

Syringomyelia as a Sequelae of the 4th Ventricular Dilatation from Traumatic Hydrocephalus and Cerebellar Atrophy

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A 32-year-old man with stuporous mental state was transferred to our hospital emergency room after a car accident. The brain computed tomography(CT) showed 4th ventricular hemorrhage. He woke up 2 weeks after admission and then discharged. However, he returned to the hospital 10 months after discharge because of both shoulder pain and weakness of both arms. His brain CT showed marked dilatation of the 4th ventricle. His MRI showed whole spinal syringomyelia without Chiari malformation. The patient then underwent ventriculo-peritoneal shunt. His symptoms dramatically improved on the immediate postoperative day, and the syringomyelia also disappeared after operation. The authors report a very rare case of syringomyelia that was developed as a sequelae, especially of the 4th ventricular dilatation without intracranial pressure elevation after traumatic hydrocephalus and cerebellar atrophy, favoring Gardner's hypothesis.

KEY WORDS : Syringomyelia · 4th ventricular dilatation · Ventricular-peritoneal shunt · Gardner's hypothesis.

Introduction

The pathophysiologic theories previously proposed for the progression of syringomyelia are controversial, and complicated mechanisms have been suspected to contribute to the pathogenesis of syringomyelia. A most widely accepted view is that cavitation occurs when cerebrospinal fluid (CSF) within the fourth ventricle is forced into the central canal of the spinal cord, either as a consequence of a caudally directed CSF pulse wave (Gardner's hypothesis), or the development of the CSF pressure gradient at the level of the foramen magnum (William's hypothesis)^{5,12,18,19}. The causative factors of syringomyelia are quite heterogeneous, for example, Chiari I malformation, posterior fossa tumor, hydrocephalus, spinal cord tumor, trauma, arachnoiditis, etc.^{7,11,13,14,16,17}.

The "hydrodynamic theory", initially proposed by Gardner and colleagues^{5,12} and the "cranial-spinal pressure dissociation hypothesis", originally formulated by Williams^{18,19}, have been proposed as the two principal theories. Ventricular

dilatation and Chiari malformation have been one of the well-known conditions for Gardner or Williams hypothesis, but isolated conditions favoring only one hypothesis is very rare. Syringomyelia favoring Gardner's hypothesis as a sequelae of the isolated normo-pressure 4th ventricular dilatation after traumatic cerebellar hemorrhage and 4th ventricular hemorrhage is not yet reported in previous literature.

In this paper, the authors report a case of syringomyelia as a sequelae of the isolated normo-pressure 4th ventricular dilatation without Chiari malformation, after traumatic cerebellar hemorrhage and 4th ventricular hemorrhage and which support the "hydrodynamic hypothesis" of Gardner.

Case Report

A 32-year-old man without specific medical history who presented with stuporous mental state was admitted to our hospital via the emergency room after a car accident. At admission, vital signs were unstable and a state of shock developed. An initial brain computed tomography(CT) showed cerebellar hemorrhage around the 4th ventricle, 4th ventricular hemorrhage, and generalized brain swelling (Fig. 1A, B). He had hemoperitoneum and a left femur shaft fracture for which an emergency operation was performed.

- Received : April 16, 2004 • Accepted : June 18, 2004
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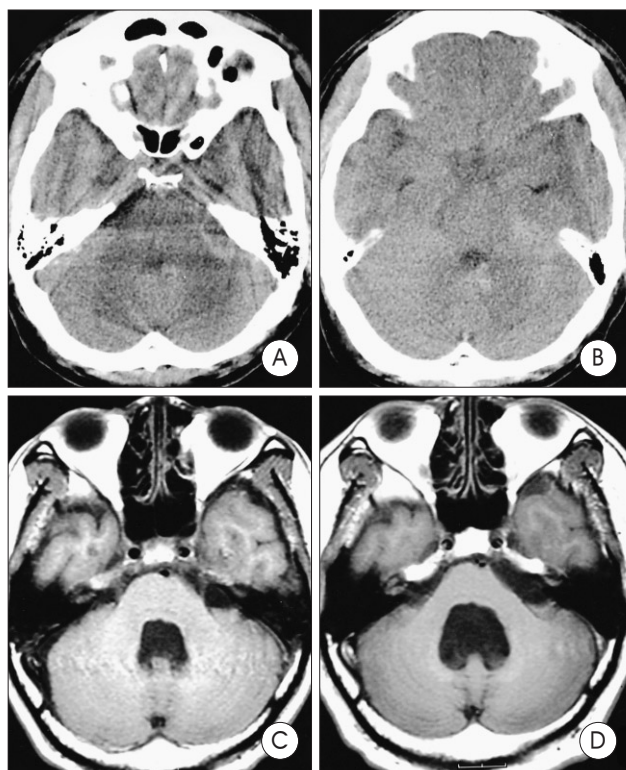


Fig. 1. A, B : Initial pre-operative brain computed tomography shows the 4th ventricular hemorrhage and cerebellar hemorrhage with swelling. C : Four week later, axial view of T1-weighted brain magnetic resonance (MR) image shows the 4th ventricular dilatation. D : At 11 months, brain MR image shows marked 4th ventricular dilatation from cerebellar malacia and mild hydrocephalus.

