Spinal Cord Edema Preceding Syringomyelia Associated With Chiari I Malformation
—Case Report—

Yasunobu ITOH, Naoyuki KUWAHARA, Toshio SASAJIMA, Kazuo MIZOI, and Jun HATAZAWA*

Department of Neurosurgery, Akita University School of Medicine, Akita; *Department of Nuclear Medicine and Radiology, Research Institute for Brain and Blood Vessels-Akita, Akita

Abstract

A 38-year-old woman with Chiari I malformation presented with spinal cord edema preceding syringomyelia manifesting as a 5-month history of nuchal pain and numbness of the upper extremities. Magnetic resonance imaging showed spinal cord edema, a poorly defined syrinx at the C-2 to T-2 levels, and distorted cerebellar tonsils. Computed tomography revealed cerebrospinal fluid (CSF) density in the center of spinal cord edema, and positron emission tomography revealed no uptake of L-[methyl-11C]methionine, indicating a non-neoplastic lesion. Craniocervical decompression achieved excellent clinical and neuroradiological outcomes. The success of surgical treatment supports the theory that patients with Chiari I malformation have increased transmural flow of CSF, causing spinal cord edema that progresses to syringomyelia. Early treatment of patients with spinal cord edema is indicated to prevent permanent spinal cord injury due to progressive syringomyelia.

Key words: Chiari I malformation, foramen magnum decompression, magnetic resonance imaging, positron emission tomography, spinal cord edema, syringomyelia

Introduction

The pathogenesis of syringomyelia associated with Chiari I malformation is controversial. Syringomyelia may be caused by the forceful diversion of cerebrospinal fluid (CSF) from the fourth ventricle into the central canal at the obex.3,4,14,15) The syrinx results from obstruction of the central canal byependymal hyperplasia occluding the rostral flow of CSF in the central canal.8,10) Therefore, syringomyelia associated with Chiari I malformation should arise from expansion of the central canal. However, neuroradiological and pathological studies rarely reveal a patent central canal in adult patients with syringomyelia, so passage of CSF from the fourth ventricle to the syrinx is unlikely to be an important mechanism of syrinx development in adults.8,10,15) Syringomyelia could also be caused by the pulsatile elevations of spinal venous pressure or pulse pressure in the spinal subarachnoid space which propel the CSF through the Virchow-Robin spaces surrounding the vascular structures of the spinal cord

and the extracellular space of the spinal cord11,13) and into the central canal.1,12) The cerebellar tonsils act as pistons in the partially enclosed cervical subarachnoid space, exaggerating the pulsatile systolic pulse waves in the spinal subarachnoid space and driving the CSF through the anatomically continuous perivascular and interstitial spaces into the central canal of the spinal cord.11,13) In this case, spinal cord edema should precede the development of a syrinx cavity.1,6,12) However, this intermediate stage in the development of a syrinx has not been well documented in patients with syringomyelia associated with Chiari I malformation.

We treated a patient with Chiari I malformation in whom neuroradiological assessment revealed spinal cord edema and a poorly defined syrinx.

Case Report

A 38-year-old woman presented with a 5-month history of dysesthetic pain of the upper extremities, which was initially noted in her left hand and progressed gradually to involve her other hand, the bilateral upper extremities, and shoulders. Treat-
Fig. 1 Sagittal T₁- (A) and T₂-weighted (B) magnetic resonance images showing spinal cord edema, a poorly defined syrinx between the C-2–T-2 levels, and distorted cerebellar tonsils.

Fig. 2 (A) Computed tomography (CT) scan revealing a low density area in the cervical spinal cord. (B) CT values plotted along the white line in Fig. 2A showing the density of cerebrospinal fluid in the center of the spinal cord.

Fig. 3 Positron emission tomography scan of the cervicothoracic spine showing no uptake of L-[methyl-11C]methionine, indicating a non-neoplastic lesion.

Neurol Med Chir (Tokyo) 42, September, 2002
Fig. 4 Three-dimensional computed tomography scan 6 months postoperatively showing the curved hydroxyapatite implant over the decompressed foramen magnum (arrow) and laminectomy of the C-1 posterior arch.

Fig. 5 Sagittal T1- (A) and T2-weighted (B) magnetic resonance images 1 year postoperatively revealing resolution of the spinal cord edema and the poorly defined syrinx, and the normal morphology of the cerebellar tonsils. Arrow indicates the hydroxyapatite plate implanted over the decompressed foramen magnum.

Ltd., Tokyo) (Fig. 4). The bilateral cerebellar tonsils were visible through the arachnoid and the inner layer of the dura. Intraoperative ultrasonography confirmed pulsatile to-and-fro movement of the cerebellar tonsils after resection of the outer layer of the dura.

The patient made an uneventful recovery and was discharged from the hospital with improvement in the upper extremity pain. Follow-up evaluation at 3, 6, and 12 months revealed excellent improvement in sensation and motor functions. MR imaging of the spine showed that the spinal cord edema and the poorly defined syrinx had resolved by 3 months after surgery. In addition, the cerebellar tonsils had returned to a normal shape and the dorsal hump of the medulla at the point of tonsillar impaction was resolved (Fig. 5).

Discussion

Spinal cord edema in the present case was consistent with the transmural passage of CSF from the spinal subarachnoid space into the spinal cord. Craniovascular decompression and resection of the outer layer of the dura resulted in complete resolution of the spinal cord edema and poorly defined syrinx, suggesting that a block in the CSF pathway at the foramen magnum was the primary cause of the spinal cord edema.

Previous evidence has indicated that spinal cord edema and syrinx fluid originate from the CSF in the spinal subarachnoid space. CT myelography of patients with syringomyelia has shown that contrast medium can pass from the subarachnoid space to the syrinx,7) and syrinx fluid obtained during syrinx-subarachnoid shunting procedures in patients with syringomyelia had identical chemical composition to the CSF. Experimental studies have shown that subarachnoid CSF can enter the spinal cord through the Virchow-Robin spaces and that these spaces are enlarged in patients with syringomyelia.1,5) CT in this case revealed CSF density in the center of the spinal cord edema.

A “presyrinx” state of spinal cord edema has been proposed in patients with nontraumatic obstruction of the CSF pathways.3) Serial MR imaging has documented progression of spinal cord edema to a syrinx without treatment.6) Therefore, spinal cord edema may be a transient condition before rapid progression to a syrinx.6) Craniovascular decompression in patients with syringomyelia and Chiari I malformation will relieve the obstruction in the CSF flow and result in resolution of the syrinx, but neurological recovery is unlikely to be completely satisfactory.6) Thus, early treatment should be considered for spinal cord edema in a patient with Chiari I malformation, because subsequent syrinx formation will lead to permanent neurological deficits as a result of irreversible spinal cord injury due to

Neurol Med Chir (Tokyo) 42, September, 2002
progressive syringomyelia.

References


Address reprint requests to: Y. Itoh, M.D., Department of Neurosurgery, Akita University School of Medicine, 1–1–1 Hondo, Akita 010–8543, Japan.
e-mail: yasuitoh@nsg.med.akita-u.ac.jp.