Miyazaki Syndrome due to Ventriculoperitoneal Shunt Treatment

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Key words
- Cerebrospinal fluid hypotension
- Miyazaki syndrome
- Overshunting-associated myelopathy
- Ventriculomegaly
- Ventriculoperitoneal shunt

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
CT: Computed tomography
MRI: Magnetic resonance imaging
VP: Ventriculoperitoneal

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BACKGROUND: **The signs and pathomechanism of Miyazaki syndrome is presented through the case of a young female patient.**

CASE DESCRIPTION: **The 33-year-old patient had undergone placement of a ventriculoperitoneal shunt with a pressure-adjustable valve for communicating hydrocephalus years before presenting to our department with the complaints of constant headache and unsteady gait. On the basis of the clinical picture and her history, plain and contrast-enhanced cranial and whole spine magnetic resonance imaging and magnetic resonance angiography examinations were performed, with the scans revealing signs indicative of cerebrospinal fluid hypotension typical of Miyazaki syndrome.**

CONCLUSION: **The article discusses the available literature suggesting the underlying cause in such cases to be the dysfunction of the Starling resistor mechanism due to an improperly adjusted ventriculoperitoneal shunt, which results in excessive cerebrospinal fluid loss accompanied by consequent cerebral venous overflow with vertebral venous engorgement and compressive cervical myelopathy.**

INTRODUCTION

The pathological alterations that make up Miyazaki syndrome were first observed and described by a Japanese neurologist in 1998.[1] Miyazaki found a dilated epidural venous system at the level of the cervical spinal cord accompanied by low cerebrospinal fluid (CSF) pressure and signs of upper cervical myelopathy as a result of venous compression in a patient with ventriculoperitoneal (VP) shunt. Looking for causative associations among these signs, Miyazaki set up a hypothesis regarding the pathomechanism of this syndrome in their pioneering publication,[1] which prompted some of the later reports to use the term Miyazaki syndrome when referring to this constellation. Many case reports have subsequently been published about similar cases; however, with only few of them discussing associations between the presenting signs and the putative pathomechanism of this syndrome.[2-10] Beside Miyazaki syndrome, the condition is commonly referred to as overshunting-associated myelopathy or myelopathy due to CSF overdrainage. In addition to the imaging signs, suspicious symptoms of CSF hypotension include head and neck pain, dizziness, and blurred vision usually orthostatic in nature, with the appearance of ataxia, unsteady gait, and numbness and clumsiness of extremities raising the suspicion of a complicating myelopathy. As a possible therapy, Miyazaki[1] recommended the closure of the shunt, which led to the full recovery of their patient.

CASE REPORT

This is the case of a 33-year-old female patient who was referred to the Department of Neurosurgery as part of a multi-institutional medical assessment of her complaints. Her past medical history was significant for premature birth at 7 months gestation and a birth weight of 1650 g. At the age of 14 years, a cranial computed tomography (CT) was performed for her complaints of headache, which revealed cerebral ventricular dilation. To assess her CSF circulation more accurately, isotope cisternography was carried out, which revealed a communicating hydrocephalus. As a consequence of the isotope study, the patient developed meningitis, from which she recovered seemingly without complications.

At the age of 23 years, her previous occipital-temporal headaches returned, which were usually dull in character but sometimes became very intense. She also complained of an unsteady gait and, according to her family, her memory deteriorated, having difficulties in learning. Her neurological examination revealed mild abnormalities (she had difficulty standing during a sharpened Romberg test). However, based on her clinical signs, intracranial hypertension was suspected. Accordingly, a pressure-adjustable VP Hakim valve system (Codman and Shurtleff Inc., Raynham, Massachusetts, USA) was implanted to the right side. During the mobilization period after the operation, the opening pressure of the valve (140 mm H_2O) proved to be low, as the patient complained of headaches. After elevating the opening pressure (to 160 mm H_2O), her complaints decreased, she became more active, and her unsteadiness started to resolve. In the follow-up cranial CT scan, the intraventricular part of the shunt was in a proper position, and no signs of excessive CSF drainage were detected (Figure 1).

In the following years, her complaints occasionally recurred, but the problems usually disappeared after increasing the opening pressure of her shunt. At the age of 33 years, after carrying heavy luggage during an overseas holiday, she presented with a gradual onset pulling sensation occipitally, accompanied by indefinite visual complaints, perioral paresthesia, and numbness and clumsiness in her extremities. These symptoms were mostly present in an upright position; however, they gradually worsened and became almost persistent over time. This was the moment when the patient sought medical help in our department.

