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Case Report

Regression of cerebellar tonsillar descent and hydrocephalus after endoscopic third ventriculostomy in a patient with a quadrigeminal arachnoid cyst

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Abstract

Background: Posterior fossa arachnoid cysts, including quadrigeminal cistern arachnoid cysts, can occasionally cause compression of the quadrigeminal plate, leading to Sylvian aqueduct stenosis and induction of cerebellar tonsillar descent into the foramen magnum. This, in turn, can result in obstructive hydrocephalus. In such cases, the characteristic of hydrocephalus is generally considered to be hypertensive.

Case Description: We present the case of a 28-year-old female complaining of chronic and progressively worsening headaches following the delivery of her first child. Magnetic resonance imaging revealed marked tri-ventriculomegaly, the arachnoid cyst located in the quadrigeminal cistern, and cerebellar tonsillar descent. Ophthalmoscopy revealed bilateral papilledema indicating a long-standing elevation of intracranial pressure. Endoscopic third ventriculostomy (ETV) was performed successfully and resulted in complete recovery from her headaches and papilledema. Postoperative MRI revealed resolution of ventriculomegaly and cerebellar tonsillar descent, suggesting that the fourth ventricle outlet obstruction was associated with the development of the hydrocephalus in this patient.

Conclusion: Our case is the first report that a quadrigeminal arachnoid cyst associated with both cerebellar tonsillar descent and hydrocephalus was well treated with ETV. It was indicated that the patient's hydrocephalus and cerebellar tonsillar descent were secondary and synergistic events, caused by the arachnoid cyst located in the quadrigeminal cistern.

Key Words: Arachnoid cyst, endoscopic third ventriculostomy, hydrocephalus, tonsillar descent

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INTRODUCTION

Posterior fossa mass lesions, such as tumors or arachnoid cysts, may lead to cerebellar tonsillar descent, which is also known as a Chiari malformation type I (CM-I).[8,10] CM-I is a structural abnormality of the cerebellum in which

elongation of the cerebellar tonsils results in a portion of it protruding through the foramen magnum, causing cerebellar tonsillar herniation, syringomyelia, and occasionally hydrocephalus. Although congenital CM-1 can also be induced or acquired following raised intracranial pressure secondary to a tumor or cyst.[1,2]

The quadrigeminal cistern arachnoid cyst (QCAC), which was first described by Di Riocco in 1993, can extend to the trigon cranially, to the supracerebellar cistern caudally, to the third ventricle anteriorly, and to the ambient cisterns laterally.[4,6] Therefore, a QCAC extending into the supracerebellar cistern can lead to not only cerebellar tonsillar herniation but also the aqueductal stenosis, a common cause of obstructive hydrocephalus.[8] This kind of obstructive hydrocephalus could be a good candidate for endoscopic third ventriculostomy (ETV). Here, we present the case of a 28-year-old female with QCAC, whose cerebellar tonsillar descent and subsequent hydrocephalus were well relieved by ETV.

CASE REPORT

A 28-year-old female presented at our hospital with a 2-year history of headaches. Her headaches, located at occipital and temporal regions, became progressively worse after the delivery of her first child 5 months ago. Her neurological status was intact except for numbness in her left arm, a symptom that appeared after the delivery. Ophthalmoscopy revealed bilateral papilledema. Her medical and family history was unremarkable.

Magnetic resonance imaging (MRI) revealed marked dilatation of the lateral and third ventricles with downward bowing of the latter's floor; the fourth ventricle was normal. The cyst, which was considered as congenital arachnoid cyst, was located at the quadrigeminal cistern extending caudally into the supracerebellar cistern [Figure 1a and b]. The cyst compressed downward into both the cerebellar vermis and the hemispheres, leading to cerebellar tonsillar descent and Sylvian aqueductal stenosis. However, heavily T2-weighted images revealed a definite discontinuity between the anterior part of the cyst wall and the third ventricle [Figure 1c]. Moreover, no syringomyelia in the cervical spinal cord was observed.

Hydrocephalus was believed to have developed as follows: the QCAC extending to the supracerebellar cistern compressed on the cerebellar tonsils, forcing them to move downward and plug the foramen magnum. This in turn blocked the flow of cerebrospinal fluid (CSF) at the fourth ventricle outlet resulting in the accumulation of CSF and hypertensive hydrocephalus.

