JEIMP
The Journal of European Internal Medicine Professionals

Case Report

Dandy Walker Malformation in a Patient Presenting with Hyponatremia

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J Eur Int Prof. Year; 2025, Volume: 3, Issue: 4
Submitted at: 04.05.2025 Accepted at: 26.10.2025 Published at: 27.10.2025

doi 10.5281/zenodo.17450717

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Abstract

Hyponatremia is defined as a serum sodium concentration of less than 135 mmol/L and is a common electrolyte abnormality. The most common causes of hyponatremia following central nervous system disorders are syndrome of inappropriate antidiuretic hormone secretion (SIADH) and cerebral salt wasting (CSW). In this case report, we discuss the diagnosis and treatment of CSW in a 30-year-old male patient with recurrent episodes of hyponatremia, in the context of the literature. CSW develops after central nervous system disorders and is characterized by hypovolemia, low serum osmolality, high urine osmolality, and high urine sodium levels.

Keywords: Dandy-Walker Syndrome, Hyponatremia, Kidney Diseases

INTRODUCTION

Dandy-Walker syndrome, or Dandy-Walker malformation, is a developmental anomaly characterized by an enlarged posterior fossa, cystic enlargement of the fourth ventricle, hypoplasia, and upward rotation of the cerebellar vermis. Affected individuals may experience psychomotor retardation, ataxia, apnea episodes, muscle weakness, occasional muscle spasms, seizures, nystagmus, and macrocephaly (1). The most common central nervous system anomalies associated with Dandy-Walker syndrome include ventriculomegaly, agenesis of the corpus callosum, holoprosencephaly, and encephalocele. Common extracranial anomalies include congenital heart disease, polycystic kidneys, and facial clefts. Less frequently observed anomalies include limb and abdominal wall abnormalities, diaphragmatic hernia, ambiguous genitalia, and fetal growth restriction (2).

Hyponatremia, defined as a serum concentration of less than 135 mmol/L, is most commonly caused by SIADH and CSW in patients with central nervous system disorders. The most commonly reported trigger for CSW is aneurysmal subarachnoid hemorrhage. Conditions leading to SIADH and CSW include head trauma, intracranial or metastatic neoplasms, carcinomatosis or infectious meningitis, subarachnoid hemorrhage, and central nervous system surgery (3). However, in rare congenital CNS malformations such as Dandy-Walker syndrome, CSW may also develop due to altered hypothalamic and autonomic regulation of sodium and water balance.

CASE

A 30-year-old male patient was admitted to a district hospital with complaints of fatigue and was diagnosed with hyponatremia (sodium 123 mmol/L). After receiving treatment, he was discharged. The patient was referred to our clinic due to recurrent hyponatremia. His past medical history was unremarkable (a ventriculoperitoneal shunt had been proposed in childhood but was not performed), and he was married with two children. His brother had a shunt placed, but they were unaware of the diagnosis.

The patient's complaints included fatigue and intermittent seizures for the past 15 days, occurring once or twice a day. Physical examination revealed a blood pressure of 110/70 mmHg, pulse rate of 78 bpm, clear

Table 1. Laboratory findings

Parameter	Reference Range	Admission	Post-treatment
Serum Sodium (mmol/L)	135–145	125	135
Serum Potassium (mmol/L)	3.5–5.0	4.5	4.3
Serum Uric Acid (mg/dL)	3.4–7.0	3.6	4.2
Urine Sodium (mmol/L)	<20 (low volume)	114	58
Urine Osmolality (mOsm/kg)	300–900	High	Normal
Serum Osmolality (mOsm/kg)	275–295	Low	Normal

lung sounds, and no pretibial edema.

Laboratory results showed a serum sodium level of 125 mmol/L, potassium 4.5 mmol/L, glucose 134 mg/dL, uric acid 3.6 mg/dL, urine density 1025, and urine sodium 114 mmol/L. The presence of high urine sodium and osmolality with low serum osmolality suggested hypovolemic hyponatremia consistent with CSW. Brain natriuretic protein (BNP) was 11.2 ng/ml, renin and aldosterone levels were within normal limits, and antidiuretic hormone level was 6.4 pmol/L.

Brain CT showed marked dilation of the ventricular system. The septum pellucidum and corpus callosum were absent. The patient was diagnosed with Dandy-Walker syndrome. A neurology consultation was requested due to epileptic seizures, and the patient was started on levetiracetam. After treatment, no further seizures occurred. Neurosurgery confirmed Dandy-Walker syndrome but did not recommend surgery. Cardiology consultation revealed an ejection fraction of 60% and mild tricuspid insufficiency.

The patient received hydration, which normalized the serum sodium level. Improvement following isotonic saline therapy supported the diagnosis of CSW rather than SIADH. Due to frequent recurrence, sodium chloride tablets were recommended but unavailable; therefore, fludrocortisone 0.1 mg twice daily was initiated. On follow-up, serum sodium levels were found to be 135 mmol/L, and the patient remained asymptomatic under outpatient monitoring.

DISCUSSION

Hyponatremia is the most common electrolyte disorder, affecting approximately 5% of adults and up to 20% of individuals over 65. The onset of symptoms is closely related to the rate of sodium decline and is categorized as mild (130–135 mmol/L), moderate (125–129 mmol/L), or severe (<125 mmol/L). Symptoms range from mild, nonspecific complaints to life-threatening cerebral edema (4–6).

CSW is characterized by renal loss of sodium and water in the setting of normal kidney function, leading to extracellular volume depletion. The exact pathophysiology remains unclear; elevated BNP and atrial natriuretic peptide levels are frequently implicated in promoting natriuresis.

Both CSW and SIADH manifest with low serum osmolality, high urine osmolality, and high urine sodium. However, CSW differs by the presence of clinical hypovolemia, hemoconcentration, and positive response to isotonic saline, whereas SIADH patients remain euvolemic or hypervolemic and may worsen with fluid administration (7–10).

In Dandy-Walker malformation, hydrocephalus and hypothalamic dysfunction may contribute to dysregulated sodium handling, occasionally triggering CSW. Previous studies have described similar cases linking Dandy-Walker malformation and hyponatremia secondary to cerebral salt wasting (11–13).

Fludrocortisone acts by enhancing renal sodium reabsorption and plasma volume expansion. Recent trials (e.g., FLUSH SALT 2023) have shown improved sodium stabilization and reduced recurrence in neurocritical CSW patients treated with fludrocortisone (9).

CONCLUSION

In central nervous system disorders, the most common causes of hyponatremia are SIADH and CSW. Differentiating between them is challenging, but a favorable response to isotonic saline and evidence of hypovolemia strongly support CSW. In our case, a young patient presenting with hyponatremia was diagnosed with CSW related to Dandy-Walker syndrome. After treatment with fludrocortisone, no further hyponatremic episodes occurred. This case emphasizes the importance of evaluating sodium balance disturbances in congenital CNS malformations and considering CSW in the differential diagnosis.

DECLARATIONS

Ethics Approval and Consent to Participate: Not applicable.

Consent for Publication: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Availability of Data and Materials: Not applicable.

Competing Interests: The authors declare that they have no competing interests relevant to this manuscript.

Funding: This manuscript did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

AI: Not Used

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