Cerebellopontine Angle Meningioma Causing Asymptomatic Syringomyelia

—Case Report—

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Abstract

A 36-year-old female was admitted with a 3-month history of headache and gait disturbance. Magnetic resonance imaging demonstrated a large tumor in the right cerebellopontine angle and syringomyelia in the upper cervical cord associated with caudal displacement of the cerebellar tonsil. Complete removal of the tumor resulted in disappearance of these associated conditions. Herniation of the cerebellar tonsil and distortion of the brain stem had probably caused disturbance of cerebrospinal fluid flow, which combined with obstruction of the spinal canal, caused the syrinx.

Key words: syringomyelia, posterior fossa, brain tumor, meningioma

Introduction

Syringomyelia is well-known to be associated with anomalies in the craniocervical junction. The combination of syringomyelia and posterior fossa tumor is rare.^{3,4,6-10,13)} We describe a case of syringomyelia associated with cerebellopontine angle tumor and discuss the pathogenesis based on neuroimaging findings.

Case Report

A 36-year-old female was admitted with complaints of headache and gait difficulty persisting for the last 3 months on our clinic. Neurological examination revealed left cerebellar ataxia with nystagmoid ocular movement, but no evidence of sensory disturbance or motor weakness in the upper trunk and arms. Computed tomography showed a high density lesion in the right cerebellopontine angle with mild ventriculomegaly. Magnetic resonance (MR) imaging with gadolinium-diethylenetriaminepenta-acetic acid also demonstrated a homogeneously enhanced lesion in the right cerebellopontine angle with marked shift of the fourth ventricle. Sagittal MR imaging indicated a peg-like deformity of the cerebellar tonsil

with syringomyelia of the upper spinal cord (Fig. 1).

After cerebrospinal fluid (CSF) drainage via a burr hole placed in the occipital region, the tumor attached to the posterolateral pyramidal wall was completely removed. Histological examination showed the large posterior fossa mass was a meningioma.

Fig. 1 left: Preoperative magnetic resonance (MR) image demonstrating a right cerebellopontine angle lesion with marked shift of the fourth ventricle. right: Sagittal MR image showing peg-like deformity and herniation of the cerebellar tonsil (asterisk) with syrinx (arrowheads), but no communication between the syrinx and the fourth ventricle.

Fig. 2 left: Postoperative magnetic resonance (MR) image showing diminishing syringomyelia with improved tonsillar herniation. right: MR image showing complete removal of the tumor.

The postoperative course was uneventful. MR imaging taken 3 months after the surgery found no caudal displacement of the lower cerebellum or evidence of syringomyelia (Fig. 2). She is now working as a housewife without neurological deficit.

Discussion

Neuroimaging of the present case revealed caudal displacement of the lower cerebellar tonsil with the same configuration as that of Chiari type I malformation. However, removal of the tumor caused the cerebellar tonsil to return to its normal position. This observation suggests that the cause of syringomyelia must be an anatomical change around the craniocervical junction.

Several theories have been proposed to explain the development of syringomyelia. Failure of the foramina of the fourth ventricle to open with continuing communication between the fourth ventricle and the cystic space within the spinal cord *via* the obex may allow increased pressure within the ventricles to be transmitted to the central canal.⁵⁾ CSF pressure waves can cause forced flow of the CSF into the syrinx along the Virchow-Robin spaces.²⁾ Partial blockage of the subarachnoid space in the region of the cisterna magna may direct CSF into the communication, providing an intermittent distending force which may be active for many years.¹⁵⁾ Obstruction at the cisterna magna associated with a high venous pressure can lead to transmedullary passage of CSF which could create a syrinx cavity.¹⁾ Excessive absorption of CSF from the spinal cord might cause Chiari type I malformation, leading to a foraminal obstruction and ultimately producing a

syrinx.¹⁴⁾

Recently, a detailed analysis of the configuration of the central canal in the normal population as well as in syringomyelia patients showed that a large portion of the normal group has an obstructed central canal except in their early stages of life.^{10,11)} The spinal cord with syrinx shows three patterns of communication with the fourth ventricle and central canal: central canal syrinx (communicating), central canal syrinx (non-communicating), and extracanalicular syrinx. Central canal syrinx (communicating) is observed predominantly in children with hydrocephalus. Central canal syrinx (non-communicating) has a cavity consisting of a focal dilation of central canal that is separated from the fourth ventricle by a syrinx-free segment of spinal cord and occurs predominantly in adult patients with various diseases which cause CSF circulatory disturbance around the cervicomedullary junction. Extracanalicular syrinx is seen as a result of spinal cord injury.

MR images of patients with brain tumors which demonstrate syrinx may suggest that the central canal has already been occluded in some locations. Phase-contrast/cine MR imaging indicates that a disturbance of CSF circulation in the spinal subarachnoid space may cause fluid to be forced into the central canal through the interstitial space of the spinal cord in such cases.¹¹⁾

Syringomyelia may occur secondary to the brain tumor within the posterior fossa but without symptoms suggestive of syringomyelia.^{3,4,6-9,12,13,16)} Sagittal MR imaging of our case showed a large and slow-growing brain tumor, which had resulted in elimination of CSF from the posterior fossa and herniation of the cerebellar tonsil through the foramen magnum. Herniation of the cerebellar tonsil and dis-

Fig. 3 Possible pathogenesis of syringomyelia due to infratentorial brain tumor. CSF: cerebrospinal fluid.

tortion of the lower brain stem may have disturbed CSF circulation in the spinal subarachnoid space, and resulted in transmedullary passage of CSF. Furthermore, the obstructed central canal may also have prevented free passage of CSF outside the central canal. Consequently, these pathological CSF flows and accumulations caused a compartment of the central canal force to dilate. The symptoms of brain tumor in the posterior fossa, as with all cases with tonsillar herniation, are too severe to remain untreated. Therefore, the cavitation of the spinal cord will not grow large enough to cause neural dysfunctions (Fig. 3).

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