Adrenocortical Tumor with Endogenous Cushing Syndrome in a Child

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Abstract
Adrenocortical tumors (ACT) are very rare in children with male to female ratio of 2:1. They commonly present with virilization, Cushing syndrome or palpable abdominal mass. Differentiation between adenoma and carcinoma is in practice somewhat arbitrary, and all patients (even those with a seemingly benign adenoma completely resected) require close follow-up initially. On occasions it is recommended to classify the tumor as “atypical adrenocortical neoplasm” or “adrenocortical neoplasm of indeterminate malignant potential.” Factors favoring malignancy include size over 5–10 cm, weight over 200 g, invasion into the periastrine soft tissues or inferior vena cava (IVC). On histopathology there are no reliable features to distinguish between adenoma and carcinoma.

Key Words: Adenoma, Carcinoma, Cushing syndrome, Virilization.

Introduction
Adrenocortical tumors are rare tumors in children. These can be Adenomas (solitary spherical well demarcated unencapsulated tumor of 50 g) or Carcinomas (multi nodular tumors with areas of hemorrhage and necrosis and of more than 100-500g). On histopathology there are no reliable features to distinguish between adenoma and carcinoma.1 Adrenal carcinomas may metastasize to lungs, liver and tumor invasion of IVC, to peritoneum, pleura, diaphragm, abdominal lymph nodes and kidneys. Survival rate of 70% for children <5 years of age and 13% for children >5 years of age. Death occurs within 1-2 years after diagnosis. Here we report a case of 5 year old boy who had adrenocortical tumor with endogenous Cushing syndrome.

Case Report
A 10 months old female infant presented with weight gain, moon like face and fragile skin. On ultrasound examination there was a well circumscribed ovoid hypoechoic mass with lobulated borders and thin echogenic capsule in left adrenal gland. It measured 5 cm and had central areas of necrosis giving heterogeneous appearance. Right adrenal gland and rest of abdomen were unremarkable. She had raised serum glucose level. She underwent hormonal assay which revealed raised cortisol and androgen levels. On CT examination with adrenal protocol she showed a well circumscribed heterogeneous mass with thin rim, variable enhancement and extension into left renal vein and IVC. There was washout of less than 40%. The patient underwent surgical resection of tumor.

Discussion
Adrenocortical tumors (ACT) are very rare in children with a worldwide annual incidence of 0.3 per million children below the age of 15 years.2 The incidence is higher in young girls with a female/male ratio of 2:1,3 whereas in adolescence the sex ratio is equal. Virilization, with early onset of pubic hair, hypertrophy of the clitoris or penis, accelerated growth, gynaecomastia or acne, is the most common presentation. The second most common manifestation is with hypercortisolism (Cushing's syndrome), whilst presentation with a palpable abdominal mass is unusual. Cushing's syndrome is a relatively more common presentation in adolescents and young adults. Very few tumors are non-hormone secreting in contrast to ACTs in adults. Hypertension may be seen in up to 43% at diagnosis.4 This may be due to either mineralocorticoid or glucocorticoid excess, increased aldosterone production or simply renal artery compression by the tumor, and the
hypertension usually resolves after tumor resection. Diagnosis of ACT is supported by raised levels of androstenedione, dehydroepiandrosterone sulphate (DHEAS), testosterone, and urinary steroids. These hormones are also useful markers for the detection of tumor recurrence during follow-up. Two syndromes have a clear association with this tumor; Li–Fraumeni syndrome is associated with mutations of the p53 gene, and Beckwith–Wiedeman which has mutations in the 11p15 region. Ultrasound is the first line investigation and is particularly useful in evaluating for IVC tumor invasion. With a negative ultrasound and the appropriate clinical context, an MRI should also be performed as left adrenal masses in particular may be difficult to visualize with US. If MRI is unavailable CT may be performed. Chest CT to exclude or detect pulmonary metastases should be performed at first diagnosis. The differentiation of benign from malignant ACTs is not clear cut and is the subject of much debate. Differentiation between adenoma and carcinoma is in practice somewhat arbitrary, and all patients (even those with a seemingly benign adenoma completely resected) require close follow-up initially. On occasions it is recommended to classify the tumor as an “atypical adrenocortical neoplasm” or “adrenocortical neoplasm of indeterminate malignant potential”. The majority of ACTs in children are interpreted pathologically as malignant in most studies. Factors favoring malignancy include size over 5–10 cm, weight over 200 g, invasion into the periadrenal soft tissues or IVC. Surgical resection is the mainstay of treatment. Lymph node biopsy should be performed in all cases and radical lymph node dissection may be necessary in some malignant cases. The role of radiotherapy is uncertain. Similarly the role of chemotherapy is limited with many centers using mitotane, as in adult ACTs, but its efficacy in children has not been well studied. Younger children, particularly those less than 5 years, with pathologically malignant ACTs, have a significantly better prognosis than older children and adults.

Conflict of interest
There is no conflict of interest to declare by any author.

References