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Arachnoid Cyst of the Velum Interpositum : Coincidence with Multiple Cranial Neuropathies

- Case Report -

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Arachnoid cyst of the velum interpositum is unusual and causes symptoms similar to those seen with a third ventricular mass. This report describes a case in which the arachnoid cyst occupied the cistern of the velum interpositum and was coincident with multiple cranial neuropathies. The patient was treated by endoscopic fenestration of the cyst. The surgery resulted in decreased in the size of the cyst but aggravation of cranial neuropathies. The patient underwent methylprednisolone pulse therapy and intravenous immunoglobulin administration under the impression of the multiple cranial neuropathies and recovered completely 3 months later.

The authors conclude that combined neurological disease that needs medical treatment should be differentiated and priority of the treatment should be determined carefully if arachnoid cyst is associated with unrelated or ambiguous neurological symptoms and signs. Careful considerations of cause and effect may avoid an unnecessary surgical manipulation.

KEY WORDS : Arachnoid cyst · Velum interpositum · Cranial neuropathy · Endoscopic fenestration.

Introduction

The cistern of velum interpositum represents a potential space between the tela choroidea and the columns of the fornices above[8]. This potential space is actually a forward extension of the quadrigeminal plate cistern. This location of arachnoid cyst is unusual[2,3,5,7]. Although arachnoid cysts are often asymptomatic, these lesions can result in serious neurologic complications by obstruction to the normal flow of cerebrospinal fluid(CSF)[4] and may need an urgent surgical intervention. But arachnoid cysts of velum interpositum may be incidentally detected or may be associated with unrelated symptoms of a concurrent disease. We report a case of arachnoid cyst of the velum interpositum accompanying multiple cranial neuropathies(III, VI, and XII cranial nerves) and discuss the priority of treatment in such case.

Case Report

A 10-year-old boy presented with a 4-day history of swallowing difficulty, diplopia, ptosis of the left eye. His medical history was unremarkable. A neurological examination on admission revealed total external ophthalmoplegia with pupillary sparing on the left side. Tongue deviation to the right side was also evident. No other neurologic deficits were noticed. Magnetic resonance(MR) imaging disclosed a 3×4cm sized cyst occupying the cistern of the velum interpositum. It had a triangular configuration on the axial plane, with the anterior apex at the level of the foramen of Monro and posterolateral extremities at the level of the choroid plexus of the lateral ventricles. The cyst wall was clearly visible. Quadrigeminal plate was mildly displaced inferiorly and posterior part of corpus callosum was mildly compressed by cyst in sagittal image(Fig. 1).

The patient underwent endoscopic fenestration of the cyst for decompression. Through a left burr hole, a 4mm flexible endoscope(Codman & Shurtleff) was introduced into the frontal horn of the left lateral ventricle. Under direct visualization through a small microchip camera, the ventricular cavity was explored and the limit between the lateral ventricle and the cyst was determined. After assaying the consistency of the cyst wall, two fenestrations were performed using

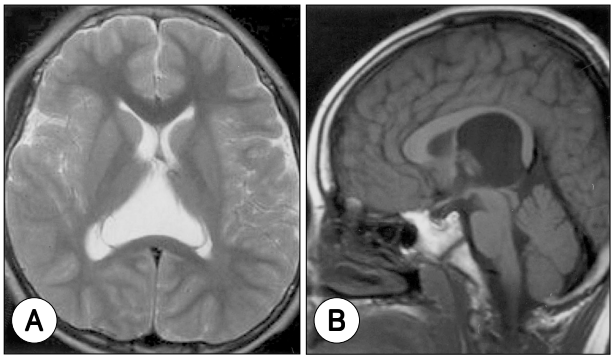

Fig. 1. A : T_2-weighted axial magnetic resonance(MR) imaging revealing a 3×4cm sized cyst occupying the cistern of the velum interpositum. It has a triangular configuration on the axial plane, with the anterior apex at the level of the foramen of Monro and posterolateral extremities at the level of the choroid plexus of the lateral ventricles. The cyst wall was clearly visible. B : T_1-weighted sagittal MR image showing that quadrigeminal plate is mildly displaced inferiorly and posterior part of corpus callosum is mildly compressed by cyst.