After induction of general anesthesia, the patient underwent ETV with the use of a fiberscope (VEF-V, Olympus Co.). Endoscopically, the floor of the third ventricle looked very thin, suggesting the existence of a long-standing hypertensive dilatation of the third ventricle. The endoscope was then advanced towards the quadrigeminal cistern, which revealed the cistern was surrounded entirely by a membrane consistent with the membrane of an arachnoid cyst. As shown on the heavily T2-weighted MRI image [Figure 1c], the cyst wall facing the posterior part of the third ventricle was opened widely [Figure 2a]. The aqueduct stenosis was easily apparent [Figure 2b]. ETV was successfully performed enabling communication between the third ventricle and prepontine cistern.

Her postoperative course was uneventful, and her headaches, left arm numbness, and bilateral papilledema were completely resolved. A postoperative MRI of the brain at the 12-month follow-up displayed marked reductions in size of the lateral and third ventricles and the quadrigeminal cyst; it also revealed normalization of the cerebellar tonsillar descent, and patency of the stoma was clearly indicated by the flow artifact, which is depicted on the postoperative T2-weighted image in Figure 3.

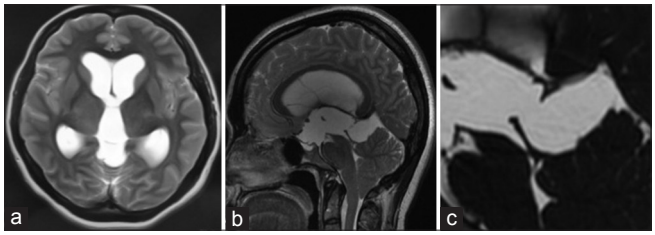

Figure 1: (a) PreoperariveT2-weighted on the axial section revealed a marked dilatation of the lateral and third ventricles. (b) Sagittal section of MRI showed that the cyst was iso-intense to the CSF, indicating that the arachnoid cyst was located at the quadrigeminal cistern and extended into the supracerebellar cistern and to the floor of the third ventricle, protruding downwards; it also revealed that the fourth ventricle was normal. (c) Heavily T2-weighted MRI image clearly revealed the discontinuity between the anterior part of the cyst wall and the third ventricle.

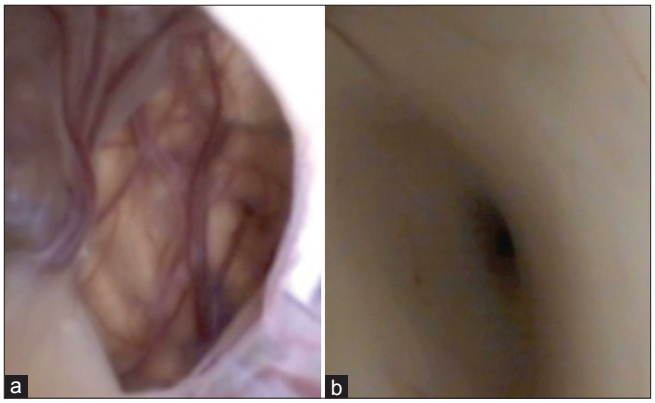

Figure 2: (a) Intraoperative endoscopic view showed that the cyst wall facing the posterior part of the third ventricle was opened widely and the cyst wall was confirmed to consist of arachnoid membrane through the endoscopic view. (b) It was demonstrated that the aqueduct was severely narrowed through the endoscopic view.

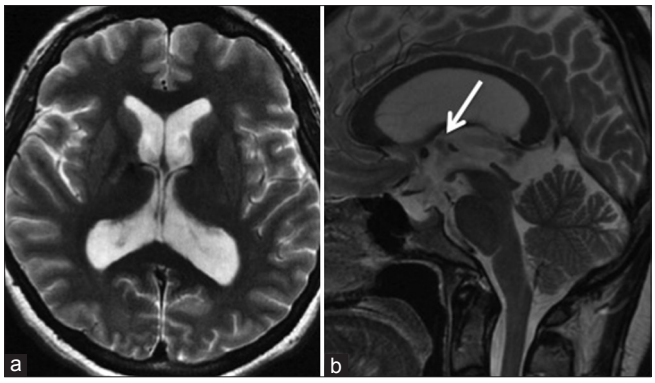

Figure 3: (a) A postoperative MRI displayed marked reductions in the size of the lateral and third ventricles. (b) The size of the quadrigeminal cistern arachnoid cyst was also reduced, and normalization of the cerebellar tonsillar descent was observed.The flow void artifact through the stoma with ETV was detected (arrow).

