Suprasellar arachnoid cyst resulting in the syndrome of inappropriate antidiuretic hormone secretion

Case report

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The syndrome of inappropriate antidiuretic hormone secretion (SIADH) is occasionally seen after hypothalamic injury or dysfunction, although it typically occurs in association with other endocrine disturbances. It has never been described as a presenting feature of a suprasellar arachnoid cyst (SAC) in the pediatric population. The authors describe the case of an enlarging SAC resulting in SIADH as the only presenting feature, with an otherwise normal hypothalamic-pituitary axis.

An SAC was diagnosed in utero in this 5-month-old girl who had a normal functioning hypothalamic-pituitary axis on presentation. Because of cyst enlargement and hydrocephalus, the patient was scheduled for surgery; however, preoperative labs revealed SIADH. After stabilizing the serum sodium concentration with fluid restriction and the administration of 3% sodium chloride, the patient underwent endoscopic cyst fenestration. Postoperatively, she had complete resolution of the SIADH.

Syndrome of inappropriate antidiuretic hormone secretion as the presenting symptom of an SAC has not been previously described. In the aforementioned patient, the proposed mechanism for SIADH was enlargement of the suprasellar arachnoid cyst causing compression of the supraoptic and paraventricular nuclei and thus overstimulating the secretion of arginine vasopressin, which resulted in SIADH.

The association of SIADH with an SAC is reportable, as is the resolution of the SIADH via cyst fenestration. The authors suggest that SIADH is an uncommon presenting feature of SACs and that syndrome resolution is possible with cyst decompression. (DOI: 10.3171/2010.8.PEDS10156)

**Key Words** • arachnoid cyst • hyponatremia • syndrome of inappropriate antidiuretic hormone secretion

**Abbreviations used in this paper:** AVP = arginine vasopressin; SAC = suprasellar arachnoid cyst; SIADH = syndrome of inappropriate antidiuretic hormone secretion.

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Hyponatremia, especially that due to SIADH, is commonly seen in neurosurgical patients. Specific neurological causes of SIADH include infection, vascular disorder, neoplasm, traumatic brain injury, hydrocephalus, and epilepsy. Hyponatremia is also seen after hypothalamic injury or dysfunction, although it typically occurs in association with other endocrine disturbances.

Patients with SACs typically present with symptoms related to mass effect leading to macrocrania and obstructive hydrocephalus. Precocious puberty, hypothyroidism, and growth hormone deficiency are the most common endocrine disturbances that have been reported. We present the case of an enlarging SAC resulting in SIADH in a patient with an otherwise normal hypothalamic-pituitary axis.

**Case Report**

*History.* An SAC was diagnosed in a girl in utero and was confirmed on postnatal head ultrasonography. An initial endocrine evaluation did not demonstrate evidence of hypothalamic-pituitary dysfunction. Notably, her plasma sodium concentration was 136 mmol/L. She was monitored with MR imaging and head ultrasonography (Fig. 1). Surgical treatment was not considered post-

This article contains some figures that are displayed in color online but in black and white in the print edition.
natally because initially her fontanel was sunken and her head circumference was less than the 98th percentile. Ultrasonography studies obtained when she was 5 months of age demonstrated enlargement of the cyst with ventricular dilation and fullness of the fontanel; hence, surgical intervention was scheduled.

Preoperatively, she was noted to have a plasma sodium concentration of 129 mmol/L. She was not receiving any medication to account for her new hyponatremia. Her serum osmolality was 270 mmol/kg/H₂O, urine specific gravity 1.010, urine sodium 133 mmol/L, and urine osmolality 431 mmol/kg/H₂O. Clinically, she was euvoletic with a positive fluid balance. A diagnosis of SIADH was made.

Her repeat endocrine labs (thyroid-stimulating hormone, T3, T4, luteinizing hormone, follicle-stimulating hormone, prolactin, cortisol, and insulin-like growth factor–I) were unremarkable. Her sodium level decreased to 121 mmol/L, and after a trial of rigorous fluid restriction and 3% sodium chloride, the sodium stabilized at 129 mmol/L.

Treatment. The SAC was then endoscopically fenestrated into the interpeduncular cistern via a right frontal precoronal bur hole (Fig. 2).

