



Unilateral expanding petrous apex cephalocele and contralateral vitreous hemorrhage in a young patient with intracranial hypertension

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ABSTRACT

Purpose: To report a case of unilateral petrous apex cephalocele (PAC) and contralateral vitreous hemorrhage in a young patient with intracranial hypertension.

Observations: A 12-year-old boy had acute visual loss in the right eye. The clinical and radiological findings were consistent with right vitreous hemorrhage with mild intracranial hypertension and left PAC. Cerebral angiography demonstrated low flow of the left inferior petrosal sinus and anterior venous high flow from the cavernous sinus to the facial vein via the superior ophthalmic vein. The presence of an expanding PAC, blocking the venous flow away from the inferior petrosal sinus, would produce an ipsilateral intraocular pressure (IOP) (20 mmHg) mildly higher than the other one (16 mmHg) although it is in the normal range, which may have caused the difference of the translaminar pressure gradient resulting from the balance between the cerebrospinal flow pressure and the IOP.

Conclusion and Importance: Unilateral expanding PAC may cause intracranial hypertension with different severity of papilledema between two eyes.

1. Introduction

Petrous apex cephalocele (PAC) is a rare cystic lesion characterized by herniation of the posterolateral wall of Meckel's cave into the petrous apex.¹ If the cyst lining consists only of arachnoid, the term of Meckel's cave arachnoid cyst is applied; if the cyst involves the dura and arachnoid, the term of Meckel's cave meningocele is applied. PAC may be incidentally detected in asymptomatic patients. Although there have been several case reports of PAC causing diplopia or trigeminal function,¹⁻⁷ no case with acute visual impairment associated with PAC has been reported previously.

We report a case of unilateral PAC and contralateral vitreous hemorrhage in a young patient with intracranial hypertension.

2. Observation

A 12-year-old boy with a 6-year history of left chronic otitis media was referred to a pediatric hospital with a chief complaint of orthostatic headache and nausea. His headache worsened at 15 minutes after assuming a standing position. Brain computed tomography (CT) showed no abnormalities except for an opacified and poorly developed mastoid

system on the left side. Post contrast magnetic resonance imaging (MRI) of the brain showed no abnormalities except for a cystic lesion of the left Meckel's cave. T2-weighted image (WI) of the spine showed a high signal intense lesion on the dorsal thoracolumbar spine. The patient was diagnosed with presumed cerebrospinal fluid (CSF) leak and underwent intravenous rehydration of 1000 ml/day along with bed rest for two weeks. His headache resolved. One month later, he noticed blurred vision in his right eye and was referred to our hospital. Upon examination, his corrected visual acuity was 0.5 in the right eye and 1.0 in the left eye. There was no relative afferent pupillary defect. Averaged intraocular pressure (IOP) was 16.0 mmHg in the right eye and 20.0 mmHg in the left eye measured by noncontact tonometer three times. Slit-lamp examinations showed no abnormalities. Funduscopic examinations showed bilateral optic discs swelling and peripapillary vitreous hemorrhage in the right eye (Fig. 1). Humphrey 30-2 visual field examination showed bilateral enlarged blind spots and a cecocentral scotoma in the right eye. MRI of the brain showed cystic mass lesion of the left Meckel's cave toward the cavernous sinus, which had high signal intensity on T2-WI, low signal intensity on T1-WI and diffusion-WI, and no enhancement on post contrast T1-WI, consistent with PAC (Fig. 2A-D). MRI additionally demonstrated left enlarged superior

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Fig. 1. Fundus photograph showed bilateral severe discs swelling and peripapillary vitreous hemorrhage in the right eye.

ophthalmic, angular veins in the left orbit (Fig. 2E). Selective left internal carotid arterial angiography of the late phase showed attenuation of the left inferior petrosal sinus and enlarged anterior venous outflow from the left cavernous sinus to the facial vein via the superior ophthalmic vein (Fig. 3). Lumbar puncture (LP) reveals an opening pressure of 250 mm H₂O without sedation in the lateral decubitus position. CSF analysis was normal. Serum laboratory evaluations were all normal. To decrease the high intracranial pressure (ICP), the patient was administered 250 mg acetazolamide twice daily for one month. Subsequently, the vitreous hemorrhage and papilledema gradually resolved (Fig. 4).

