Endoscopic Fenestration of Posterior Fossa Arachnoid Cyst for the Treatment of Presyrinx Myelopathy
—Case Report—

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Abstract

A 32-year-old man presented with an arachnoid cyst of the posterior fossa manifesting as cervical syringomyelic myelopathy. Magnetic resonance (MR) imaging demonstrated edematous enlargement and T2 prolongation of the cervical spinal cord, indicating a “presyrinx” state. MR imaging showed the inferior wall of the cyst, which disturbed cerebrospinal fluid (CSF) pulsatile movement between the intraspinal and intracranial subarachnoid spaces. The cyst wall was fenestrated with a neuroendoscope. The presyrinx state and the CSF movement improved. Posterior fossa arachnoid cyst, as well as Chiari malformation, can cause CSF flow disturbance at the craniocervical junction and syringomyelia. Endoscopic fenestration is less invasive than foramen magnum decompression and should be the procedure of choice.

Key words: arachnoid cyst, syringomyelia, fenestration, neuroendoscope

Introduction

Syringomyelia is caused by disturbance of cerebrospinal fluid (CSF) movement at the craniocervical junction in patients with Chiari malformation and other etiologies.1,5,10,14) However, patients with syringomyelia that can be treated by posterior decompression do not always have tonsillar herniation.6) We treated a patient with syringomyelic myelopathy, without tonsillar herniation but with a posterior fossa arachnoid cyst, by endoscopic fenestration.

Case Report

A 32-year-old man had undergone cyst-peritoneal (CP) shunting at age 3 years because of posterior fossa arachnoid cyst (Fig. 1). He was hospitalized in our ward again because of paresthesia of both hands, finger movement clumsiness, and gait disturbance. The deep tendon reflexes were exaggerated in both legs. The symptoms indicated cervical myelopathy at the C4–T1 levels. The CP shunt was still functioning.

Cervical radiography showed the anteroposterior diameter and interpedicular distance at C-4 were 12 mm and 22 mm, respectively, and less than the lower limits in a normal adult. Magnetic resonance imaging showed edematous enlargement and T2 prolongation of the cervical spinal cord, indicating a “presyrinx” state.
(MR) imaging showed the cervical spinal cord was enlarged, with T2 prolongation. These neurological and radiological findings indicated a presyrinx state. Constructive interference in steady state (CISS) imaging demonstrated the inferior wall of the cyst as concave, indicating that the CSF pressure in the major cistern was higher than that in the cyst (Fig. 2 left). Cine MR imaging showed no CSF pulsatile movement at the cranioventricular junction. The wall of the cyst disturbed CSF movement between the intraspinal and intracranial subarachnoid spaces.

Endoscopic surgery was performed. A neuroendoscope was inserted directly into the cyst through the midline suboccipital burr hole. The cyst wall surrounded the medulla, and fenestration of the wall was achieved on the right (Fig. 3). On the day after surgery the hypesthesia of the hands and the finger clumsiness had improved. Several days after surgery, the patient was able to walk and go up and down stairs without difficulty. Postoperative CISS imaging did not show the cyst wall, possibly because of increased wall motion after the fenestration (Fig. 2 right). Cine MR imaging showed CSF pulsatile movement between the cyst and the major cistern.

Discussion

MR imaging may show edematous enlargement of the cervical spinal cord and T2 prolongation in some patients before the formation of syringomyelia. "Presyrinx" state is used to describe this pathology.3) The presence of a presyrinx state or syringomyelia suggests CSF flow disturbance at the cranio-

cervical junction.

Several mechanisms have been proposed to explain the relationship between syringomyelia and CSF flow disturbance at the cranioventricular junction.5,10,14) Under normal conditions, CSF in the spinal subarachnoid space moves by pulsatile movement to the intracranial subarachnoid space and by influx to the spinal parenchyma.11) Disturbance of CSF movement into the intracranial space, together with obstruction of the central canal, results in accumulation of CSF in the spinal parenchyma, leading to syringomyelia formation. Rapid downward movement of the tonsil in the cardiac systolic phase exaggerates the spinal pulse wave and forces CSF into the spinal cord.3)

In our patient, the arachnoid cyst occupied the posterior fossa. The major cistern was so small that CSF movement between the spinal and intracranial subarachnoid spaces was limited. The wall of this arachnoid cyst had the same function as the herniated tonsil in Chiari malformation in disturbing CSF movement at the cranioventricular junction (Fig. 4 left). Two cases of syringomyelia were caused by invagination of an arachnoid cyst into the cervical spinal canal.13) Our case indicates that CSF flow distur-
bance at the craniocervical junction can occur even if cyst invagination is absent.

Foramen magnum decompression was not necessary to improve CSF flow disturbance in our patient. Fenestration of the inferior wall of the arachnoid cyst allowed the CSF in the spinal subarachnoid space to move freely into the intracranial space. The overload of CSF against the spinal parenchyma was reduced, and the presyrinx state improved (Fig. 4 right). The syringomyelia improves with reduction of the trans-spinal CSF flow (small arrows).

Our patient had spinal canal stenosis, which appeared to be a complication of long-term CP shunting.3) Canal stenosis can cause syringomyelia3) or can manifest as syringomyelic symptoms.7) The presyrinx state may have been affected by the canal stenosis and could have been treated by laminectomy. However, endoscopic fenestration is minimally invasive and should be the procedure of choice for the treatment of a posterior fossa arachnoid cyst presenting with syringomyelia, provided that a target wall is detected. In our patient, the inferior wall of the cyst was detected preoperatively by CISS imaging. The patency of the fenestrated channel was evident in the CSF pulsatile movement revealed by postoperative cine MR imaging. CISS and cine MR imaging2,4,12) are useful tools for pre- and postoperative examination.

References


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