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(CASE REPORT)

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Cervical Myelopathy from the Overshunting of Cerebrospinal Fluid: Miyazaki Syndrome

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Abstract

This case report details Miyazaki Syndrome in a patient who is status post right frontal strata ventriculoperitoneal shunt (VP). This condition results from the overshunting of cerebrospinal fluid (CSF), leading to intracranial hypotension and epidural venous dilation (1). Increased fluid within the venous plexus can compress the cervical spine, resulting in myelopathy (1). Neurosurgeons who consistently manage intracranial shunts should be knowledgeable about this uncommon pathology.

Keywords: Miyazaki Syndrome; Ventriculoperitoneal shunt; Intracranial hypotension; Neurosurgery

1. Introduction

Miyazaki Syndrome is a rare myelopathy that results from overshunting of CSF as a complication of ventriculo-peritoneal or ventriculo-atrial shunts (1). The pathophysiology of this condition has not been definitively established in the literature. The most prominent proposed mechanism suggests that CSF pressure changes from over-drainage lead to intracranial hypotension and epidural venous dilation. Venous dilation may result in compression of the spinal nerve roots, leading to the symptoms of Miyazaki Syndrome (2). Miyazaki Syndrome is typically characterized by cervical myelopathy, increased volume in the venous plexus in the cervical spine and the absence of headache and nausea as well as other symptoms of intracranial hypotension (3). Definitive treatment is valve or shunt adjustment (1).

2. Case Presentation

A 72 year-old-female with a history of HTN, DM, and NPH with right frontal strata VP programmable shunt placed on 1/10/2018 presented with headache for several days with associated dizziness, numbness and tingling of extremities. Patient had a 5 year history of these headaches and was started on Doxepin 25 mg in August 2023. CT Head revealed the presence of small, bilateral subdural hygromas. MRI revealed no acute findings and an intact ventriculostomy shunt catheter. Lab results revealed BS>900, Bicarbonate >25, pH of 7.42, negative urine ketones and plasma osmolality of 314. Patient was admitted for HHS.

One week prior to presentation in the ED, the patient underwent an MRI of the cervical spine. MRI revealed prominence of the ventral epidural soft tissues, which represented engorgement of the anterior epidural venous plexus in the cervical spine. This contributed to severe narrowing of the thecal sac at the level of C2-C3 and C3-C4 with near-complete effacement of the subarachnoid space around the cervical cord. There was flattening of the left and right ventral surface of the cervical cord at the level of C2 without adjacent cord signal abnormality.

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Neurosurgery consultation revealed decreased rapid finger movements and weakness of the right upper extremity and decreased sensation on the right hand. The patient reported diffuse dysesthesias. Shunt settings were increased from 1.5 to 2.0. Over the next several days, the patient reported gradual improvement in upper extremity symptoms and improvement in hand dexterity and numbness. The patient's headache resolved, and she was able to ambulate in the hallway without issue. Repeat MRI showed improvement in high cervical stenosis due to vascular congestion. Patient was discharged with instructions for follow-up with Neurosurgery.

3. Discussion

This case matches the clinical criteria for Miyazaki Syndrome quite well. Our patient's MRI findings were also consistent with this characterization, demonstrating anterior epidural venous plexus engorgement of the cervical spine, further supporting the conclusion that this patient was enduring an overshunting associated myelopathy (4). The definitive cure for Miyazaki Syndrome is limited to shunt or valve management (1). In this case, our patient had prompt resolution of her symptoms, both clinically and on imaging, following adjustment of shunt settings, which supports our suspicion of Miyazaki Syndrome in this patient.

This case is similar to the other reported cases of Miyazaki Syndrome in that positional headache was absent as a reported symptom. Other cases of Miyazaki Syndrome share this common feature, which is peculiar given that the proposed pathophysiology of this syndrome relies on intracranial hypotension and its downstream effects (5).

Further research is necessary to properly characterize the mechanism by which this syndrome occurs and may help to explain why the classical symptoms of intracranial hypotension are consistently absent on presentation. The Monro-Kellie hypothesis could potentially provide a means for understanding this phenomenon (6).

4. Conclusion

The purpose of this case report was to provide a demonstration of a rare but conceivable pathology known as Miyazaki Syndrome, which results from the overshunting of CSF in patients with VP shunts. The underlying mechanism of action, clinical course, radiologic findings, and treatment are discussed within this case. This will hopefully bring further awareness of this condition to physicians who routinely manage VP shunts and will contribute to the current understanding and characterization of this syndrome.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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