Case Report

Endogenous Cushing’s Syndrome with Precocious Puberty in an 8-Year-Old Boy due to a Large Unilateral Adrenal Adenoma

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Adrenocortical tumors (ACTs) causing Cushing’s syndrome are extremely rare in children and adolescents. Bilateral macronodular adrenocortical disease which is a component of the McCune-Albright syndrome is the most common cause of endogenous Cushing’s syndrome. We report the case of a boy with Cushing’s syndrome who presented with obesity and growth retardation. The child was hypertensive. The biochemical evaluation revealed that his serum cortisol levels were 25.80 𝜇g/dL, with a concomitant plasma ACTH level of 10.0 pg/mL and nonsuppressed serum cortisol on high-dose dexamethasone suppression test (HDDST) to be 20.38 𝜇g/dL. Computed tomography of the abdomen demonstrated a 8 × 6 × 5 cm left adrenal mass with internal calcifications. Following preoperative stabilization, laparotomy was carried out which revealed a lobulated left adrenal mass with intact capsule weighing 120 grams. Histopathological examination revealed a benign cortical neoplastic lesion, suggestive of adrenal adenoma; composed of large polygonal cells with centrally placed nuclei and prominent nucleoli without capsular and vascular invasion. On the seventh postoperative day, cortisol levels were within normal range indicating biochemical remission of Cushing’s syndrome. On followup after three months, the patient showed significant clinical improvement and had lost moderate amount of weight and adrenal imaging was found to be normal.

1. Introduction

Adrenocortical tumors (ACTs) are quite rare in children and adolescents. Iatrogenic hypercortisolism is the most common cause of Cushing’s syndrome (CS) in infancy and childhood [1]. In infants and children less than 7 years of age, adrenal tumors and predominantly malignant adrenal carcinoma constitute the most common causes of Cushing’s syndrome [2] and that of those older than 7 years is ACTH secreting pituitary adenoma. ACTH-independent CS in children has been reported to be due to bilateral macronodular adrenocortical disease encountered in cases of McCune-Albright syndrome (MAS) [3]. The prevalence of the syndrome is reported to be between 1/100,000 and 1/1,000,000 and has been observed more commonly in females [4], with a tendency of severe presentation.

We report the case of an 8-year-old boy who was diagnosed with Cushing’s syndrome due to a left sided adrenal adenoma who had presented with generalized obesity and growth retardation with features of precocious puberty. Subsequently, a left adrenal adrenalectomy was performed and clinical stabilization resulted in weight loss and biochemical resolution of Cushing’s syndrome.

2. Case Report

An 8-year-old Bangladeshi boy presented to our out patient department (OPD) with complaints of gaining weight for the last 9 months. The parents were also concerned that the boy was not attaining the height according to his age. The child was born to nonconsanguineous parents at term, by
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Figure 3: Contrast-enhanced abdominal computed tomography (CECT) scan in axial view revealed a large, well-circumscribed mildly enhancing adrenal mass (8 × 6 × 5 cm approx.) having few internal calcifications at left suprarenal region, which had displaced left kidney slightly downwards (arrow-mass; triangle-left kidney).

Figure 4: Enlarged view of CT scan image in cross section, showing a large well-circumscribed mildly enhancing mass having few calcification at left suprarenal region.

The physical changes that occur in Cushing’s syndrome such as the moon face, hirsutism, and acne as well as the bulging of the cervicodorsal region (buffalo hump) are a result of the intense action of the glucocorticoids which favor the accumulation of fat in the abdomen, chest, and face (central obesity). Growth hormone and β-adrenergic receptor antagonists also induce lipolysis which facilitates increase in triglycerides and free fatty acids. Hypertension is a consequence of increased renin substrate and sodium retention, facilitating the expansion of extracellular volume. In our case, the patient presented with precocious puberty which is in line with the observations made by Michalkiewicz et al. [9] who found in a registry of 254 pediatric patients with ACTs that 55% with virilization alone. Twenty-nine percent presented with mixed overproduction of adrenal hormones. Only 5.5% percent of this group presented with isolated Cushings syndrome, and this tended to occur in older children (median age 12.6 years).

Several laboratory investigations are helpful in establishing the diagnosis and differentiating between suprarenal or hypophyseal origin. These include serum cortisol levels, plasma ACTH, and high-dose dexamethasone suppression test (HDDST) which has better sensitivity [10]. Radiological evaluation includes ultrasonography, computed tomography (CT) scan of abdomen, and MRI of the brain. CT scan has been shown to be more sensitive in identification and localization of tumor mass [11]. The recommended procedure is surgical removal of the tumor (adrenalectomy) [12], which resulted in rapid weight loss in our patient. Postoperative hydrocortisone supplementation following surgery for adrenal adenoma causing CS is necessary as the contralateral adrenal gland is usually hypoplastic secondary to prolonged suppressed ACTH secretion from the pituitary due to CS.

Pediatric adrenocortical tumors (ACTs) are most commonly encountered in females and in children less than four years. But our case being an 8-year-old boy forms a rare presentation of endogenous Cushing’s syndrome due to adrenal adenoma.

References