His brain swelling and hemorrhage was managed by conservative treatment. He woke up 2 weeks after admission with mild quadriparesis, and was discharged with more improvement 4 weeks after admission.

The patient returned to the hospital 10 months after discharge because of both shoulder pain and weakness of both arms, especially of the right side. Follow-up brain magnetic resonance (MR) image and cervical spine MR image showed cerebellar atrophy, fourth ventricular dilatation (Fig. 1C, D), and syringomyelia of the whole spine without Chiari malformation (Fig. 2A). The maximum diameter of the syringomyelia was estimated to be 8mm. To evaluate the CSF flow dynamics and intracranial pressure, we performed radioisotope cisternography (RIC) and ICP monitoring. The RIC showed normo-pressure communicating hydrocephalus, and the spinal pressure was 115mm H₂O (Fig. 3). The range of ICP during the ICP monitoring was normal, from 100mm H₂O to 150mm H₂O, and there were no abnormal waves. The patient was treated by a ventriculo-peritoneal shunt (V-P shunt) (Fig. 2B, C). He showed rapid clinical improvement on the immediate postoperative day. Postoperative cervical

spine MR image showed remarkably decreased syringomyelia size, from the pre-operative 8mm in maximum diameter to 3mm within 2 weeks after the operation (Fig. 2D).

The follow-up cervical spine MRI at 6 months and 10 months after shunt operation showed that the syringomyelia had near completely disappeared to 1mm in diameter. The patient eventually recovered from the previous symptoms completely.

Discussion

Syringomyelia is a malformation marked by the presence of a fluid-filled cavity dilatation within the spinal cord. Several theories on the pathogenesis of syringomyelia have been put forward, but is still a subject of debate. Especially in the hind-brain pathology associated cases, tonsillar herniation is a cause of foramen magnum obstruction. And such tonsillar herniation is developed from various conditions such as Chiari malformation, arachnoiditis, posterior fossa tumorous condition, and craniocervical junction malformation^{7,11,13,14,16,17}. The CSF flow dynamic dysfunction due to foramen magnum obstruction was mainly related formation and progression of syringomyelia^{5,11,18}. But report on mechanisms regarding syringomyelia from foramen magnum obstruction or CSF flow dynamic dysfunction are controversial.

The two main theories that are usually accepted are the hydrodynamic theory of Gardner, and the CSF pressure dissociation theory of Williams. Gardner's hypothesis relates the persistence and enlargement of the central canal to the conservation of a communication between the fourth ventricle and the central canal of the spinal cord through the obex by the pulsatile water-hammer effect^{5,12}.

In contrast, Williams focused attention on the CSF pressure dissociation that arises between the intracranial and vertebral compartments as a result of the sudden rises in pressure that occur during coughing, sneezing, and the Valsalva maneuver, all of which initially cause a rise in intravertebral pressure, which is transmitted to intracranial pressure. When the perimedullary venous pressure returns to normal, the mechanism is reversed, with the return of the pressure wave from the intracranial to the spinal compartment. The presence of an obstruction to the drainage of CSF at the foramen magnum, as in Chiari malformation or arachnoiditis, gives rise to a pressure dissociation between the intracranial and spinal compartments, the CSF being sucked into the central canal with the formation of a syringomyelia^{18,19}. According to the above two main theories, actually the patent central canal or patent obex is indispensable^{4,13}. The patent central canal

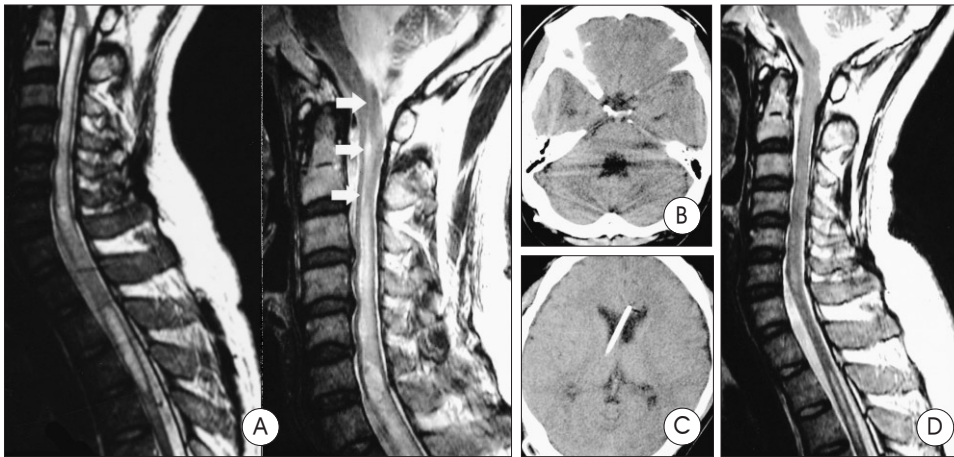


Fig. 2. A : Pre-operative sagittal cervical T2-weighted magnetic resonance(MR) image shows cervicothoracic syringomyelia and communication between the 4th ventricle and syringomyelia (white arrow). B, C : Post-operative brain computed tomography shows decreased 4th ventricle dilatation and shunt proximal catheter. D : Sagittal cervical T2-weighted MR image obtained 2 weeks postoperatively. The syringomyelia has nearly completely disappeared.

