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Case Report Open Access

An Unusual Cause of Central Diabetes Insipidus in a Young Female

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Abstract

Central Diabetes Insipidus (CDI) in adults is most commonly occurs as a result of hypothalamo- pituitary surgery, head injury or various inflammatory and infiltrative disorders. CDI with mass lesions in the sellar-suprasellar area occurs due to infiltrative disorders like lymphoma, Langerhan's cell histiocytosis, and inflammatory disorders like hypophysitis, mass lesions like germinoma, craniopharyngioma and even metastases. Treatment and correct diagnosis depends on histopathology. Rosai-Dorfman Disease (RDD) is a rare disorder of unknown etiology characterized by abnormal proliferation of histiocytes. Extra nodal involvement is found in 40-50% of cases; with Central nervous system (CNS) involvement being uncommon hence in majority of instances intracranial disease is not suspected. Due to paucity of the reported cases, optimal treatment options are not known. We report a young female who presented with CDI, hyperprolactinemia and other features of hypopituitarism along with systemic manifestations including nodal and skeletal involvement. The patient was treated with combination of surgical debulking followed by oral glucocorticoid for 6 months with gratifying results.

Keywords: Rosai–dorfman disease; Sellar-suprasellar mass; Diabetes insipidus.

Introduction

Central diabetes insipidus (CDI) is usually caused by damage to posterior pituitary gland, stalk or hypothalamus with most common causes in adults being pituitary surgery, traumatic brain injury, and infective, inflammatory or infiltrative diseases in the sellar region such as meningitis, tuberculosis, hypophysitis, sarcoidosis, lymphoma, langerhans cell histiocytosis. Benign histiocytic proliferation and infiltration of hypothalamo-pituitary area as an etiology of CDI is uncommon.

Rosai-Dorfman disease (RDD) or Sinus Histiocytosis with Massive Lymphadenopathy (SHML) was first described in 1969 by two pathologists Juan Rosai and Ronald Dorfman [1]. It is a benign, idiopathic histiocyte proliferative disorder with systemic symptoms such as fever, generalized weakness, fatigue, lymphadenopathy and pathognomonic histological and immuno-histochemical characteristics. RDD is equally prevalent in males and females [2] but central nervous system involvement though rare, occurs twice as common in males. RDD presenting as sellar-suprasellar mass is extremely rare with only few cases mentioned in the literature [2-8]. Here we report a case of RDD presenting as suprasellar mass causing CDI, hypopituitarism with hyperprolactinemia.

Case Report

A 32 year-old- female presented to our institute with secondary amenorrhoea, polyuria, polydipsia and weakness for 9 years, with galactorrhoea, headache, and irritability for the last four years. She had a significant weight gain of 40 kg in past 6 months. There was no family history of any lympho-proliferative disorder, malignancy or immunodeficiency syndrome. Before coming to us, a primary care physician prescribed her levothyroxine 100 mcg, cabergoline 0.5 mg once a week and combined estrogen and progesterone pills for 5 months, on basis of her clinical picture and hormonal profile. As her condition remained statusquo, she stopped all medicines 4 months before being admitted with us, except for levothyroxine which she restarted from last 15 days.

The patient was obese (BMI-40.8 kg/m2), normotensive with no palpable lymphadenopathy or hepatosplenomegaly. Her routine investigations were normal except for hypercholesterolemia (269 mg/dl, 160-200) and hypertriglyceridemia (344 mg/dl, 100-150). Hormonal investigation revealed LH <0.1 mIU/ml (2.4-12.6), FSH 0.518 mIU/ml (1.5-1.5), testosterone 0.095 nmol/L (0.2-2.9), estradiol 39.5 pg/ml (12.5-166), ACTH 4.33 pg/ml (5-60), prolactin 64.8 ng/ml (4.79-23.3), iPTH 23.22 pg/ml (15-65), morning cortisol 301 nmol/L (171-536), T_3 1.34 ng/ml (0.8-2.0), T_4 12.28 µg/dl (4.8-12.7), TSH 0.018 µIU/ml (0.27-4.2). The baseline urine and serum osmolality were 59 mOsmol/kg (150-700) and 304 mOsmol/kg (275-290), respectively. With water deprivation, urine osmolality rose to 149.78 mOsmol/kg following administration of aqueous vasopressin (5 U s/c), suggestive of CDI. In the light of aforementioned investigations, she was started on oral desmopressin (100 mcg/day) and levothyroxine (100 mcg/day), following which she obtained relief.

As a part of imaging studies, gadolinium enhanced MRI of brain showed a T1 hypointense, T2/FLAIR mildly hyperintense, lobulated mass in supasellar region, measuring 2.8×2.5×1.8 cms, with homogeneous enhancement post-contrast. The mass was abutting the optic chiasma and left cavernous sinus, with pituitary stalk not separately visible (Figure 1A and 1B). Fluoro deoxy glucose positron emission computed tomography (FDG PET-CT) (Figure 2A and 2B) was done that revealed FDG avid suprasellar soft tissue mass (standard uptake value max. 37.7), multiple cervical level II, peri-pancreatic, portocaval,

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