

Recovery of diabetes insipidus after Chiari malformation decompression: a case report

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Abstract

This paper aims to introduce a patient with Chiari type 1 malformation presented with upper extremity pain and diabetes insipidus. After laboratory examinations, we confirmed central diabetes insipidus diagnosis. The patient has undergone posterior fossa decompression. In follow up patients symptoms relieved gradually.

Recovery of diabetes insipidus after Chiari malformation decompression: a case report

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consent statement:

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy

Abstract

This paper aims to introduce a patient with Chiari type 1 malformation presented with upper extremity pain and diabetes insipidus. After laboratory examinations, we confirmed our case's central diabetes insipidus diagnosis. The patient has undergone posterior fossa decompression, tonsilopexy and duraplasty. After six months of follow-up, pain and diabetes insipidus were improved. We introduced a rare presentation of Chiari malformation in this article.

Introduction

CM (Chiari malformation) could have ambiguous presentations, ranging from cervical pain to endocrine complications¹. It has been classified into four groups. In adulthood, type 1 is more common². CM is a complex disorder with extreme diversity in presentation. Chiari type 1 is defined as 5 mm descending cerebellar tonsils inferior to the basion-opisthion line (Mcrae line). It can be accompanied by syrinx or not³.

Case presentation

A 40 years old man came to our clinic complaining of feeling pain in his right hand. He also suffered from polyuria and polydipsia, which caused him difficulty sleeping at night. The symptoms started three months before his admission. Pain in the right upper extremity was progressive.

In the examination, he had an average height and weight (174 cm and 71kg). Stature seems to be normal. Vital signs, including blood pressure, were in the normal range. Force of upper and lower extremities was intact. There was no wasting or atrophy in muscles. DTRs of extremities were in the normal range. Urine volume was more than 5 liters in 24 hours, and urine osmolality was 250 mOsm/kg. Urine-specific gravity was 1008. After the water deprivation test, urine osmolality did not rise significantly, but after desmopressin administration, it increased to 600 mOsm/kg. Central DI was the most compatible diagnosis for polyuria. We also performed a cervical MRI and laboratory examination. MRI showed a tonsillar herniation of 5 mm and a compact posterior fossa (figure1). There was no sign of cord tethering in the whole spine MRI. Sella region was normal in brain MRI. Laboratory examination revealed an elevated serum sodium level confirmed by rechecking (Na: 150 – 155). The endocrine profile was normal. After the nephrology consultation, the patient underwent an abdominal ultrasound examination with no abnormal findings. Due to symptoms and imaging, we did a posterior fossa decompression, tonsilopexy, and duraplasty without C1 laminectomy. The intraoperative image is shown in figure 2.

In one month's follow-up, the pain in the upper extremity was relieved. In three months follow-up, urine frequency was reduced, and he had no complaint of polydipsia. The laboratory findings revealed normal serum Na levels.

Discussion

The exact pathology of CM is unknown. The key to diagnosing this condition is a combination of clinical findings and imaging⁴. Previous literature reported some endocrinologic manifestations of CM, including precocious puberty¹. It seems to be a relation between hypopituitarism and neurologic condition in Chiari patients. We see a higher incidence of Chiari malformation associated with a growth hormone deficiency, but there is no determinant response with growth hormone replacement⁵⁶. It could be related to dynamic disturbances of intracranial or regional pressure on the hypothalamic-pituitary axis. In our case, the patient presented with polydipsia, polyuria, and high serum sodium. We confirmed central DI diagnosis through our laboratory studies. Genetic studies have shown that NFIA haploinsufficiency is associated with both urinary tract and CNS malformations⁷. As we know, CNS malformation can be seen with urinary tract abnormalities. In previous studies, it has been proposed that injury to renal descending sympathetic fibers in cervical cord injury leads to water retention and sodium⁸⁹. A tethered cord may cause the presence of urinary symptoms in CM type 1 with a traction mechanism¹⁰. Our patient did not show any clinical signs of tethered cord, and there was not any sign of tethering in the whole spine MRI. In 128 Chiari cases analysis in 2007, Guo and his colleagues reported 2 cases of diabetes insipidus presentation, but there is no information about improvement after surgery and decompression of posterior fossa¹¹. We did not find any reported case of central DI in CM type 1 patients in the literature. This is the first CM type 1 case reported with confirmed central DI, which has been resolved after decompression surgery.

Conclusion

Chiari type 1 can manifest with different presentations. DI could be one of the rare complications caused by CM type 1. DI responds well to posterior fossa decompression and duraplasty.

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