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Petrous apex cephalocele and empty sella/arachnoid cyst coexistence: a clue for cerebrospinal fluid pressure imbalance?

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PURPOSE
To reveal the magnetic resonance imaging (MRI) properties of incidental petrous apex cephalocele (PAC) and coexisting empty sella-arachnoid cyst.

MATERIALS AND METHODS
We reviewed our archive from June 2005 to July 2008. Four patients were diagnosed with PAC (four females; age range, 41–60 years; mean, 48.5). All patients underwent MRI examination of the cranium. We evaluated the lesions for extension into the neighboring structures, content, signal intensity, enhancement, and relation to Meckel's cave, petrous apex and for the presence of empty sella.

RESULTS
The presenting symptoms included headache for three patients and diplopia for one patient. All patients had bilateral PAC, more prominent on one side. All lesions were centered posterolateral to the Meckel's cave. They were isointense to cerebrospinal fluid signal intensity and continuous with Meckel's cave on T1W, T2W and FLAIR sequences. In two patients, there was no diffusion restriction on diffusion-weighted MR images and the ADC map. Three patients had empty sella. One patient had arachnoid cyst.

CONCLUSION
Coexistence with empty sella-arachnoid cyst raises the possibility of cerebrospinal fluid inbalance in the etiology.

Key words: • *magnetic resonance imaging* • *petrous bone* • *arachnoid cyst*

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Petrous apex cephalocele (PAC) is a congenital or acquired herniation of the posterolateral wall of Meckel's cave into the petrous apex. It is also called arachnoid cyst and meningocele (1). It is an uncommon incidental lesion. The differential diagnosis includes congenital cholesteatoma, trapped fluid, petrous apicitis, mucocele, cholesterol granuloma, and Meckel's cave trigeminal schwannoma. Preoperative differentiation is necessary before planning the appropriate operation. Histopathologically, one or all three layers of meninges may be present in PAC. The pathogenesis and natural history are yet unclear. There are two theories: congenital or acquired. Chronic cerebrospinal fluid (CSF) pulsations against the thin anterior wall of a pneumatized petrous apex might result in dehiscence (1). Coexistence with empty sella might support the second theory (2). Our purpose in this study is to reveal the magnetic resonance imaging properties of incidental PAC and coexisting empty sella or arachnoid cyst.

Material and methods

We reviewed the imaging archives between June 2005 and July 2008. Four patients were diagnosed with petrous apex cephalocele. All patients underwent magnetic resonance imaging (MRI) exam (four females; age range, 41–60; mean, 48.5). All examinations were performed on a 1.5 T MRI system (Excite, General Electrics, Milwaukee, Wisconsin, USA) with a 33 mT/ m maximum gradient capacity. Spin echo T1W (TR, 500 ms; TE, 9.6 ms; slice thickness, 5 mm; interslice gap, 1.5 mm; FOV, 24 × 18 cm; matrix, 320×192; NEX, 2) and fast-recovery fast spin echo T2W (TR, 4,240 ms; TE, 98.1 ms; slice thickness, 5 mm; interslice gap, 1.5 mm; FOV, 24 x 18 cm; matrix, 352 x 224; NEX, 2) and fluid attenuated inversion recovery (FLAIR) (TR, 8,402 ms; TE, 95.5 ms; slice thickness, 5 mm; interslice gap, 1.5 mm; matrix, 288 x 192) images were obtained. Diffusion-weighted sequence (TR, 10,000 ms; TE, 85.8 ms; slice thickness, 4 mm; interslice gap, 1 mm; matrix, 128 x 128) was performed with echo planar single shot spin echo imaging with b values of 0 and 1000 s/mm^2. Diffusion gradients were applied in three orthogonal directions to generate three sets of diffusion weighted imaging (x, y, z axes). Apparent diffusion coefficient (ADC) values were calculated automatically. The images were evaluated for extension into the neighboring structures, signal intensity, relation to Meckel's cave and petrous apex, lesion margins and coexisting empty sella.

