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Case Report:
Cortisol Secreting Adrenal Adenoma in a 5 Year Old Child

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Abstract:
Adrenal adenoma is a rare endocrinal tumor in children. It can present with features of Cushing's syndrome. We present a case report of five years old female child who came with morbid obesity and hypertension. The patient manifested polyphagia, weight gain, and changes in sleep patterns. During physical examination we found a full-moon face, bulkiness in the cervico-dorsal (buffalo-hump) region, high blood pressure.
Key Words: Adrenal adenoma; Cushing's syndrome; Hypercortisolism

Introduction:
Adrenocortical tumours mostly secrete hormones, resulting in Cushing's syndrome, virilisation, hyperaldosteronism or feminization, while the non-functioning ones are unusual. Two syndromes have a clear association with adrenocortical tumour : Li Fraumeni syndrome is associated with mutations of the p53 gene, and Beckwith Wiedemann syndrome which has mutations in the 11p15 region.[1] Cushing's syndrome is defined as the combination of biological and clinical manifestations resulting from the presence of abnormally high and sustained circulating concentrations of glucocorticoids, whether of exogenous or endogenous production.

Case Report:

A 5 years old girl presented to us in the emergency department with complaints of excessive irritability and severe headache since 12 hours. History of abnormal body movements were present since past 4 hours which were associated with frothing from the mouth and incontinence of urine and stools followed by unconsciousness.

Child was born to 1st gravid mother to a non consanguineous marriage. Perinatal period was uneventful. Birth weight of the child was not known to the mother, but child was not overweight as per mothers observations. Child was fed breast milk till the age of 8 months after which complementary feeds were started. Milestones were normal for her age. Child started gaining weight since the age of 3 years. There has been a gradual increase in the weight of the child till present date. History of polyphagia and sleep disturbances was present. No H/o intake of exogenous steroid was present.

Childs Anthropometric measurements showed weight of 35 kg(>97th percentile), head circumference of 52.5cm(>50th percentile), height was 106cm(25th percentile), abdominal circumference of 82.5cm and mid arm circumference of 27.5cm with BMI > 95th percentile.

On examination child was markedly obese with a BMI of, with BP of 140/90 mmHg. Child had a plethoric moon face with truncal and subcutaneous and centripetal obesity and abdominal cutaneous striae. Hirustism was not present. Bone age was corresponding to the chronological age. Fundus examination, electrocardiograph (ECG) and other investigations, including blood glucose (and glucose tolerance test), serum cholesterol and renal function tests were normal. Laboratory investigations revealed leukocytosis and normal serum electrolytes and normal fasting and post-prandial glucose levels. Cushings syndrome was suspected and further endocrinological tests were performed to determine the etiology. Diurnal variation of plasma cortisol was not observed. The plasma cortisol level was not suppressed by dexamethasone suppression test. Plasma ACTH levels were markedly suppressed (less than 10pg/ml). Plasma free T_3, free T_4, TSH (thyroid stimulating hormone) were within normal limits.

Chest X – ray was normal. Ultrasonography of abdomen didn't reveal much so Computed tomography of the abdomen was done which showed a small hypodense (approx 5× 6 mm) lesion in medial limb of left adrenal gland s/o adenoma. MRI brain was normal.

Exploratory laparotomy through the left supra umbilical transverse incision was done. There was a left adrenal mass measuring 6×6 mm. Tumor was excised, it was not adherent to the surrounding structures.

Discussion:
The cause of hypercortisolism with clinical manifestations of Cushing's syndrome is most often as a result of the administration of synthetic glucorticoids.[2,3] Endogenous causes of hypercotisolism may be due to hypersecretion of adrenocorticotropic hormone (ACTH) by pituitary or by excessive secretion of the suprarenal glands. Overproduction of cortisol from the suprarenal tumor is a very uncommon pathology in children < 5 years of age.[4]

Fig 1: 5 years old patient weighing 35 kgs.

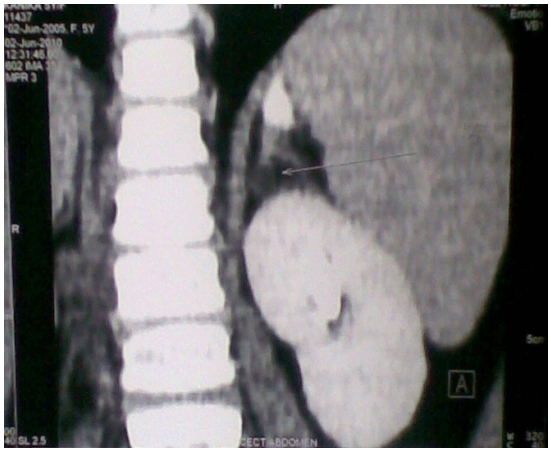

Fig 2: Cut section of CT abdomen showing suprarenal mass.

Adrenal cortical tumors (ACTs) constitutes less than 0.2% of all pediatric neoplasms and account for 6% of all adrenal tumors in children with an estimated incidence of 0.3 million population.[5] There is bimodal occurrence by age, with a peak incidence at less than 5 years and a second peak in the 4th and 5th decades of life. Because of rarity of these tumors, little is known about their causation and the influence of genetic factors.

Cushing's syndrome with the "full moon" characteristics of the face, hirsutism, acneiform lesions in the chest and arms, as well as a "hump" and bulging of the superior segment (trunk) in the cervicodorsal region. All the physical changes are as a consequence of the persistent and intense action of the glucorticoids, which favored the accumulation of fat in the abdomen, chest and face, condition central obesity. With the action of other compounds such as growth hormone and β- adrenergic receptor antagonists, these induce lipolysis, facilitating increase of triglycerides and free fatty acids. Thinning of the extremities is usually observed with proximal weakness. Decrease in protein synthesis leads to atrophy of the muscle fibers, a circumstance accentuated by the decrease in potassium.[6] Hypertension is usually seen as a consequence of increased renin substrate, inducing as a consequence the pressor response of angiotensin and catecholamines, and sodium retention, facilitating the expansion of extracellular volume. Apart from presence of stria on thighs and abdomen, other skin changes that tend to be present are capillary fragility, ecchymosis and hematomas.

There are laboratory studies to establish the diagnosis and help to discern between hypophyseal or suprarenal origin. These include serum cortisol level, free cortisol in the urine, determination of 17,hydroxycorticosteroids that has been substituted by free cortisol in the urine test in order to obtain better sensitivity, suppression test with nighttime dexamethasone, cardiac rhythm of cortisol, plasma ACTH.[7]

Radiological investigations include Ultrasonography, CT scan(abdomen) and MRI (brain) etc. CT scan is more sensitive than ultrasonography in identifying tumor mass and CT scan seems to be the best for localization of tumor masses.[8]

Tumors measuring more than 5 cm are usually malignant. Surgical removal of the tumor (Adrenalectomy) remains the procedure of choice.[9]

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