3. Discussion

Our patient had an expanding PAC and contralateral severe vitreous hemorrhage resulting in acute visual loss. Friedman et al. published the revised diagnostic criteria for the idiopathic intracranial hypertension in adult and children in 2013, including elevated LP opening pressure (≥ 250 mm CSF in adults and ≥ 280 mm CSF in children [250 mm CSF if the child is not sedated and not obese]).⁸ Additionally, to account for the possibility of patients with the clinical syndrome of intracranial hypertension having ICP below this cutoff, current criteria allow for borderline ICP of 200–250 mmH₂O when symptoms (pulsatile tinnitus), ophthalmologic examination findings (Frisen grade 2 papilledema or sixth nerve palsy) or high ICP-associated neuroimaging findings are present.^{9,10} Our patient was not sedated at the time of the LP and not obese. Additionally, he had bilateral fundus pictures of Frisen grade 5 papilledema, which would be diagnosed as intracranial hypertension secondary to PAC.

It is unknown whether our patient definitely had intracranial hypertension, as he did not undergo LP when he complained of orthostatic headache and nausea. Some previous reports describe that PAC may produce the clinical manifestation related to a CSF fistula.^{1,4,11–13} The lesion triggered a CSF fistula in these patients when a tear occurred in the cyst wall, allowing CSF to leak into the petrous apex. PAC of our patient may be the possible source of the CSF leak.

In our patient, PAC at the time of the management of intracranial hypertension was larger than at the time when the patient was initially diagnosed as CSF leak (Fig. 5). On the basis of endoscopic observation of suprasellar prepontine arachnoid cyst, a ball-valve mechanism is suspected to be the basis of enlargement.^{14,15} Harani et al. proposed that one-way slit valve is responsible for the net influx of CSF into the cyst and for its enlargement.¹⁶ In any case, the presence of enlarged PAC of our patient, blocking the venous flow away from the left inferior petrosal sinus and causing a retrograde flow back to the facial vein, would produce the low flow in the inferior petrosal sinus draining on the same side.

I have no explanation of the reason why vitreous hemorrhage in our case occurred on the opposite side of PAC. Watson et al. described a similar case of unilateral vitreous hemorrhage in a 14-year-old boy with idiopathic intracranial hypertension and speculate that the degree of optic nerve head swelling could conceivably have caused mechanical stretching and consequent rupture of a small vessel resulting in vitreous hemorrhage.¹⁷

However, there is no description about the asymmetry of the fundoscopic findings. Bidot described that “very asymmetric” papilledema define as a ≥ 2 modified Frisen grade difference between the two eyes.¹⁸ In our patient, it appears that vitreous hemorrhage with papilledema in the left eye may be slightly more severe than that of the right eye although the term of “very asymmetric” papilledema does not apply. Several mechanisms such as asymmetrical structural change in the lamina cribrosa,¹⁹ along the optic nerve sheath,²⁰ or into the optic canal¹⁸ have been suggested to explain the asymmetry of papilledema, but its mechanism remains unclear. The pathogenesis of papilledema depends on the translaminar pressure gradient resulting from the balance between the CSF pressure and the IOP. If the gradient is different between both eyes, asymmetric papilledema may be produced.^{21–23} Our patient had normal IOP for either eye but with left eye (20 mmHg) higher than the right eye (16 mmHg), which probably might be secondary to the orbital venous congestion due to arising in the low flow in the inferior petrosal sinus draining. The vitreous hemorrhage occurred on the right side, which had a higher translaminar pressure gradient (ICP minus IOP: +2.6 mmHg) as compared to the left side (ICP minus IOP: –1.4 mmHg). Venous congestion of the left orbit causing ipsilateral IOP higher than that of the other eye may have restrained the severity of papilledema by reducing the translaminar pressure gradient.