Results

Three patients presented with headache and one with diplopia. There was no history of cerebrospinal fluid leak or trigeminal neuropathy. The lesions were bilateral. They were centered in the posterolateral portion of Meckel's cave and were continuous with it. They extended to the level

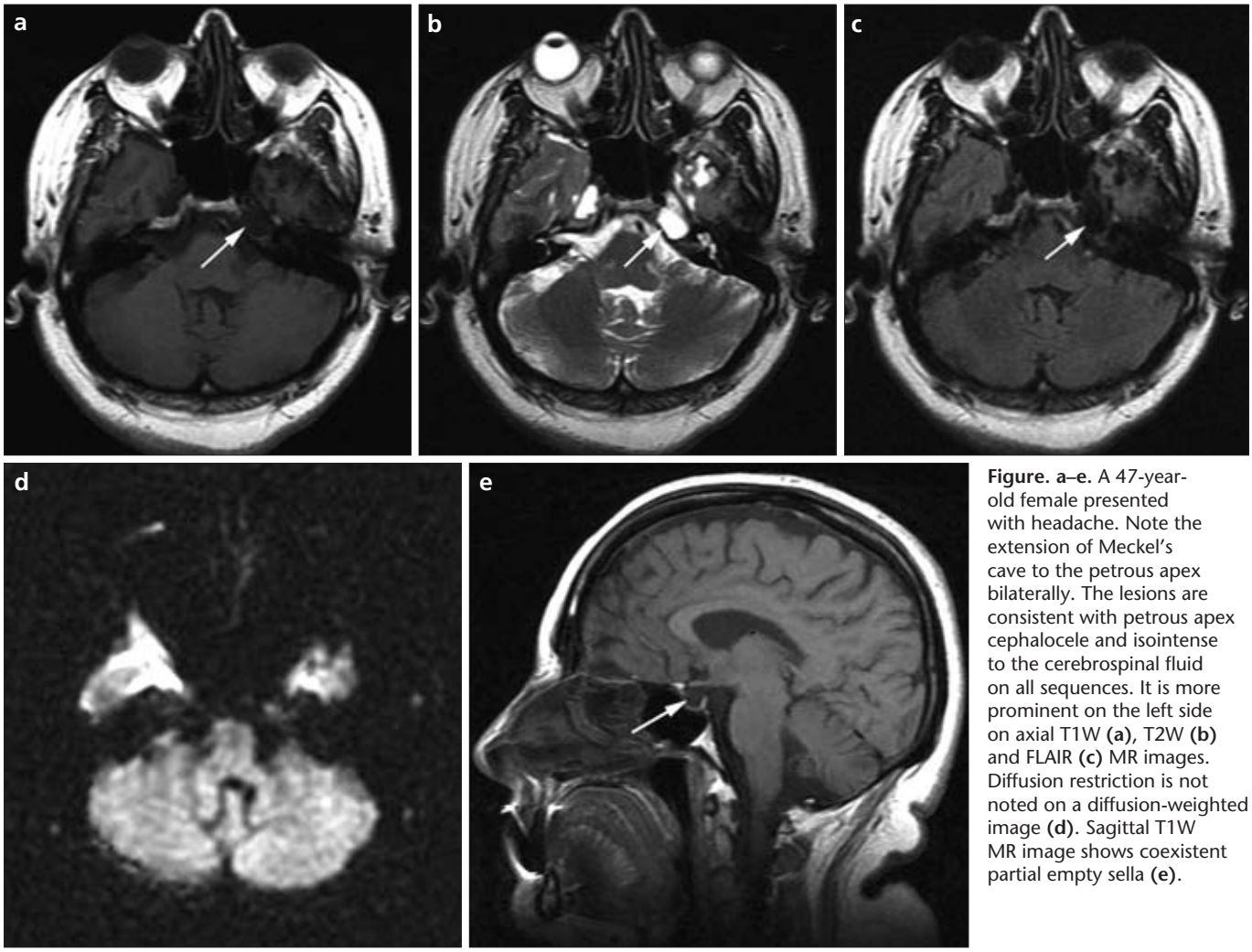

Figure. a–e. A 47-year-old female presented with headache. Note the extension of Meckel's cave to the petrous apex bilaterally. The lesions are consistent with petrous apex cephalocele and isointense to the cerebrospinal fluid on all sequences. It is more prominent on the left side on axial T1W **(a)**, T2W **(b)** and FLAIR **(c)** MR images. Diffusion restriction is not noted on a diffusion-weighted image **(d)**. Sagittal T1W MR image shows coexistent partial empty sella **(e)**.

of the internal carotid artery. None of them were related to the inner or middle ear structures. Cerebellopontine angle and internal acoustic canal were intact in all cases. The lesions were isointense to CSF signal intensity on T1W, T2W and FLAIR sequences (Fig. a–c). In two patients, there was no diffusion restriction on diffusion-weighted images and the ADC map (Fig. d). We did not obtain diffusion-weighted sequence in the other two patients. Three patients had coexisting partial empty sella (Fig. e). There was arachnoid cyst in the sylvian fissure in one patient. The findings are summarized in Table.

