Case Report

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A Rare Case of Central Diabetes Incipidus Due to Post Pituitary Gland Agenesis: A Case Report

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ABSTRACT

Background: Diabetes insipidus is a syndrome that begins with polyuria and is often associated with polydipsia. Three significant differential diagnosis are important to consider in evaluating the causes of polyuria, including primary polydipsia, diabetes mellitus, and diabetes insipidus. In diabetes incipidus, ADH hormone is not synthesized and secreted in central DI or the hormone has lost its function in the renal tubules in nephrogenic DI type.

Case Presentation: The present case was about 5.5 year-old-boy with polyurea and polydipsia from the beginning of the infancy. The patient had the serum osmolality of 277 mOsm/kg with the 24 hours urine volume of 4500 cc and urine osmolality of 200 mOsm/kg. The patient underwent water deprivation test for 12 hours. After administration of 20 µg of desmopressin spray, the urine osmolality increased to 720 mOsm/kg. Central diabetes insipidus was diagnosed. There was posterior hypophyseal agenesis in the brain MRI.

Conclusion: The above patient seemed to be one of the rare cases of autosomal recessive central DI that became symptomatic with polyuria and polydipsia from the early days of life. This rare and interesting case had isolated posterior pituitary agenesis. Desmopressin treatment resolved the patient's complaints. The patient was asymptomatic, and had normal growth in one year follow up.

Introduction

Diabetes incipidus (DI) manifested clinically with polyurea and polydipsia.[1] Polyuria is defined as urine volume of more than 2 liters per square meter of the body the in 24 hours.[2] It should be distinguished from the frequent urination and nocturnal enuresis that are not associated with an increasing urine volume per day. The

disease diagnosis requires a precise history of the onset of symptoms, accurate measurement of urine volume, serum and urinary osmolality levels. Diabetes incipidus has two types. The central type occurs due to failure to secrete the antidiuretic hormone (ADH) from the posterior pituitary gland and urine concentrations does not occur in the urine collecting tubules. As a result, urine volume increases and its osmolality decreases.[3] A rare type of familial central DI may be present at birth in infants with homozygote mutation of the arginine vasopressin (AVP) hormone. However, in the nephrogenic diabetes insipidus, the synthesis and secretion of this hormone from the pituitary gland is normal, but there are different degrees of resistance to this hormone in the urinary collecting tubules, which lead to similar symptoms.[4] The distinction between these types is made by the therapeutic response to desmopressin. The therapeutic response is seen in the central type of diabetes incipidus but there is no therapeutic response to desmopressin in the nephrogenic type of diabetes incipidus.[5]

Case Presentation

The patient was a 5.5-year-old boy who had been suffering from polyuria and polydipsia since the age of 2 months. Due to frequent nocturnal waking ups for urination and sometimes urinary incontinence till 5.5 year old, the patient's mother had been referred for examining the etiology and treating her son. The parents had consanguinity marriage. They were cousins and there was no similar case in their family. The patient's history suggested that the child was a term infant who was born via vaginal delivery with no history of hospitalization in the neonatal period due to cyanosis, jaundice or seizure. The patient had no history of head trauma. His weight and height were 18 kg and 115 cm, respectively. The boy's growth chart showed that he was in the 25th percentile for weight and 75th percentile for height. The patient had the serum osmolality of 277 mOsm/kg with the 24 hours urine volume of 4500 cc and urine osmolality of 200 mOsm/kg (Table 1).

Table 1. Laboratory Tests

Variable	
FBS	89mg/gl
UREA	13mg/dl
CREA	0.8
Serum NA	135mg/dl
Serum osmolarity	277
Urine SG	1005
Urine volume	4500cc
Urine osmolarity	200

Regarding the history of polyuria and polydipsia and laboratory tests, the patient underwent water deprivation test for 12 hours under the direct supervision of a physician. After administration of 20 μg of desmopressin spray, the urine osmolality increased to 720 mOsm/kg, which confirmed the diagnosis of central DI. Brain magnetic resonance imaging (MRI) was performed to evaluate the cause. There was posterior pituitary agenesis in the brain MRI (Figure 1). Diagnosed with the central DI, the patient was treated by desmopressin spray, one puff every 12 hours. In a one year follow-up, the patient was asymptomatic, and the growth in height and weight was increasing according to the standard curve.

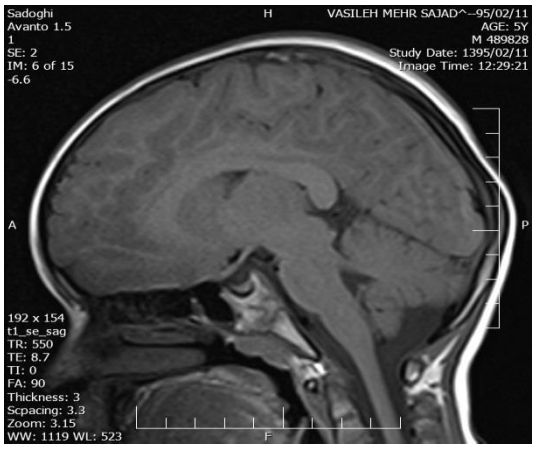

Figure 1. A Mid Sagital T1 MRI of Brain. No Evidence of Hypersignal in Posterior of Sella and Suggestive of Agenesis of Neurohypophyses.

Discussion and Conclusion

The prevalence of diabetes insipidus (DI) in the general population is 1 in 25000 people with a higher incidence rate in males. Some cases of inheritance by autosomal dominant and

recessive have been reported both in central DI and nephrogenic DI.[6] The patient described above was diagnosed with one of the rarest types of central diabetes insipidus due to the isolated agenesis of posterior pituitary. Regarding the onset of symptoms from beginning of infancy and his parents' consanguinity marriage the above patient was clinically diagnosed as familial central DI which might be related to homozygote mutation of the arginine vasopressin hormone. However the genetic study did not performed for definitive diagnosis because of the cost. The central diabetes insipidus has many causes.[7] The idiopathic type accounts for 31 to 51% of cases, which is due to the failure in synthesizing and secreting the hormone from the ADH generating cells.[8] Other common causes include head injury, neurosurgical operation, and primary or inflammatory brain tumors.[9,10] The congenital central DI cases reported already have had septo-optic dysplasia (SOD) or hypopituitarism with or without ectopia of the posterior pituitary lobe, which were often autosomal dominant and due to a gene mutation.[11] But, in this case, there was no similar case in the family despite the parents' close familial ties. Similarly, the height-age ratio for this patient was on the 75 percentile of the curve, which was different from the reported congenital cases that were associated with deficiency of other pituitary hormones, including the growth hormone.[12,13] In the patient's brain MRI, the full agenesis of the posterior pituitary gland with no involvement of the anterior lobe or agenesis of the corpus callosum or other middle-line anomalies was seen, and the patient had no abnormal neurological symptoms.

Conflict of Interests

Authors have no conflict of interests.

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