INTRODUCTION

CM (Chiari malformation) could have ambiguous presentations, ranging from cervical pain to endocrine complications. It has been classified into multiple types and subtypes. In adulthood, type 1 is more common. CM is a complex disorder with extreme diversity in presentation. Chiari type 1 is 5 mm descending cerebellar tonsils inferior to the basion-opisthion line (Mcrae line). It can be accompanied by syrinx or not.

CASE PRESENTATION

A 40-year-old man presented to our clinic with 3 months of radicular, moderate, and intermittent pain in his right arm along with polyuria and polydipsia impairing his sleep. Pain in the right upper extremity was progressive.

In the examination, his BMI was 23.45 (height:174 cm and weight: 71 kg). Stature seems to be normal. Vital signs, including blood pressure, were in the normal range. The strengths of the upper and lower extremities were intact.
Atrophy or wasting of the muscle mass was absent. DTRs of extremities were in the normal range (2+). Urine volume was more than 5 liters in 24 h, and urine osmolarity was 250 mOsm/kg. Urine-specific gravity was 1008 (normal range: 1005–1030). After the water deprivation test, urine osmolarity 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 (Figure 1). There was no sign of cord tethering in the whole spine MRI. The sella region was normal in the brain MRI. Laboratory examination revealed an elevated serum sodium level confirmed by rechecking (Na: 150–155 mEq/L). 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 1 month of follow-up, the pain in the upper extremity was relieved. In 3 months of follow-up, urine frequency was reduced, and he had no complaint of polydipsia. The laboratory findings revealed normal serum Na levels.

3 | DISCUSSION

The exact pathology of CM is unknown. The key to diagnosing this condition is a combination of clinical findings and imaging. Previous literature reported some endocrinologic manifestations of CM, including precocious puberty. It seems to be a relationship 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 replacement. 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. The common differential diagnosis for these findings is diabetes mellitus, nephrogenic DI, central DI (CDI), and psychogenic polydipsia. The majority of CDI cases occur after neurosurgical procedures. We confirmed central DI diagnosis through our laboratory studies. Genetic studies have shown that NFIA haploinsufficiency is associated with urinary tract and CNS malformations. As we know, CNS
malformation can be seen with urinary tract abnormalities. Previous studies have proposed that injury to renal descending sympathetic fibers in the cervical cord leads to water retention and sodium. A tethered cord may cause urinary symptoms in CM type 1 with a traction mechanism. 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 the posterior fossa. We did not find any reported case of central DI in CM type 1 patients in the literature. The current article is the first CM type 1 case reported with confirmed central DI, which has been resolved after decompression surgery.

4 | CONCLUSION

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

AUTHOR CONTRIBUTIONS

Faramarz Roohollahi: Methodology; visualization; writing – original draft; writing – review and editing. Arad Iranmehr: Methodology; writing – original draft. Taher Musavi: Data curation; investigation; writing – original draft. Mohammadreza Hajiabadi: Conceptualization; project administration; supervision; validation; writing – review and editing.

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CONFLICT OF INTERESTS

There is no conflict of interests regarding the authors of this manuscript to declare.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CONSENT

We must mention that written informed consent was obtained from the patient to publish this report in accordance with the journal’s patient consent policy. Also, we have to clarify that all the writers involved in this article are researchers with educational targets in medical universities, and they have no government involvement or any other official representative.

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