Patient consent

Consent to publish the case reports was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

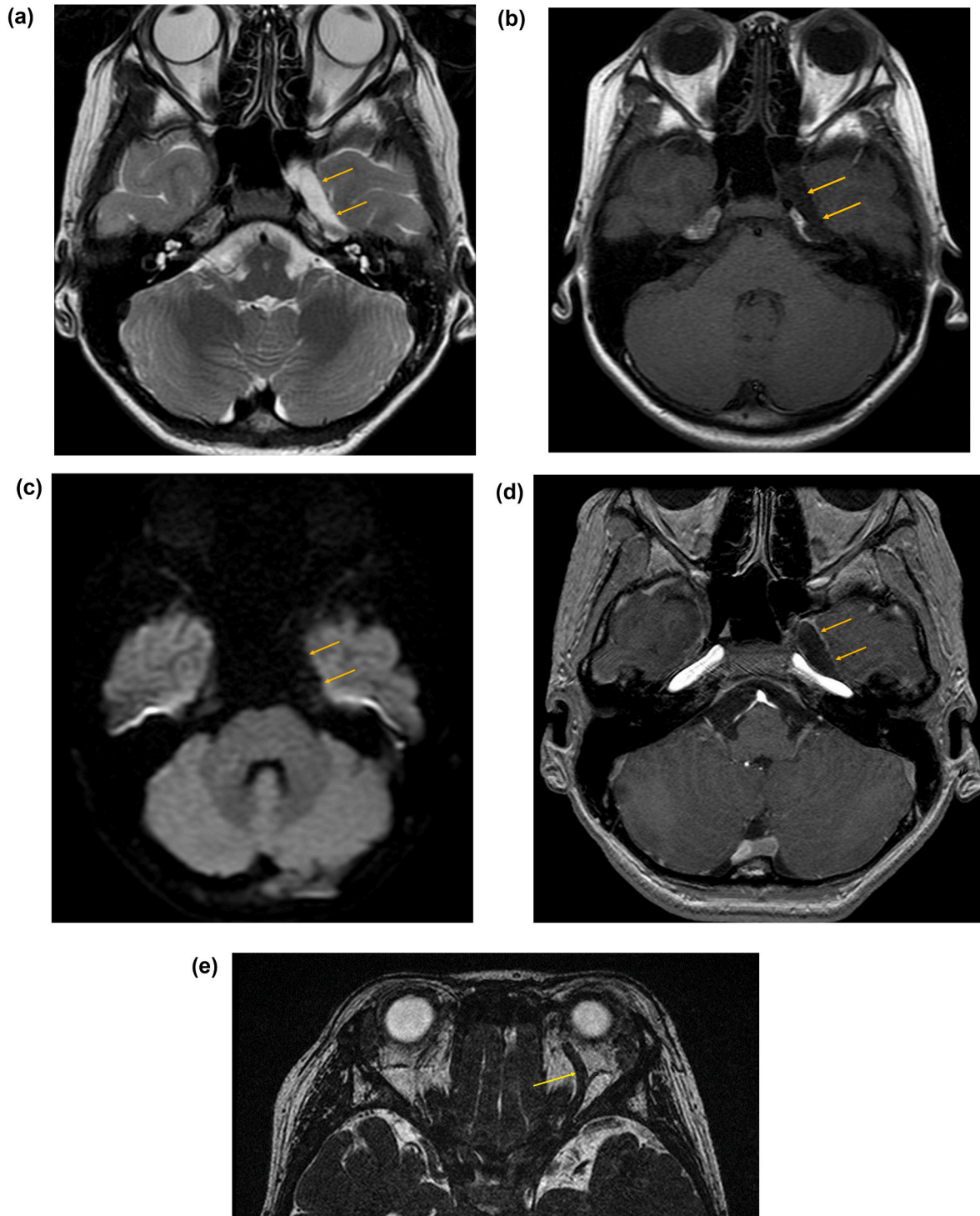


Fig. 2. Brain MRI showed cystic lesion (arrows) arising from the left Meckel's cave and extending along the petrous apex, which had high signal intensity on T2-weighted image (WI) (A), low signal intensity on T1-WI (B) and diffusion-WI (C) and no enhancement on post contrast T1-WI (D). MRI of the orbit showed left enlarged superior ophthalmic vein (arrow) (E).