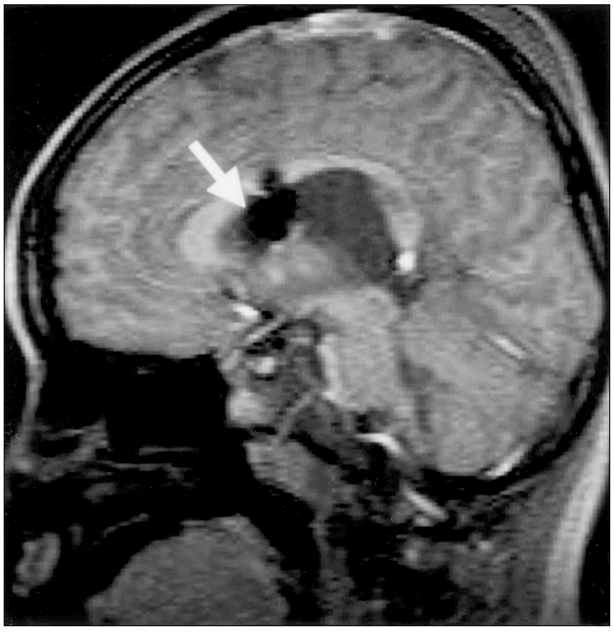

Fig. 2. Postoperative T_1-weighted sagittal magnetic resonance image demonstrating decreased size of the cyst and air(white arrow) in the lateral ventricle.

monopolar radiofrequency coagulation. After fenestrations had been completed, free flow of CSF between the cyst and the lateral ventricle was confirmed under the direct vision.

Postoperatively, the preoperative symptoms and signs were not resolved and progressed. Three days later, a neurological examination revealed paresis of the right oculomotor nerve with pupillary sparing, and MR image demonstrated decreased size of the cyst and air in the lateral ventricle(Fig. 2). The patient was referred to neurologist for evaluation of the unusual cranial nerve palsies. Nerve conduction test revealed normal velocity of both side. Administration of methylprednisolone(10mg/kg/day for 7 days) and immunoglobulin(0.4 mg/kg/day for 5 days) under the impression of the multiple cranial neuropathies were done promptly. His symptoms and signs was slightly improved 3 weeks later and recovered completely three months later.

Discussion

The cistern of the velum interpositum is formed during the interhemispheric cleavage in relation to the development of the corpus callosum. It is located between the fornix above, the roof of the third ventricle below and the choroids plexuses of the ventricles laterally ; it is entirely surrounded by pia mater, which is invaginated between the fornix and the roof of the third ventricle. This cistern extends anteriorly to the foramen of Monro, whereas posteriorly it opens in the cistern of the vein of Galen[8]. As arachnoid cysts have been shown to arise between the layers of the arachnoid tissue, this arachnoid cyst probably arose from the tela choroidea and bulged upward into the space of the cistern of the velum interpositum.

Arachnoid cyst of the velum interpositum is unusual and only several cases have been reported[2,3,5,7]. But a cyst of the velum interpositum is confused with cystic dilatation of the cavum veli interpositi. The cavum velum interpositum communicates with adjacent cisterns and typically does not cause mass effect or hydrocephalus. A cyst of the velum interpositum, in contradistinction, is prone to obstruct the ventricles, causing clinical symptoms that can mimic those of a third ventricular mass[5,7]. In present case, the cyst was not associated with obstructive hydrocephalus and presented with multiple cranial neuropathies that imply brainstem lesion. In midline cut of sagittal images, brainstem was mildly compressed by cyst, but that was not enough to develop brainstem signs. However, we thought that endoscopic fenestration for decompression of the cyst was the best option of the treatment in acutely progressing clinical course because arachnoid cyst seemed to compress the brainstem. In conclusion, arachnoid cyst of the velum interpositum was asymptomatic and was associated with the other neurological disease entity that needs medical treatment.

Several cranial nerves may be affected by a single disease process. One of the clinical problems that arise is whether the lesion lies within or outside the brainstem. Lesions lying on the surface of the brainstem are characterized by involvement of adjacent cranial nerves and by late and only slight

if any involvement of the long sensory and motor pathways and segmental structures lying within the brainstem. Multiple cranial neuropathies may precede or accompany viral infections of cranial nerves[9]. Although antibody titers to a variety of viruses are sometimes increased, such findings are quite nonspecific and do not establish a viral etiology. Some of cases probably represent a variant form of the Guillain-Barre syndrome, insofar as they may be preceded by a nonspecific infection and accompanied by areflexia, evanescent paresthesias and/or weakness of the extremities, and an elevated spinal fluid protein without pleocytosis[1]. In rare instances, none of these features are evident and the neuropathy is strictly confined to cranial structures-in which case the status of the cranial neuropathy remains uncertain[6]. In either case, recovery is usually prompt, in a matter of several weeks, and quite complete.

In summary, we present the arachnoid cyst of the velum interpositum associated with multiple cranial neuropathies. Although endoscopic fenestration for decompression of the cyst was successful, the patient's symptoms and signs were aggravated. Thereafter, the patient was administrated with steroid and immunoglobulin under the impression of multiple cranial neuropathies. Preoperative symptoms and signs recovered completely. Arachnoid cyst may be associated with unrelated or ambiguous neurological symptoms and signs. Combined neurological disease that needs medical treatment should be differentiated and priority of the treatment should be determined carefully. Careful considerations of cause and effect may avoid an unnecessary surgical manipulation.

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