CASE REPORT

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Acquired Chiari Malformation

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Perioperative lumbar drainage of cerebrospinal fluid is commonly used in neurosurgical practice. However, the relationship between lumbar drainage and acquired Chiari malformation is not well established. The authors present an unusual case of paraplegia as a result of acquired Chiari malformation after lumbar drainage. Acquired Chiari malformation can induce compression of cervicomedullary junction and syrinx formation. Foramen magnum decompression is recommended for the solution of such problems.

KEY WORDS : Arnold-Chiari malformation · Iatrogenic disease · Cerebrospinal fluid · Drainage.

Introduction

The perioperative use of lumbar drainage of cerebrospinal fluid(CSF) is a relatively common procedure in neurosurgical practice. The development of acquired Chiari malformation after lumbar drainage is not well aware of. The incidence is extremely low, but its neurological sequelae might to be very severe and critical[2].

Case Report

A 35-year-old woman with huge sellar and suprasellar mass underwent TSA with partial removal of tumor (Fig. 1A). Intraoperatively bleeding was severe and arachnoid membrane was ruptured. Postoperatively, lumbar drainage of CSF was started in order to prevent CSF leakage. However, on the fourth day after surgery, CSF rhinorrhea was noted, and high fever with neck stiffness developed. Lumbar drainage (5~8cc/hr) was continued and antibiotics treatment was started. On the seventh day after surgery, lumbar drain was revised. On the ninth day after surgery, lumbar drain was again revised at 6PM. Opening pressure was 25cmCSF. At 8PM, patient complained of four extremities tingling sense and pain. At 10PM, her condition was rapidly deteriorated to paraplegic state. Neurological examination showed hypesthesia below T12 dermatome, decreased anal tone, and decreased knee and ankle jerk. Brain CT revealed diffuse brain swelling and crowding of the foramen magnum. Lumbosacral spine MR images revealed contrast enhancement in the anterior and posterior surface of the spinal

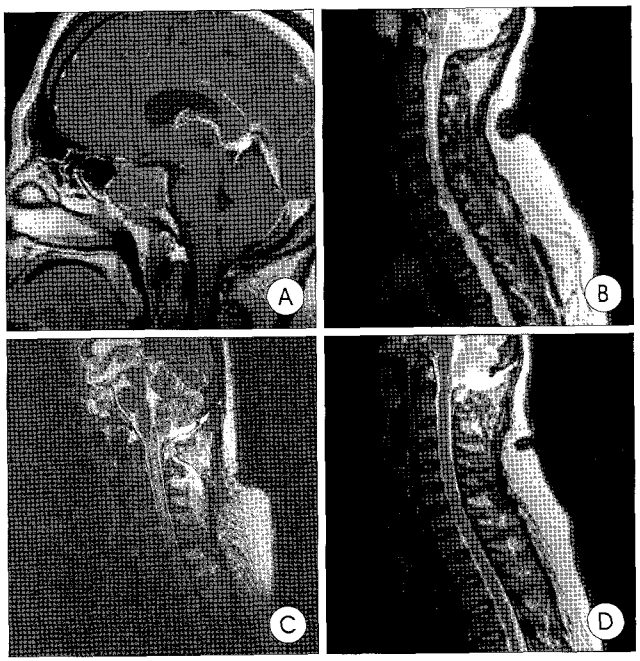

Fig. 1. Preoperative, sagittal, gadolinium-enhanced, magnetic resonance image(MRI) demonstrated a huge sellar and suprasellar mass (A). Sagittal T2 weighted cervicothoracic MRI showed tonsilar descent and syrinx formation (B). On 2 days after foramen magnum decompression(FMD), Sagittal T2 weighted cervical MRI showed much decrease of syrinx (C). On 18 days after FMD, Sagittal T2 weighted cervicothoracic MRI demonstrated complete decompression of cervicomedullary junction and disappearance of syrinx (D).

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cord and nerve root, suggesting leptomeningeal inflammation. There was no epidural abscess or hematoma causing cord compression. Subsequent cervicothoracic spine MR images showed syrinx formation at C2-T8 level and tonsilar descent (about 5mm below the foramen magnum) (Fig. 1B). Under the impression of acquired Chiari malformation, the patient underwent emergent foramen magnum decompression(FMD). Postoperatively her weakness was gradually improved. Follow up cervical spine MR images showed also gradual resolution of syrinx (Fig. 1C). On the 18th day after FMD, her weakness was improved to motor power grade IV+. Cervicothoracic spine MRI showed complete subsidence of syrinx and complete decompression of cervicomedullary junction (Fig. 1D). On the 54th day after FMD, her weakness was completely improved to motor power Grade V. Voiding function was also normalized.

Discussion

The acquired Chiari malformation is radiologically indistinguishable from congenital one[6,8,10]. Chiari I malformation is defined as a caudal descent of the cerebellar tonsil more than 5mm below the foramen magnum. It was generally not associated with caudal descent of the brainstem, and hydrocephalus is uncommon. The pathogenesis of Chiari I malformation involves cephalocranial disproportion attributable to underdevelopment of the posterior fossa and the development of a craniospinal pressure gradient[2,7,8,10]. Cephalocranial disproportion theory can not account for adult patients whose skull growth is arrested. A craniospinal pressure gradient can develop naturally and iatrogenically[2,9,11]. Circumstances in which the normal cephalic absorptive pathways are disrupted, with maintenance of absorptive pathways in the spine, may lead to a downward pressure gradient. Similar circumstances can occur iatrogenically by lumbar puncture or drainage[10], lumboperitoneal shunts[4,7-9], or spontaneous spinal CSF leakage[2].

The authors postulate that continuous lumbar CSF drainage resulted in acquired Chiari malformation. Other factors, that might be operative in causing downward displacement of the cerebellar tonsil, could be postoperative diffuse brain swelling and disrupted CSF absorption by postoperative meningitis in this patient. Once a negative craniospinal pressure gradient developed, tonsilar descent with concomitant compression of the cervicomedullary junction decreased the normal CSF circulation around the foramen magnum. This hydrodynamic change of CSF circulation caused the cord swelling and the formation of syrinx[3,5]. The mechanism of syrinx formation and resolution arises outside, not inside, the spinal cord. The restoration of CSF flow dynamics by decompressive operation is essential in the treatment of syrinx and cord swelling[3,5]. This patient achieved a good long-term outcome following emergency foramen magnum decompression.

The incidence of symptomatic acquired Chiari malformation from perioperative lumbar drainage is rare[1]. Acquired Chiari malformation, however, can induce catastrophic results such as cardio-pulmonary arrest and/or quadriplegia[2]. Before the lumbar drainage, stringent assessment is necessary to exclude radiological or clinical evidence of a Chiari malformation. In order to prevent and/or detect earlier, intracranial pressure has to be evaluated, and overdrainage should be prevented. And close observation for early symptoms needs to be performed at intensive care unit[2]. Once the acquired Chiari malformation is suspected, the lumbar drainage should be clamped immediately and imaging study of cervicomedullary junction should be followed.

Conclusion

Acquired Chiari malformation can induce compression of cervicomedullary junction and syrinx formation. Decompressive operation is essential in the solution this two problems. Foramen magnum decompression is recommended.

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