Refractory epilepsy and recurring hyponatremia are leading in Primary Empty Sella Syndrome: A Case Report

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ABSTRACT Background: Empty Sella syndrome is a neuroendocrine disorder rarely in population. Often occurred with a female: male = 4:1, in middle age, and obesity. **Report:** We present a case of a 27-year-old male, short structure, BMI 20,4 kg/m2, undescended testes. He was diagnosed with epilepsy symptomatic. The first symptoms occurred when he was 25 years old with the general-tonic clonic pattern; blood test was showed normal only found low hyponatremia, brain MRI and EEG were abnormal. Unfortunately, his seizure was not relieved with medical management, phenytoin for two years. Prolong seizure occurred; he had status epilepticus. Brain MRI showed empty Sella, moderate encephalopathy with EEG, severe serial hyponatremia (105 mEq/L), and loss of sodium urine. Subsequently, seizure relieves controlled with normal sodium level by mineralocorticoid drug. **Conclusions:** Clinicians should be aware of the typical signs of an empty sell for its proper diagnosis & management.

KEYWORDS Short structure, undescended testes, severe hyponatremia, seizure

Introduction

Empty Sella syndrome (ESS) is a neuroendocrine disorder rare case in populations. Radiological imaging showed sella turcica without pituitary gland.[1]. ESS divided into primary and secondary ESS. Primary ESS figured with empty pituitary gland in Sella cavity. Primary ESS is also is known as arachonidocele. Hypertension and obesity are associated. Secondary ESS defect pituitary gland after an injury with pituitary function impairment. The association of primary ESS and seizure in the same patient is, to our knowledge, very rare. We report a case of a patient who presented with clinical signs of an intractable seizure in whom primary ESS was diagnosed on the radiological investigation.

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Case presentation

A younger male, 27 years old, was referred to Sanglah General Hospital for intractable seizure evolving for two months. He was a product of normal term pregnancy, with birth weight 3100g, birth length of 44 cm and the circumference of 33cm. Her mother complained of seizure that had not been investigated, but he had, however, received anti-epileptic drugs for seizure. Upon clinical examination, the patient with short structure, height was weight 40kg 140cm. The patient has a small body with body mass index (BMI) 20,4 kg/m2 with undescending testes. Hormone analysis showed; Cortisol 37,31 ug/dL (4,3-22,4 ug/dL), Testosteron <2,5 ug/dL /(249-836 ug/dL) Free T4 0,53 ug/dL (0,9-1,7 ug/dL); TSH 1,32 microIU/mL (0,27-4,2 microIU/mL), periodic hyponatremia with the lowest level 105 mEq/L (figure 1). Growth hormone secretion (Ornithin test) is available only in a few institutions in Indonesia and it was not used in this patient.

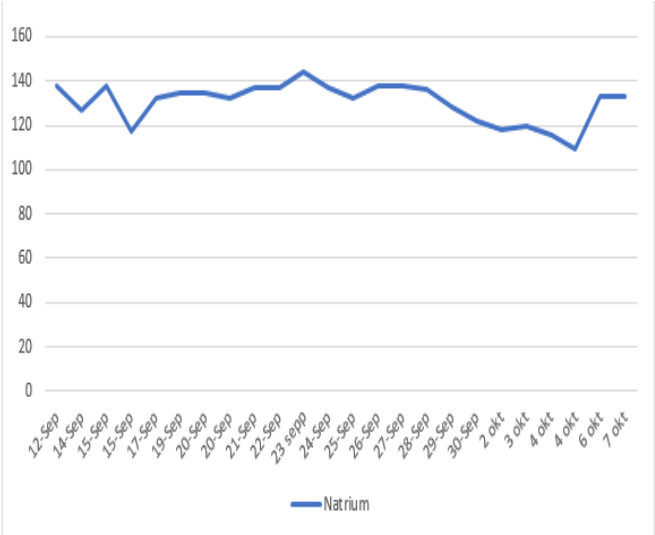

Figure 1: Sodium blood level in mEq/L.

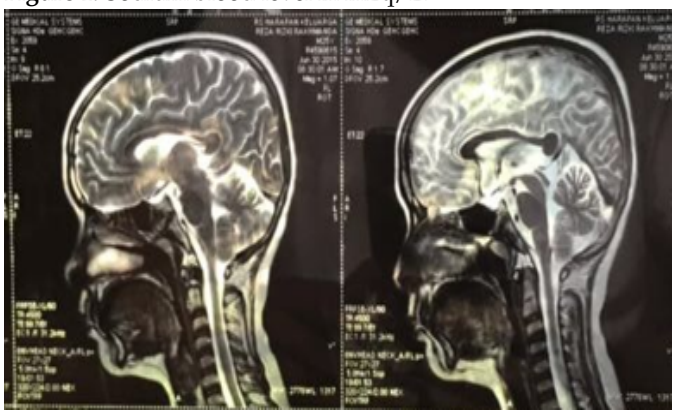

Figure 2: MRI brain (T-2) sagittal showing empty sella. Hyperintense signal intensity in the region of sella turcica suggestive of CSF filled sella (orange's narrow).