was in part supported by laboratory studies performed by Becker, et al., who demonstrated that kaolin-induced hydrocephalus resulted in marked dilatation of the central canal². Other authors have argued that because the obex frequently does not communicate with the central canal, this theory cannot adequately account for all syrinx cavity formations. Dayan focused on the disturbance of intraspinal CSF flow to the foramen magnum, and suggested that increased intraspinal pressure from local blockage of intraspinal CSF flow permits CSF flow shifting into the spinal cord through the Virchow-Robin spaces along the transparenchymal vessels¹. The CSF flow shifting into the spinal cord through the Virchow-Robin space was demonstrated by intraoperative ultrasonography and phase-contrast MRI of syringomyelia in the Oldfield study¹¹, and by the movement of water-soluble contrast in the Dubois report³.

The exact function of the central canal as an ependymal-

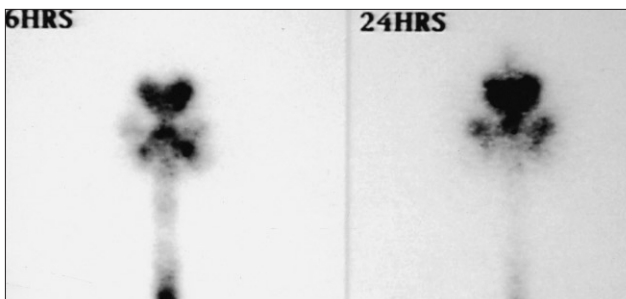


Fig. 3. The preoperative radioisotope cisternography shows communication between the 4th ventricle and syringomyelia without obstruction with normal pressure.

lined structure from the fourth ventricle to the conus medullaris in the spinal cord remains unclear^{4,15}. Milhorat and Storer presumed that it serves in a "sink-like" capacity, clearing waste products from the spinal cord^{10,15}. The central canal is patent in prenatal life and it undergoes age-related stenosis. After all it is obliterated during adulthood^{9,6}. But Holly, et al. demonstrated the persistent central canal by review of their experience in diagnosis and management of 32 patients

with slitlike syrinx cavities⁴. Also, Milhorat, et al. found three types of syrinxes-communicating central canal syrinxes, non-communicating central canal syrinxes, extra-canalicular syrinxes-in an analysis of 105 autopsy cases⁸. According to his opinion, patent central canal is one of the pathological basis of syringomyelia formation^{8,10}.

In our case, fourth ventricle hemorrhage and cerebellar swelling was caused by trauma which progressed to cerebellar atrophy and mild hydrocephalus, resulting in marked dilatation of the fourth ventricle. The fourth ventricular dilatation permitted large pulsatile pressure of CSF to be transmitted into the central canal, that developed in to canal dilatation in the spinal cord, progressing to a longer and larger size. In the previous description, it is difficult to conclude which theory, Gardner's or William's hypothesis, is more compatible with syringomyelia formation, because all syringomyelia had partial CSF flow obstruction at the foramen magnum. Because there was no CSF flow obstruction at the foramen magnum in our case, pulsatile water-hammer effect to the central canal of Gardner's hypothesis should be considered the driving force of the syringomyelia formation. The 4th ventricle was more predominantly dilated, the pressure of ventricle wall tension was very highly surged. This mechanism was explained by the law of Laplace. The elevated pulsatile pressure of ventricle wall tension was transmitted to central canal. And syringomyelia was developed. This hypothesis is supported by direct connection between the fourth ventricle dilatation area and syrinx, demonstrated by the cervical MRI and by CSF flow connection to the syrinx shown in the RIC.

Treatment of syringomyelia are very multifarious. Many neurosurgeons attempt many different kinds of treatment modality, for example, syringomyelotomy, various syringomyelia shunting, posterior fossa decompression, duroplasty, adhesion lysis, and etc. For treatment of our case, we supposed that only dissolution of the pulsatile water-hammer effect from the fourth ventricle to the central canal may be an effective treatment modality for syringomyelia. Therefore, we performed a V-P shunt, which improved the symptoms rapidly. Also, the syringomyelia dramatically disappeared in follow-up imaging studies 2 weeks after the operation.

In our case, there was no cerebellar tonsillar herniation and the syringomyelia was disappeared after CSF flow pressure diversion without other treatment modality. These finding also support our hypothesis favoring Gardner's theory.

We present a case of syringomyelia after traumatic hydrocephalus and cerebellar atrophy which supports the Gardner's theory.

Conclusion

The authors reported a very rare case of syringomyelia that developed as a sequelae, especially of the 4th ventricular dilatation without Chiari malformation, and intracranial pressure elevation after the traumatic hydrocephalus and cerebellar atrophy favoring Gardner's hypothesis that pulsatile water-hammer effect causes syringomyelia. This case was treated by a V-P shunt subsequently resulting in complete improvement.

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