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Case Report

Partial empty Sella syndrome in women-the significance of obstetric and lactational history

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ABSTRACT

Empty Sella syndrome is an uncommon condition characterized by the shrinking or flattening of the pituitary gland, resulting in the filling of the Sella turcica with cerebrospinal fluid rather than the normal pituitary gland. In this report, we present a case of undiagnosed partial empty Sella syndrome, which was found to be caused by pituitary hypophysitis with an idiopathic etiology. The patient, a middle-aged individual, presented atypically with acute adrenal insufficiency induced by a lower respiratory tract infection. The diagnosis was made following an investigative work-up that took into consideration the presence of hypotension, electrolyte imbalances, and a history of two post-partum lactational failures. Hormonal supplements were used to manage the patient conservatively, and no significant complications were observed.

Keywords: Empty Sella, Pituitary gland, Hypophysitis, Adrenal insufficiency

INTRODUCTION

Empty Sella is a medical condition that refers to the herniation of the subarachnoid space within the Sella, which is often accompanied by varying degrees of pituitary gland flattening. This condition can be subdivided into primary or secondary. Primary empty Sella (PES) is an idiopathic insult to the pituitary resulting in inflammation i.e., pituitary hypophysitis and is diagnosed after ruling out prior pituitary pathological conditions such as previous surgical, pharmacological, or radiotherapy treatment of the sellar region. On the other hand, secondary empty Sella (SES) can occur following brain trauma, pituitary tumors treated via trans-sphenoidal surgery, pharmacological or radiotherapy; pituitary infectious or autoimmune disease, or rare causes such as spontaneous pituitary necrosis (ischemia or hemorrhage) i.e., Sheehan syndrome.[1,2]

Empty Sella syndrome can also be classified as partial or complete. The border between these two categories is commonly defined in the current literature as partial empty syndrome when less than 50% of the Sella is filled with cerebrospinal fluid (CSF), and pituitary thickness is equal to or greater than 3 mm. In contrast, the latter is when more than 50% of the sella is filled with CSF, and pituitary thickness is less than or equal to 2 mm.

In clinical practice, PES is observed in around 8-35% of general population, with female-to-male preponderance of 5:1.[3-6] A major prevalence of PES is noted among women with a history of at least one completed pregnancy in their physiological history, as in our case.

Presentation of idiopathic pituitary hypophysitis can vary depending on extent of gland involvement and resulting hormonal deficiencies. Variability makes it challenging to diagnose and treat this condition effectively.

CASE REPORT

A female patient in her late 30s presented to the hospital with a 1 week history of low-grade, intermittent fever and cough with vomiting and loose stools. This was associated with generalized fatigue. Otherwise, there was

no other medical, trauma, surgical, or recent medication history. She had normal growth and sexual development. The patient's oxygen saturation was found to be 94%, as a result of which lower respiratory tract infection was suspected and the patient was started on 2 liters oxygen via nasal cannula. The patient was admitted and started on empirical antibiotics after sending the blood sample for culture. On examination, the patient was drowsy and found to be repeatedly hypotensive, which was followed up by hormone assays such as serum cortisol levels that came out to be repeatedly low for the amount of physiological stress the patient was undergoing-4.46 µg/dL and 2.95 µg/dL (reference range, 4.82-19.5 µg/dL); ACTH was found to be 41 pg/ml. Serum TSH was normal (3.360 uIU/ml) with low free T4 of 0.3 ug/dl. Other laboratory investigations showed severe hyponatremia (sodium 109 mmol/L) with calculated serum osmolality 217 mOsm/kg H_2O (reference: 275-295 mOsm/kg H_2O). Her spot urine sodium was 63 mmol/L and urine osmolarity was 363 mOsm/Kg. Her serum sodium level not improving with routine salt supplementation and intravenous saline administration. Further probing of her past history revealed a history of irregular menstrual cycles that recently progressed to amenorrhea in the past few months, as well as lactational failure for the last two postpartum periods without any history of post-partum hemorrhage, hence a tentative diagnosis of central hypopituitarism was made and co-syntropin test was ordered to know response of adrenals to ACTH, which did not increase serum cortisol level significantly (12.28, 15.4, 17.6 pg/ml at baseline, 30 and 60 min after cosyntropin administration) (Table 1).

Brain magnetic resonance imaging (MRI) demonstrated a thin and flat pituitary gland located in the Sellar floor with prominent cerebral spinal fluid space (Figure 1), compatible with hypoplasia of the anterior pituitary. A diagnosis of empty Sella syndrome as a consequence of possible pituitary hypophysitis was made.

Table 1: Baseline investigations.

Biochemistry	Value (at presentation)	Reference range
Serum cortisol	4.46 and 2.95 µg/dl	4.82-19.5 µg/dL
ACTH	41 pg/ml	10-60 pg/mL
Serum TSH	3.360 µIU/ml	0.34-5.6 µIU/ml
Free T4	0.3 ng/dl	0.7-1.9 ng/dl
Serum sodium	109 mmol/l	136-145 mmol/l
Serum osmolality	217 mOsm/kg	275-295 mOsm/kg
Urine sodium (spot)	63 mmol/L	~20 mmol/l
Urine osmolality	363 mOsm/Kg	50-1200 mmol/kg
Co-syntropin stimulation (Min)		
30	15.4 pg/ml	>18-20 pg/ml
60	17.6 pg/ml	>20 pg/ml