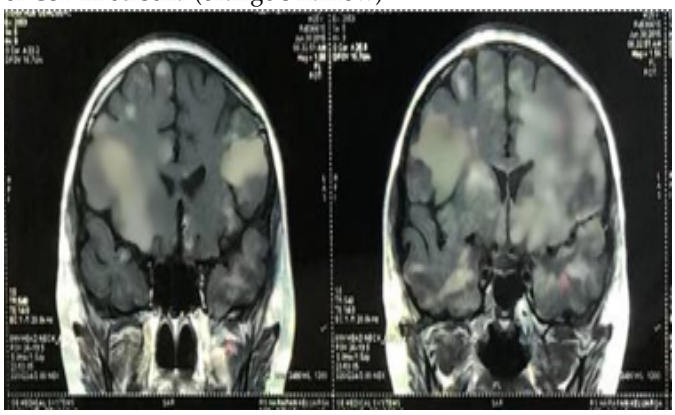

Figure 3: MRI brain (T-2) coronal view is showing empty sella. (orange's narrow).

The Electroencephalography showed encephalopathy frontobilateral (abnormal I). The brain MRI presented empty pituitary gland in the sella turcica. There were no signs of raised intracranial pressure.

The patient was managed with mineralocorticoid therapy for hyponatremia. He was reviewed, clinically three times a year. He presented without seizures and hyponatremia.

Discussion

ESS has primary and secondary types.[3] In this case, the patient happened ESS which is associated with congenital abnormalities in the process of forming the diaphragm sella and the pituitary gland impingement due to increase intracranial pressure when the process of expansion subarachnoid space into Sella tursica. Fat women with many children are often found in cases of ESS. [2] In this patient there is no compression of the pituitary gland on the stalk, because of hyperprolactinemia is not found. [3]

Neuroendocrine impairments are the most common symptoms found in this case. In this case, it is found that hormone levels of FT4 are low while TSH is normal suggests a malfunctioning of the thyroid gland. In some scientific literature described primary empty sella syndrome is associated with hypothyroidism. In population, prevalence hypothyroidism with ESS 1: 80,000 to 1: 120,000 regardless of race and gender and the highest incidence of secondary empty sella syndrome. [2] Most thyroid hormone disorders are found in the case of the secondary type of empty sella syndrome with the aetiology of macroadenoma, radiation and surgery in the pituitary gland.[2] In this case, impaired thyroid gland function occurs in the primary empty sella syndrome.

This patient also had experienced recurrent hyponatremia. In patients with hyponatremia and hypogonadism, a screening test should be carried out immediately to distinguish the location of hypopituitarism or adrenal cortex insufficiency.[3] Yammato et al. conducted research and observations in some patients with hyponatremia, and hypopituitary found mild dehydration with blood pressure abnormalities, low body weight, abnormal blood nitrogen urea levels, total protein and hematocrit.[4]

Primary ESS occurred hormonal disorders 25-35% with the most considerable abnormality in growth hormone by 30-60% of cases.[5] Clinical examination, there was a disorder of growth and development, the structure of the height did not match the age, hypogonadism with the sign of undescending the testes with low levels of testosterone hormone.

We reported patients with severe hyponatremia induced general tonic-clonic seizures. Anti-epileptic drugs therapy for two years cannot stop seizures. In severe hyponatremia with levels <120 mM showed clinical neurology seizures.[6] The frequency of seizures increases when blood sodium levels <110mmol.[7] Hyponatremia will cause interference with the blood-brain barrier by affecting the environment in and outside the cell. Hyponatremia causes disruption in membrane neuron for ion exchange, changes in excitatory and inhibitory neurotransmitter levels, neurotransmitter receptor disorders, and adenosine triphosphate (ATP) production in excitatory neurons. [7] Hyponatremia causes oedema of brain neurons and glial cell oedema and reduces space between cells. Increases the occurrence of interactions between membrane excitation neurons and surrounding neuron groups to trigger seizures.[8] Hyponatremia is very difficult to correct by administering sodium intravenous but is very responsive to hydrocortisone.[2] Hyponatremia occurs because of a disruption of water secretion

in the renal tubular system due to reduced glucocorticoid hormones. Disorders of water excretion occur through the ADH Dependent mechanism and ADH-Independent. In the dependent ADH mechanism, a decrease in an extracellular fluid with low levels of glucocorticoid hormones causes the release of the hormone ADH and changes in renal hemodynamic autoregulation function. In the ADH-independent mechanism, the decrease in fluid levels in the distal renal tubule causes a significant decrease in cardiac output, as well as a decrease in renal blood flow.[9,10]

When a secondary adrenal insufficiency is diagnosed, hyponatremia must be corrected immediately by administration of hydrocortisone (15-25 mg a day).[11] The glucocorticoid combination with sodium-rich fluid should be of more concern to prevent the onset of osmotic demyelination syndrome.[12]

Conclusion

Clinicians should be aware of the typical signs of empty sella for its proper diagnosis & management.

Competing Interests

There were no financial supports or relationships between authors and any organization or professional bodes that could pose any conflict of interest.

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Abbreviations

MRI = Computed tomography, EEG = electro-enchepalo-graphy

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