Based on the patient's history and previous medical findings, cranial and whole spine magnetic resonance imaging (MRI) and magnetic resonance angiography examinations were performed. The cranial MRI revealed dilation of the lateral ventricles in a colpocephalic configuration with left-sided predominance. In this region, thinning of the periventricular white matter was observed. The rest of the cerebral ventricles and the external CSF spaces were normal (Figure 2A). The tip of the ventricular catheter located in the right frontal horn was in a proper position. There was no change in the signal intensity in the periventricular area, indicating no CSF resorption secondary to CSF hypertension. Diffuse thickening and intense contrast enhancement of the dura mater were also observed (Figure 3A) together with the dilation of venous sinuses. The surface of the pituitary gland became strongly convex (Figure 3B). These signs were together indicative of CSF hypotension. As an incidental finding, a subependymal heterotopic gray matter mass was noticed adjacent to the left frontal ventricular horn (Figure 2B).

In addition, noncontrast spinal MRI and time-resolved imaging of contrast kinetics (TRICKS) magnetic resonance angiography scans revealed a pronounced dilation of the epidural venous system almost along the whole spine. These dilated vessels narrowed the spinal canal and also compressed the spinal cord predominantly in the cervical region, which was associated with slightly inhomogeneous signal intensity indicating myelopathy (Figure 4).

On the basis of the findings, we established the diagnosis of Miyazaki syndrome (i.e., CSF hypotension developed as a result of VP shunt implantation accompanied by subsequent epidural venous engorgement, leading to spinal cord compression and myelopathy). Accordingly, we recommended the temporary ligation of the VP shunt and potentially its definite closure, depending on the improvement of the patient's condition.

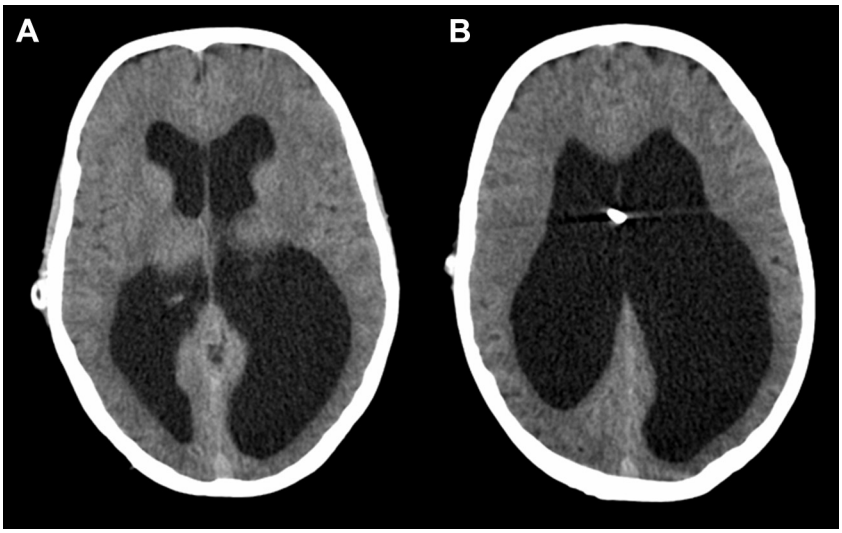

Figure 1. Axial computed tomography scans of the brain with signs of ventriculomegaly in a colpocephalic configuration (**A**), confirming the proper position of the intraventricular part of the ventriculoperitoneal shunt after implantation (**B**).

The patient returned to her physician and, based on our recommendation, the opening pressure of the Hakim valve was increased significantly (to 200 mm H_2O). One month later, as the patient recovered, the valve was replaced (Certas Plus; Codman and Shurtleff Inc., Raynham, Massachusetts, USA), and by further increasing the opening pressure, the valve was basically closed. Beside the replacement of the valve, an antisiphon (Siphonguard; Codman and Shurtleff Inc., Raynham, Massachusetts, USA) device was built into the shunt system to prevent venous overdrainage. The rationale behind the replacement of the old valve with a modern one was to enable a complete closure with supraphysiologic opening pressure levels, yet maintaining the possibility of careful drainage in the future, if it becomes necessary.

After replacement, the condition of the patient showed progressive improvement, and at her 6-month follow-up MRI, the lesions in the cervical spine demonstrated regression (Figure 4C). The valve is still closed with no apparent clinical or radiographic need for reopening at present. The status of the hydrocephalus as the primary indication of the VP shunt placement is virtually unchanged (Figure 5).

DISCUSSION

The various symptoms of CSF hypotension, developed due to excessive drainage through the VP shunt, are well known in the literature (the most common ones include orthostatic headache, cervical pain, and signs of III, IV, and VI cranial nerve palsy) and so are its diagnostic signs (e.g., dural enhancement, rounded pituitary gland, and cerebellar prolapse). However, the subsequent development of epidural venous engorgement leading to spinal cord compression is one of the rare complications of this disease. We found the description of similar signs in a total of 9 previously published case reports. The indication of VP shunt implantation was different in each case; however, the subsequently developed symptoms were similar. A summary of these cases is presented in Table 1. The literature search of the previous reports was conducted in the database of PubMed using the key words CSF hypotension, CSF overdrainage,