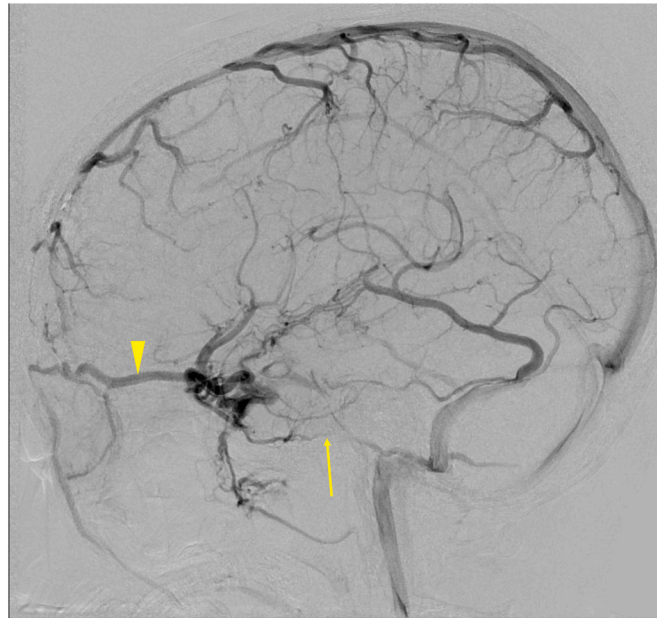


Fig. 3. Fluoroscopic image from left internal carotid arteriogram in delayed venous phase on the sagittal image showed attenuation of left inferior petrosal sinus (arrow) and enlarged anterior venous outflow from the left cavernous sinus to the facial vein via the enlarged superior ophthalmic vein (arrowhead).



Fig. 4. The patient's papilledema and vitreous hemorrhage resolved after management of intracranial hypertension.

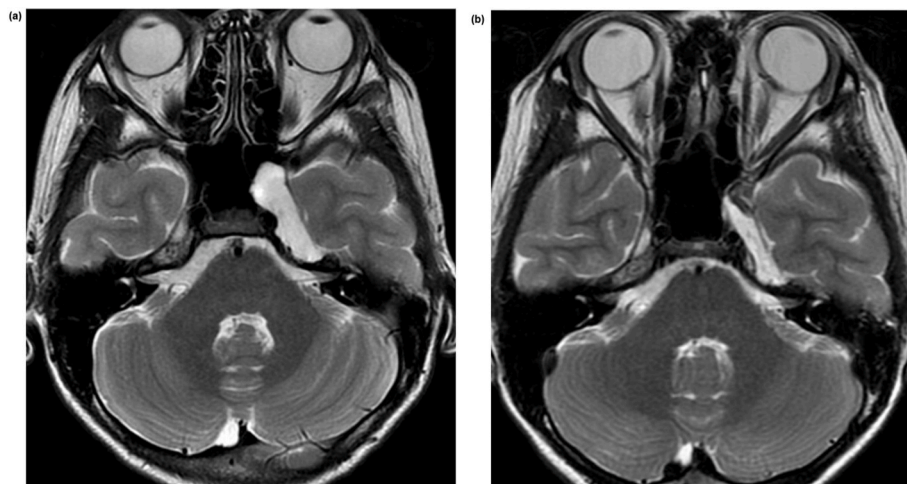


Fig. 5. PAC (A) at the time of the management of intracranial hypertension (size: 39 × 14 mm) is larger than (B) at the time when he was initially diagnosed as CSF leak (size: 32 × 9 mm) on MRI.

Declaration of competing interest

The authors have no financial disclosures.

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