Virilizing adrenal tumor- a case report.

ARUNKARTHICK THANGAVADIVEL
Department of Paediatrics,
MADURAI MEDICAL COLLEGE AND HOSPITA

Abstract:
We present a case report of a 6 year old female child who presented with virilization secondary to an adrenal tumor. The girl, with no previous history of disease or treatment, presented with clitoromegaly and pubic hair development. The child was investigated and found to have an adrenal tumor. The tumor was 6.3 7.5 6.1 cm and was secreting sex steroids. The tumor was surgically removed. The pathology of the tumor showed large polygonal cells with abundant intensely eosinophilic cytoplasm without mitosis and necrosis. These findings were consistent with an adrenocortical tumor secreting hormones as the cause of virilization in this child.

Keyword: virilization, adrenocortical tumor, androgens, serum DHEAS.

CASE REPORT:
A girl of 6 years presented to our OPD with the complaints of increase in the size of the clitoris and appearance of pubic hair. The total duration of the complaints was for a period of six months. The child was apparently normal prior to this period.
EXTERNAL GENITALIA OF THE CHILD SHOWING CLITOROMEGALY AND EARLY PUBIC HAIR GROWTH

An USG-Abdomen showed a mass in the left adrenal region. A CT scan/plain & contrast) of the abdomen showed 6.3 x 7.5 x 6.1 cm sized well defined mass lesion with areas of marginal calcification arising from the left adrenal region showing intense heterogeneous enhancement with contrast; with nonenhancing areas representing necrosis. The lesion compresses the upper pole of left kidney and displaces it inferiorly and posteriorly. No e/o fat density in the lesion. No e/o invasion of renal vein or inferior vena cava. No e/o ascites or adenopathy. The rest of the organs were normal.

As the parents were not affordable, MRI could not be done.

The child’s blood investigations revealed normal renal functions and electrolytes; serum DHEA = 1020 µgm/dl; serum testosterone = 1540 ng/dl; serum cortisol = 11.61 µg/dl. Tumor was found to be secreting DHEA and testosterone.

So a diagnosis of adrenocortical tumor was made. Since the tumor was limited to adrenal, total excision of the tumor was planned. The child was started on nifedipine and hypertension was controlled (B.P: 102/66 mm Hg) before surgery. Total excision of the adrenal tumor was done by a transperitoneal approach.

The gross appearance was a piece of grayish white tumor partially encapsulated measuring 5.5 x 5 x 3 cms. Microscopic examination of the sections showed encapsulated tumour composed of sheets of large polygonal cells with abundant intensely eosinophilic cytoplasm and central vesicular nuclei with prominent nuclei separated by delicate vascular stroma. Bizarre, multinucleate giant cells seen. Mitosis and necrosis is not seen. Focal calcification is present.
The postoperative period was uneventful. The child was on follow-up. During follow-up, the child should be monitored for regression of virilization symptoms, hormone levels and recurrences using imaging studies.

DISCUSSION

Adrenocortical tumors (ACTs) are rare in children, accounting for only 2 to 12 percent of all childhood cancers\(^1\). The worldwide incidence of ACTs is 0.3 to 0.38 cases per million persons under the age of 15 years\(^2\). The distribution of ACTs according to patient age is bimodal. In the pediatric population, the median age of occurrence tends to be approximately 3 years. The female-male ratio is 2.3:1. 25% of ACTs are adenomas; the rest are carcinomas. Virilization is the most common symptom of ACTs in pediatric patients, occurring in 80% of cases.

More than 50% of ACTs secrete increased amounts of androgens; the rest secrete other hormones. Symptoms include deepening of the voice, acne, hirsutism, increase of muscle mass, and proliferation of sebaceous glands and their secretions, with a characteristic adult odour. Sex-specific changes in females include clitoral enlargement, facial and pubic hair with male escutcheon, amenorrhoea, and occasional temporal balding. In males, penile enlargement and precocious isosexual pseudo-puberty may occur. One third of patients with ACTs exhibit Cushing’s syndrome. Conn’s syndrome is the manifestation of primary aldosteronism; in most cases, the associated pathological disorder is bilateral cortical hyperplasia. Hypertension is associated with approximately 50% of ACTs. This symptom could be explained by an excess of glucocorticosteroids or mineralocorticoids or by the activation of the rennin angiotensin system because of compression of the renal vascular system by the tumor. Non-functional ACTs are uncommon in pediatric patients, accounting for only 5% of childhood ACTs. Because these tumors do not demonstrate endocrine hyperactivity, they usually cause symptoms of abdominal pain or fullness only after they have become quite large or have metastasized. The presence of acne in an infant's presentation should be directed toward the detection of androgens, including the plasma concentration of testosterone, the urinary and plasma conc. DHEA & DHEAS, and the urinary concentration of 17-ketosteroid (17-KS), a nonspecific measure of androgenic metabolites. The most specific assessment of adrenal androgen production is the plasma concentration of DHEAS; an elevation of the urinary concentration of 17-KS almost always indicate adrenal hyperactivity\(^5\).

Patients with Cushing’s syndrome exhibit hypercortisolism and the loss of diurnal variation. These symptoms are confirmed by detection of elevations in the plasma concentration of 17 hydrocortisone and the urinary concentration of free cortisol. Imaging studies are as important as biochemical confirmation.
o f a d r e n o c o r t i-

cal overactivity in the diagnosis of ACTs. The

e studies localize and stage the tumor to fa-

cilitate surgical intervention. CT can detect ad
eral gland tumors as small as 0.5 cm in diameter

and can reliably detect lesions larger than 1 cm in diameter. In the pediatric population, the lack of retroperito
eral fat may pose some difficulty in the detection of ACTs. How-
ever, CT can be used to determine whether tumor thrombus is present in the adrenal or renal vein, the IVC, or the contralateral adrenal gland. CT can also be used to ensure contralateral renal function should ipsilateral nephrectomy be necessary. Only the presence of regional invasion or distant metastatic lesions (liver, lung, or brain) can reliably differentiate benign lesions from malignant tumors on CT images.

It is difficult to distinguish benign ACTs from malignant ACTs solely on the basis of histopathological features, unless patients have obvious signs of malignancy such as metastatic disease or local invasion. The clinicopathologic studies of Hough and Weiss (6,7) attempted to distinguish benign lesions from malignant lesions in adults; however, many ACTs display both benign and malignant morphologic characteristics.

Prognosis is good in pediatric population. Curitiba reported a series of 54 pediatric patients, 56% of whom were disease-free after initial treatment (1). The time to tumor recurrence after surgery ranged from 1 to 48 months (median, 6 months). Recurrences were rapidly fatal; in nearly all cases, patients died within 11 months. Michalkiewicz et al reported 20 pediatric patients with small, surgically resected adrenocortical tumors; the overall survival rate was 90% with a median follow-up time of 2.3 years (8).

To conclude, Virilization is the most common symptom of ACTs in pediatric patients. Palpable abdominal mass was rather uncommon and considered a late manifestation with poor prognosis. CT scan remains the standard accurate modality for their localization and during follow-up. Complete removal of the tumor is the only effective treatment.

REFERENCES


