A case: Hydrocephalus secondary to suprasellar arachnoid cyst with reset osmostat and Isolated Growth Hormone Deficiency

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INTRODUCTION

Arachnoid cyst (AC) is a rare situation and its prevalence in children changes between 1.7% and 2.6%. Patients with a suprasellar arachnoid cyst (SAC) typically present with symptoms related to mass effect leading to macrocrania and obstructive hydrocephalus. Precocious puberty, hypothyroidism, and growth hormone deficiency (GHD) are the most common endocrine disturbances that have been reported patients with a SAC.

Persistent asymptomatic hyponatraemia, secondary to a rare subtype of syndrome of inappropriate antidiuretic hormone secretion (SIADH), reset osmostat described in the literature are rare. The syndrome of SIADH is occasionally seen after hypothalamic injury or dysfunction, although it typically occurs in association with other endocrine disturbances. Reset osmostat, a subtype of SIADH, has never been described as a presenting feature of SAC in the pediatric population. We present a case that presented with short stature and was found to have borderline hyponatremia. He was diagnosed with SAC-hydrocephalus-reset osmostat, a subtype of SIADH and GHD following the various.

CASE REPORT

A male aged 6 years and 3 months was brought with a symptom of short stature that had become marked as the child became older. He was born 3300 g term and the parents were not relatives. His height was 104.6 cm (-2.6 sds), body weight 18.2 kg (-1 sds), bone age 4 years 6 months, and arterial blood pressure 110/70 mm Hg. Turgor and tonus was normal, and prepubertal and systemic findings were normal on physical examination.

Target height was 175.5 cm (+0.36 sds) (mother 160 cm, father 178 cm).

Laboratory results
Glucose: 97 mg/dL
Sodium: 131.6 mEq/L (135-143)
Potassium: 4.85 mEq/L

Thyroid function: Euthyroid
Cortisol: 26.86 µg/dL
ACTH: 33.8 pg/mL
Blood lipid level: Normal
Urine sodium 251.9 mmol/L
Spot urine beta-2 microglobulin: 0,22 (0-0,3 mg/g creatinine)
Blood gases: Normal
Urine and blood amino-acid levels: Normal
Renal ultrasonography: Normal
Renin: 0.54 (0.5-5.9 ng/ml/hour)
Aldosterone: 11.93 (5-80 ng/dL)

Laboratory results
The ability of the kidney to concentrate

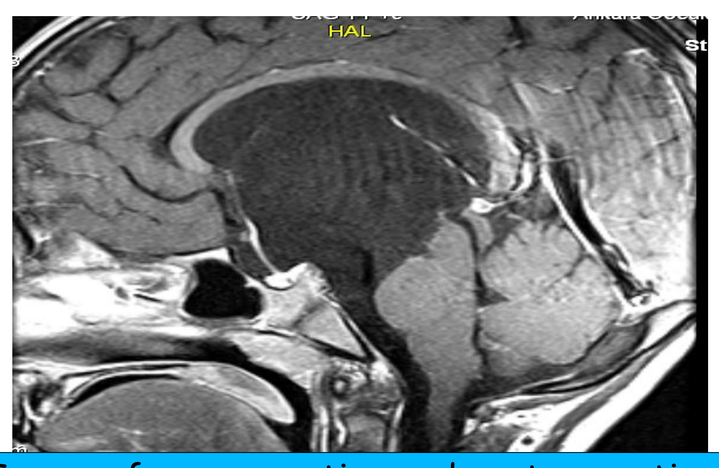
Density of the initial urine in the morning: 1027
Serum sodium: 130 mEq/L
Spot urine: 146.1 mmol/L

Dilute (urinary density after fluid intake): 1007
Serum sodium: 128,8 mEq/L
Spot urine sodium: 131.3 mmol/L)

Treatment
Fludrocortisone and 2 gr/day oral salt- Persiste hyponatremia

Diagnosis
Reset osmostat

Short stature → Mild hyponatremia → Reset osmostat → SAC-hydrocephallus → GHD



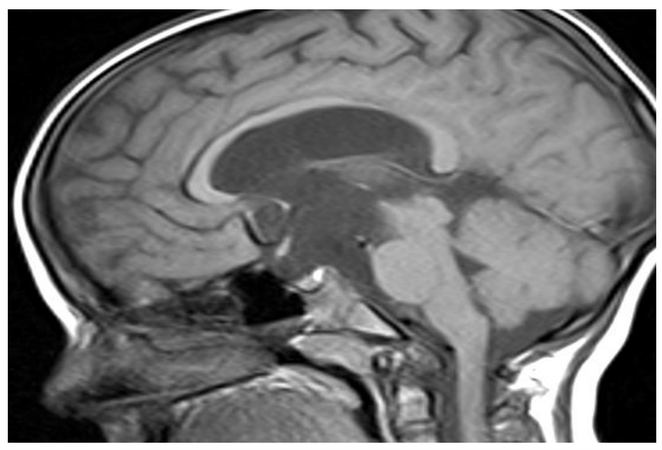


Image of pre-operative and post-operative suprasellar arachnoid cyst

The growth percentile during the postoperative 4 years of follow-up.

GH treatment was started
age was: 10 y 7 mo

Body weight 31.1 kg (-0.5 sds)
Height 124.2 cm (-2.63 sds)
Bone age 8 years
Growth rate 4.3 cm
Testis volumes: 3/3 ml

CONCLUSION

About 95% of AC cases are diagnosed with a neurological symptom and 5% due to endocrine disease. Reset osmostat is the C subtype of the four SIADH subtypes. The normal plasma osmolality is over 280 mosm/kg but the plasma osmolality threshold and plasma sodium concentration are low in this condition. Reset osmostat should be suspected in a patient with SIADH with moderate hyponatraemia (usually 125-135 mmol/L) that remains stable despite variations in water and sodium intake. The diagnosis is important as it is unnecessary and ineffective to try to treat the hyponatremia. The diagnostic criteria of reset osmostat are: euvolaemia; normal renal, adrenal and thyroid function; ability to concentrate urine; ability to excrete fluid overload, and hyponatraemia despite salt overload.

In conclusion, these patients should be treated and followed-up by keeping in mind that hyponatremia is an important indicator and that endocrine problems can be coexistent with SAC or can develop during follow-up with individual variations in each patient.

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