Table. The summary of cases

No.	Sex	Age	Symptom	Petrous apex cephalocele on MRI	Empty sella on MRI	Other MRI findings
1	F	47	Headache	Bilateral, left>right	+	Ischemia-gliosis
2	F	60	Headache	Bilateral, left>right	+	Ischemia-gliosis
3	F	41	Diplopia	Bilateral, left>right	+	Atrophy
4	F	46	Headache	Bilateral, right>left	-	Atrophy, arachnoid cyst

Discussion

Petrous apex cephalocele is a rare lesion (3). It is usually an asymptomatic incidental finding in adults. However, it should be considered as a possible cause of CSF rhinorrhea, otorrhea, and recurrent meningitis in children (3–5). The smallest patient diagnosed in the literature was 2-year-old. She had recurrent meningitis due to CSF fistulae (4). Moore et al. reported the largest series of 10 adult patients with petrous apex cephalocele (3). In their group, there was female preponderance (80%). The most common symptoms were trigeminal neuropathy (30%) and CSF fistula (10%), but the majority was asymptomatic (60%). On the aother hand, among pediatric cases in the literature only one was asymptomatic. The rest presented with recurrent meningitis, postural headache and conductive hearing loss. Surgical intervention should be considered only when symptoms are clearly linked to the presence of this lesion (3). Clinical differentiation of petrous apex lesions is impossible. Multislice computed tomography (CT) and volume-rendered images might be useful to differentiate the cystic nature, extent and location in

the bony structure. However, Hounsfield density measurements often turn out to be insufficient because of beam hardening artifacts. CT cisternography might be helpful to differentiate the communication with the arachnoid space. MRI has enabled the characterization of much common cystic lesions like cholesterol granuloma, cholesteatoma and mucocele from PAC with the use of different sequences and advanced techniques like diffusion-weighting. In order to avoid unnecessary surgery, PAC should not be confused with cholesteatoma. Arachnoid cysts have low, cholesteatomas high signal intensity on fluid attenuated inversion recovery images (6, 7). As a result of diffusion restriction, cholesteatomas are hyperintense on diffusion-weighted images (8). Cholesterol granulomas are hyperintense on T1W images. After contrast administration mucoceles might enhance peripherally. Due to increased proteinaceous content, they might be hyperintense on T1W images (6, 9). Inflammatory lesions are typically centered within the bone itself and may show expansion of the apex. PAC typically arises from the Meckel's cave and secondarily extends into the petrous apex (3). In cases complicated with CSF fistula and otorrhea, middle ear or mastoid effusions might accompany.

When we reviewed our database there were four cases with PAC. Three of them were coexistent with empty sella and one of them was coexistent with arachnoid cyst. The lesions were isointense to CSF in continuation with Meckel's cave posterolaterally. They were bilateral but prominent on one side. The CSF imbalance within the cranium might result in the development of empty sella or arachnoid cyst. Disturbance in CSF absorption results in intracranial hypertension, which leads to herniation of meninges and CSF through the weak points in the skull. Empty sella develops from deficient diaphragma sella. Spontaneous CSF leak incidence in association with empty sella is 63–100% in the literature (10–12). It is 11% in patients with nonspontaneous CSF leak (13). Almost one third of PAC cases in the literature is presented with spontaneous CSF leak (2). Arachnoid cyst is lined with arachnoid cells and continuous with the surrounding normal arachnoid matter (6). The pulsatile pressure of CSF might cause protrusions of arachnoid granulations through weak areas in the overlying dura (13). The lobulations in the borders of PAC might be due to arachnoid pits (2). In our study, all cases were associated with either empty sella or arachnoid cyst. None of them presented with CSF leak. Our findings support the results of a recent study by Alorainy where all patients had some degree of empty sella and one patient had arachnoid cyst in the middle cranial fossa (2). Even though this observation requires additional large series for statistical inferences or definite conclusions, we believe the overall failure of CSF absorption might be the explanation for coexistence of PAC and empty sella or arachnoid cyst.

The most common mistake in the preoperative radiological diagnosis is between cholesteatoma and PAC (3, 6, 14, 15). In this study, we obtained diffusion-weighted imaging in two patients, which helped the differential diagnosis from cholesteatoma. Therefore, we recommend addition of this sequence in cases of suspicion.

As a conclusion, it is necessary to differentiate PAC from more aggressive tumors in the petrous apex. The combination of MRI with diffusion-weighted imaging should be used in characterization of such lesions. Coexistence with empty sella and/or arachnoid cyst raises the possible role of disturbance of CSF circulation in pathophysiology.

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