DISCUSSION

The quadrigeminal cistern is a rare location for the origin of an arachnoid cyst.[11,12] Typical symptoms and signs of QCAC include headaches, vomiting, and visual complaints caused by compression against the quadrigeminal plate, which in turn can develop into Parinaud's phenomenon. It can also bear down on the aqueduct of Sylvius leading to obstructive hydrocephalus.[1,9]

The QCAC can extend to the regions of the trigon cranially, to the supracerebellar cistern caudally, to the third ventricle anteriorly, and to the ambient cisterns laterally. The QCAC can cause not only aqueduct stenosis but also cerebellar tonsillar herniation following extension into the supracerebellar cistern; both in turn can lead to the development of obstructive hydrocephalus. This kind of obstructive hydrocephalus can be effectively treated with ETV.[8,10] In our patient, both aqueductal stenosis and cerebellar tonsillar herniation might be attributable to the development of obstructive hydrocephalus. Chronic elevation of the intracranial pressure was indicated by persistent headaches and papilledema.

The diagnosis of the QCAC was confirmed by endoscopic observation; the whole of the cyst wall consisted of the arachnoid membrane. In our case, the development of communication between the third ventricle and the arachnoid cyst remains unclear. However, we speculate that it may have occurred as follows: the QCAC extended into the supracerebellar cistern causing the aqueduct stenosis and cerebellar tonsillar herniation, and subsequent hypertensive obstructive hydrocephalus. Therefore, in our patient, the headaches gradually worsened. In addition, the excess elevation of intracranial pressure associated with the delivery of her first child might have induced the tear in the cyst wall, enabling subsequent communication between the arachnoid cyst and the third ventricle. This communication could have led to the direct transmission of the elevated intracranial pressure into the cyst leading to the subsequent progression of the aqueduct stenosis and the cerebellar tonsillar herniation. It is also possible that the excess elevation of intracranial pressure associated with the delivery of her first child could have attributed to the cerebellar tonsillar herniation directly. It was suggested that her left arm numbness was derived from the tonsillar herniation. As a result, this mal-circuit developing intracranial hypertension might have happened after the delivery.

ETV is reported to be quite effective for the treatment of obstructive hydrocephalus, especially when associated with tumors and cysts.[3,7] We speculated that ETV would release the elevated pressure in the third ventricle out into the interpeduncular and prepontine cistern sufficiently. Following the procedure, the patient's symptoms were relieved, concurrently with a reduction in the volume of the lateral and third ventricles, the QCAC, and regression of the cerebellar tonsillar descent.

In some CM-I patients with posterior fossa arachnoid cysts, such as quadrigeminal portion, or in patients with aqueductal stenosis, their tonsillar descents have been improved following CSF alteration procedures, such as cyst peritoneal shunting or endoscopic procedures.[5,10] Galarza *et al*. reported 10 cases of posterior arachnoid cysts associated with cerebellar tonsillar descent, one of which was located at the quadrigeminal cistern, whereby the patient exhibited signs of ataxia and nystagmus. However, this 45-year-old male required no surgical intervention.[8] To the best of our knowledge, this case is the first to report that a quadrigeminal arachnoid cyst associated with both cerebellar tonsillar descent and hydrocephalus was well treated with ETV. It was indicated that the patient's hydrocephalus and cerebellar tonsillar descent was a secondary and synergistic event, caused by the arachnoid cyst located in the quadrigeminal cistern.

CONCLUSION

Cerebellar tonsillar descent and hydrocephalus were derived from an aqueductalstenosis due to the downward extension of quadrigeminal arachnoid cyst. It is very important to recognize that these events were secondary and synergic, and treated well with ETV.

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