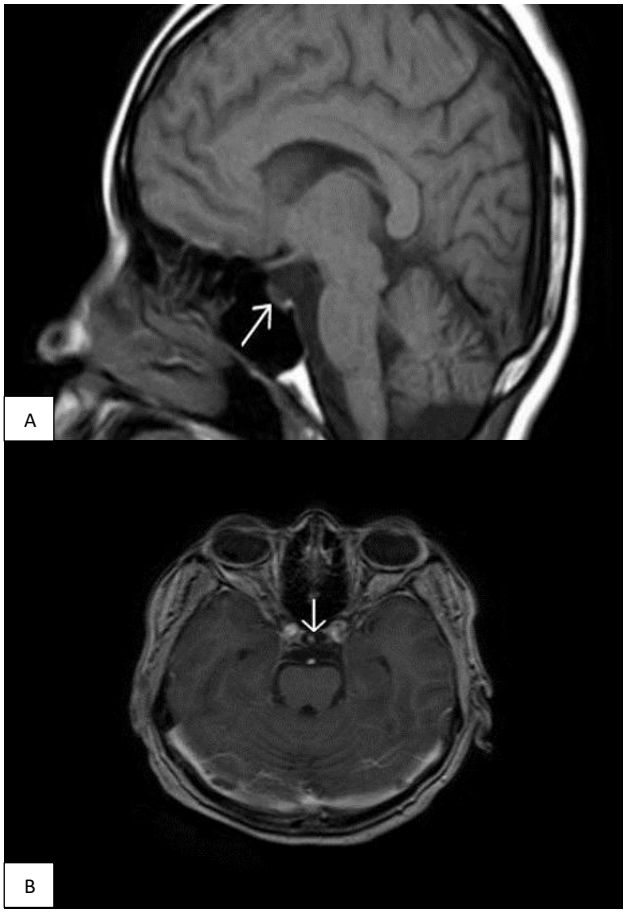

Figure 1 (A and B): Illustrate hypoplasia of the anterior pituitary lobe leading to empty Sella. Anteroposterior dimension of Sella appears increased. Posterior pituitary and pituitary stalk appear normal.

This case shows long-term silent hypopituitarism unmasked by lower respiratory tract infection presenting as acute adrenal insufficiency. This case is reported for its rarity of presentation.

Treatment and follow-up

The patient was started on intravenous hydrocortisone 100 mg every 8th hourly for 7 days post which the serum sodium levels gradually rose from 109 to 136 mmol/L. The patient eventually recovered and returned to baseline health. She was discharged after 11 days of hospitalization. The patient was maintained on 5 mg prednisolone in the morning and 2.5 mg prednisolone in the night and thyroxine 75 mcg daily without any sequelae when followed at the outpatient department.

DISCUSSION

Empty Sella patients are usually asymptomatic, although some patients develop panhypopituitarism.[7] Other aspects of pituitary deficiency include secondary adrenal insufficiency (AI), as seen in our patient, hypothyroidism, hypogonadism, and growth hormone deficiency.

Posterior pituitary involvement is less frequent which can be partly explained by the vascular supply of the two regions. The inferior hypophyseal arteries arising from the cavernous portion of the internal carotid artery divide into medial and lateral arteries. These arteries join with those from the opposite side forming an anastomotic ring around the infundibular process of the neurohypophysis and protect it from excessive damage.[8]

In our patient, severe hyponatremia was encountered. The exact prevalence of hyponatremia due to empty Sella and consequent secondary adrenal insufficiency is not known. Diederich et al reviewed the causes of hyponatremia in 185 patients over a 20-year period, where 28 (15%) had secondary adrenal insufficiency as the main cause, and 12 of these (6%) had empty Sella. This study was selective for patients with complex or refractory hyponatremia therefore the true prevalence of hyponatremia due to empty Sella is probably lower than 6%, but perhaps more common than generally thought.[9]

Hyponatremia in empty Sella is a result of excessive vasopressin secretion due to hypo-cortisolism that fails to suppress vasopressin, and not hypoaldosteronism.[10,11] Unlike primary AI, the renin-angiotensin-aldosterone system (RAAS) is intact in secondary AI, and therefore, hypoaldosteronism and the electrolyte abnormalities secondary to it such as hyperkalemia and metabolic acidosis are not seen here. Failure of cortisol release on cosyntropin stimulation seen in this case is likely due to long-standing absence of stimulation from the pituitary gland.

Contrary to the usual hyponatremia management, i.e., fluid restriction and saline administration, empty Sella syndrome-associated hyponatremia is promptly corrected by glucocorticoid supplementation and conversely can have catastrophic outcomes if missed. Twice or thrice daily cortisone is preferred over longer-acting steroids such as dexamethasone as it most closely mimics physiologic cortisol release.

Obtaining a comprehensive obstetric and lactational history is crucial in the diagnosis of endocrine conditions in parous females. In particular, the inability to lactate during the postnatal period, even in the absence of postpartum hemorrhage, can serve as an important diagnostic clue. It is noteworthy that these conditions can present at any age and may have atypical manifestations, such as extreme electrolyte disturbances such as hyponatremia and hyperkalemia, fatigue that is exacerbated by intercurrent infections or stress. Timely diagnosis and management can be lifesaving. Furthermore, it is crucial to administer additional doses of steroids during times of stress. Additionally, individuals with endocrine conditions should carry a steroid card, as it can aid in the prompt recognition and treatment of adrenal insufficiency during emergency situations.

CONCLUSION

This case is presented to highlight the importance of long-term undiagnosed and silent pituitary malfunction requiring adequate evaluation at the earliest available opportunity and treating the underlying condition to prevent a major catastrophe.

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