Postoperative Course. Postoperatively, she had a rapid resolution of the SIADH. By the 1st postoperative day her serum sodium concentration was 132 mmol/L, despite having discontinued the 3% sodium chloride immediately after the operation. Fluid restriction was weaned over the next 3 days, and her serum sodium concentration remained normal despite ad lib feeding. She was discharged on postoperative Day 5 with a sodium concentration of 135 mmol/L. 9 days postoperatively as well as 7 weeks postoperatively, which confirmed the resolution of SIADH.

Discussion

The incidence of SACs in the pediatric population is reportedly rare.3,5,14 However, the European cooperative study states that, based on a cohort of 285 patients, SACs account for 11.3% of all intracranial cysts in patients younger than 16 years of age.9 Moreover, 9.8% of all intracranial cysts in the article by Crimmins et al.2 were SACs. Although they have also been seen after trauma, these cysts are believed to be congenital lesions.2,3 Crimmins et al.2 state that fetal SACs develop late in gestation, most commonly in the last trimester of pregnancy. Their theory of pathogenesis proposes that there is a diverticulum or splitting of the Liliequist membrane with upward herniation of the apical membrane via CSF pulsation.2,3

An alternative theory is that SACs can result from cystic dilation of the interpeduncular cistern.3

On review of several studies, the typical presentation of a patient with an SAC includes macrocrania, obstructive hydrocephalus, psychomotor retardation, incidental discovery of the cyst prenatally, head bobbing, seizures, neurological deficits due to mass effect, and endocrine dysfunction.2,3,5,11 The typically reported endocrine disturbances have been precocious puberty, hypothyroidism, and growth hormone deficiency.1,2,8,11 Most authors have found that endocrine disorders do not regress postoperatively.2,11,15

The occurrence of SIADH in association with an SAC in the pediatric population has not been previously described. Hyponatremia has been seen in association with a Rathke cleft cyst; however, it is typically seen with other endocrine disturbances such as hypothalamic dysfunction and precocious puberty.5,10

Syndrome of inappropriate antidiuretic hormone secretion is typically defined as hyponatremia with resulting hypoosmolality. It results from the inability to excrete free water. Arginine vasopressin is produced by neurons in the paraventricular and supraoptic nuclei of the hypothalamus. It is then released in neurosecretory granules from the posterior pituitary. The AVP acts on the V2 vasopressin receptor in the kidney and allows water to be retained. Arginine vasopressin secretion is mainly regulated by changes in plasma osmolality, and basal levels of AVP do not increase until the plasma osmolality exceeds 280 mOsm/kg.13
The diagnosis of SIADH requires normal renal, thyroid, and cortisol function. The patient should be euvoletic to slightly hypervolemic with a plasma osmolality < 275 mOsm/kg, have less than maximally dilute urine, and demonstrate natriuresis. Some authors recommend obtaining an AVP level, but this level may be normal in 10%–20% of patients with SIADH. The main diagnostic dilemma is between cerebral salt wasting and SIADH. Cerebral salt wasting represents a hypovolemic state that is associated with hyponatremia, less than maximally dilute urine, and natriuresis. Volume status appears to be the main distinguishing factor between cerebral salt wasting and SIADH. Suggested clinical indicators of volume are weight, fluid balance, jugular venous distension, hematocrit, blood urea nitrogen, creatinine, uric acid, bicarbonate, central venous pressure, and pulmonary wedge pressure. The treatment of SIADH involves the removal of the offending agent and fluid restriction; however, severe hyponatremia requires correction with hypertonic saline. As regards the patient in the above case, we suggest that the cause of SIADH was enlargement of the SAC causing compression of the supraoptic and paraventricular nuclei. This compression in turn overstimulated the secretion of AVP resulting in SIADH. As this compression was not longstanding, the patient had rapid resolution of the SIADH once the cyst was fenestrated and decompressed.

Conclusions

Syndrome of inappropriate antidiuretic hormone secretion has not been reported in association with pediatric SACs. Moreover, the almost immediate resolution of the SIADH with cyst fenestration, as seen in the reported patient, is important to note because most endocrine disorders typically do not regress postoperatively. We suggest that SIADH is an uncommon presenting feature of SACs and that syndrome resolution is possible with cyst decompression.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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