Figure 2: CT scan shows bilateral petrous apex lesion.

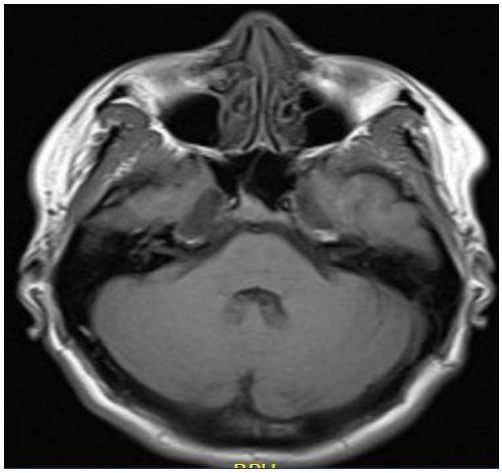


Figure 3: Axial T1 MRI image shows a hypointense bilateral apex lesions.

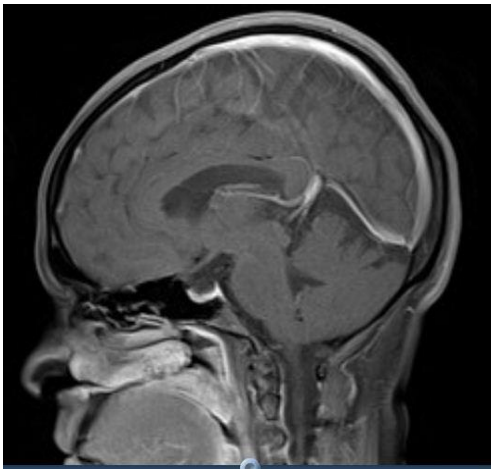
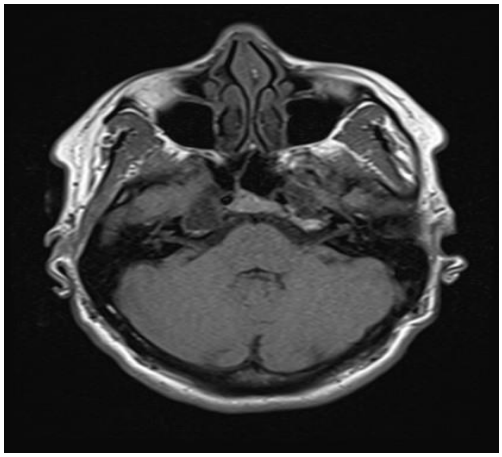


Figure 4: Sagittal MRI image shows a partial empty sella.



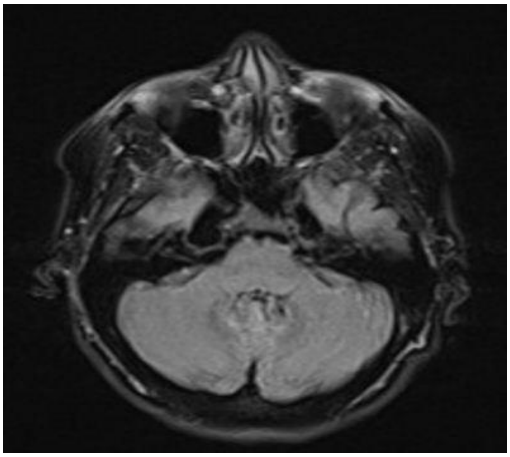


Figure 5a-b: Axial MRI images on 2013 and 2016 showing stable lesion, no significant growth or invasion to labyrinth or adjacent neurological structures.

CONCLUSION

The correct management of cephalocele requires one to make the correct diagnosis before recommending treatment and ensuring that presenting symptoms are indeed caused by cephalocele. Incidentally discovered cephalocele can be followed with serial imaging studies, whereas symptomatic lesions may require surgical intervention.

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