



Androgen Secreting Adrenal Adenoma – A Case report

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Introduction

Adrenocortical tumors (ACTs) are rare in children, comprising <0.2% of all childhood neoplasms. The most common ACT are adrenocortical carcinoma (ACC) and adrenocortical adenoma (ACA). Benign and malignant tumors of the adrenal gland might be functional or silent. The most frequent clinical presentation is virilization, alone or in combination with hypercortisolism.¹

Pure androgen-secreting adrenal tumors are very rare and their diagnosis represents a clinical challenge. Virilization is characterized by clitorimegaly, hirsutism, male pattern baldness and deepening of the voice.²

Case Summary

A 22 months old girl presented with enlargement of clitoris for one year, pubic hair and excessive body hair for 4 months. She had muscular build, her weight was 14 kg, on 90th centile, height was 86 cm, on 95th centile and normotensive (Fig1).



Fig 1: Muscular build with hirsutism

She had mild hirsutism (Ferriman-Gallwey modified score was 11), clitoromegaly (length was 2.3cm) and pubic hair Tanner stage 2. She had no thelarche, menarche, axillary hair or labial fusion.

Her basal cortisol, basal Adrenocorticotrophic hormone (ACTH) and 17-hydroxyprogesterone (17-OHP) was normal, Dehydroepiandrosterone sulfate (DHEAs) and serum Testosterone was high. Rapid ACTH stimulation test showed normal Cortisol and 17 OHP level. Her investigation findings are as follows:

Hormone	Result	Ref. value
Basal cortisol	259.6 nmol/L	136-690nmol/L
Basal ACTH	11.2 pg/ml	0-46pg/ml
17-OHP	0.17ng/ml	0.1-3.1ng/ml
DHEAs	500.30µg/ dl	19-144µg/dl
Serum Testosterone	6.33nmol/L	0.38-1.97 nmol/L
Rapid ACTH stimulation test		
a) Cortisol	basal- 306.00 nmol/L	at 60 min- 699.00 nmol/L
b) 17-OHP	basal- 0.27 ng/ml	at 60 min - 0.30ng/ml
X-ray bone age	Advanced (5 year)	
USG of abdomen	Left adrenal gland prominent, uterus and both ovaries- present and normal for age.	
CT scan of KUB	Calcified left adrenal mass (5.5 x 3.2cm) and no distant metastasis (lung, liver) was noted	



Fig 2 : Bone age was 5 year



Fig 3: CT scan showed calcified left adrenal mas

Bone age was advanced (5 year), (Fig. 2) CT scan of KUB region showed calcified left adrenal mass (5.5 x 3.2cm) and no distant metastasis (lung, liver) was noted (Fig 3). USG of abdomen revealed Left adrenal gland prominent, uterus present (2.32 x 1.38 x 0.9) normal for age of this child, both ovaries (right ovary-0.88cm x

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0.68cm x 0.76cm) (Lt ovary-1.42cm x 0.79cm x 1.18cm) normal for age of this child.

A left Adrenalectomy was performed, excision of whole mass (4 X 3 X 1.4 CM) was done and no extension to surrounding lymph node or structure was detected.

Histopathology report showed an encapsulated tumor, mixed architecture of zona fasciculata and zona reticularis, the tumor was composed of cells mostly having granular cytoplasm mixed with cells with clear cytoplasm arranged in trabeculae and nests and some of these cells have mild to moderate nuclear pleomorphism (Fig 4). Foci of infarction and dystrophic calcification are also seen and no vascular or capsular invasion was seen. Adrenocortical adenoma was diagnosed.

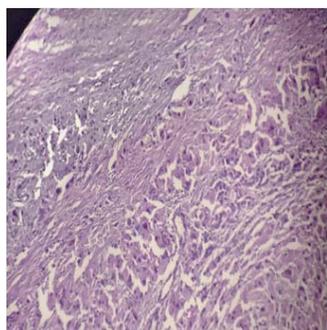


Fig 4: Histopathological slide

One month after surgery the S. DHEAs and S. testosterone were normalized and clinical signs receded.

Discussion

Our female patient began to develop her symptoms at 10 months of age which was consistent with this study of Michalkiewicz et al.¹

ACT most commonly presented with the signs of virilization and/or hypercortisolism (Cushingoid features). Signs and symptoms of virilization were the most common presenting clinical manifestation (>80% of patients).³ Our case presented with features of virilization. About 89.5% from different case series presented with signs of virilization with or without Cushingoid features.^{1,4,5}

Moreno et al. described 21 cases with pure androgen-secreting adrenal tumors among them only 2.4% were due to pure androgen-secreting adrenal tumors in which hirsutism was found in all patients. Tumor size had a mean of 9 cm in the adenomas and DHEAs were elevated in virtually every patient.⁷ In our patient tumor size was 4cm and DHEAs was high.

Cordera et al. reported hypertension was found in 30% of his 11 cases, testosterone level was elevated in 6 cases and DHEAs was elevated in 5 patients.⁵ In our patient blood pressure was normal and testosterone was high.

Our patient presented with puberchy at the age of 10 months. Premature pubarchy in children under four years of age be considered as a feature of ACT until proven otherwise.¹

In the Curitiba series, the height and weight of the children with ACT often exceeded the 50th percentile at the time of diagnosis.¹ Bone age was advanced more than 1 year in 68% of the patients.⁷ In our patient weight was on 90th centile, height were on 95th centile and bone age was advanced. Histopathological examination following adrenalectomy is required for diagnosis. The overall prognosis of adrenal adenoma is excellent, in contrast to adrenal cortical carcinoma.⁸

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