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Virilizing adrenocortical oncocytoma in a toddler

Merna El Khansa [a], Amal Naous [a, *], Nabil Daoud [b], Mariam El Rajab [a]

[a] Pediatric Department, Makassed General Hospital, Beirut, Lebanon
[b] Pediatric Surgery Department, Makassed General Hospital, Beirut, Lebanon

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ABSTRACT

Functioning adrenocortical oncocytomas are extremely rare and most reported patients are 40–60 yr of age. To our knowledge, only 6 cases of functioning adrenocortical oncocytomas have been reported in childhood. We report a case of functioning adrenocortical oncocytoma in a 3 year 4 month-old female child presenting with hirsutism, acne and virilization. She presented with coarse features, deepening of the voice and excessive hair growth, and elevation of plasma testosterone and dehydroepiandrosterone sulfate. She had an adrenalectomy. The completely resected tumor composed predominantly of oncocytes without atypical mitosis and necrosis. A discussion of this case and a review of the literature on this entity are presented.

1. Introduction

An oncocytoma is a tumor that is composed predominantly of polygonal oncocytes with abundant granular and intensely eosinophilic cytoplasm. It occurs in virtually every organ, most commonly the kidney, salivary glands, and thyroid. An oncocytoma that arises in the adrenal gland is very rare and usually benign and nonfunctioning. Because of the unusual occurrence of functioning adrenocortical oncocytoma, particularly in childhood, we report here a case of adrenocortical oncocytoma in a girl presenting with hirsutism, acne, and virilization.

2. Case report

A 3 year 4 month-old girl presented with coarse features, deepening of the voice and excessive hair growth, acne, change in behavior as well as change in body habitus since about 8 month prior to admission. She was born at full term without perinatal problems and had no special health problems previously. No history of consanguinity and no family history of similar symptoms in the female family members. Her mother worried about failure of multiple topical cream for her acne. Her body weight was 18 kg (90th percentile), height was 104 cm (90th percentile) and body mass index was 17 kg/m^2. Her blood pressure was normal. Physical examination revealed coarse features, acne on the face and a male pattern of excessive hair on the chin, thigh, pubic area, axilla (Fig. 1). Her breast was Tanner stage 1. She had moderate clitoromegaly. We performed a hormonal study for evaluation of virilization, which revealed increased plasma testosterone (>18 pg/ml, reference range: 0–1 pg/ml) and dehydroepiandrosterone sulfate (DHEA-S) (702 μg/dL, reference range: ≤260 μg/dL) levels. Plasma 17-hydroxyprogesterone (17-OHP) was also increased (1.1 ng/mL, reference range: 0.1–0.9 ng/mL). Plasma adrenocorticotrophic hormone (ACTH) was 5.1 pg/mL (reference range: 5–37 pg/mL) and plasma cortisol was 3 μg/dL (reference range: 6–22 μg/dL). Plasma lutenizing hormone (LH) was 3 mIU/mL and plasma follicle-stimulating hormone (FSH) was 0.2 mIU/mL. The abdominal computed tomography (CT) scan showed a 4.8 × 4 cm sized well defined left adrenal mass showing internal calcification (Fig. 2). According to the rapid progression of symptoms and virilism, with high level of DHEA-S, testosterone and the presence of adrenal mass, she had a left adrenalectomy, and her postoperative course was uneventful. The resected tumor was a huge encapsulated round mass, measuring 5 × 4 cm in dimension and 1100 gm in weight. The outer surface was smooth and glistening (Fig. 3). Microscopically, the tumor was composed of large cells with eosinophilic cytoplasm. Capsular or sinusoidal invasion was not noted and the subcapsular area showed a compressed normal adrenal cortex. Atypical mitosis and necrosis were absent. These findings were compatible with the diagnosis of oncocytoma (Fig. 4).

A month after the operation, during the first follow-up visit the girl was feeling well. The acne was less expressed and facial hair was reduced. Her voice remained deep. The hormonal evaluation

* Corresponding author.
E-mail address: amalnaous@hotmail.com (A. Naous).

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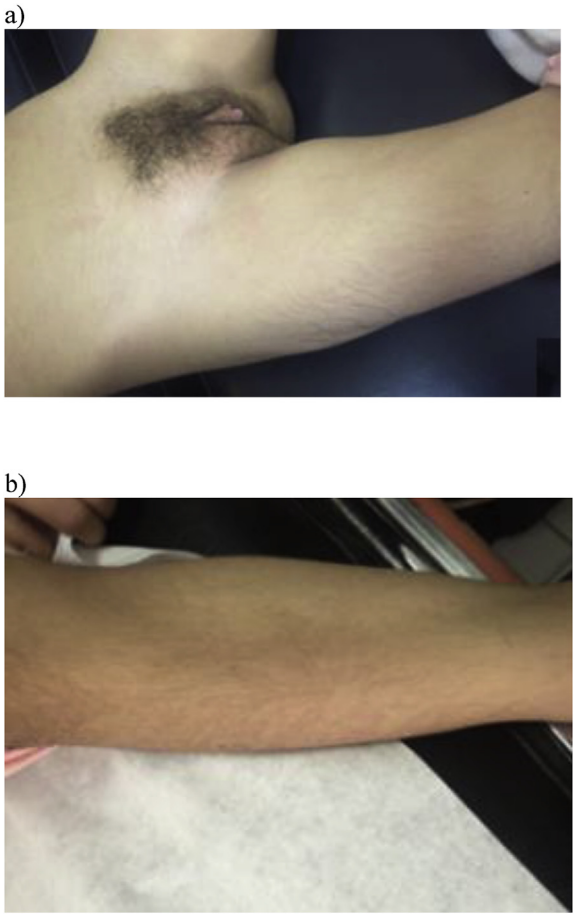

Fig. 1. a) Excessive hair pattern b) Excessive hair in arm and forearm.

revealed normal testosterone and DHEA-S (Table 1).

The ultrasonography of the pelvis and abdomen were consistent with prepubertal stage, without further abnormalities.

On the follow-up visit 12 months after the surgery, the girl's appearance was less masculine, with significantly reduced body hairs but still no changes in the voice (Fig. 5). Hormonal, biochemical and imaging investigations were in the reference ranges for gender and age. Growth velocity was improving (Fig. 6). There was no further progression of pubertal development.

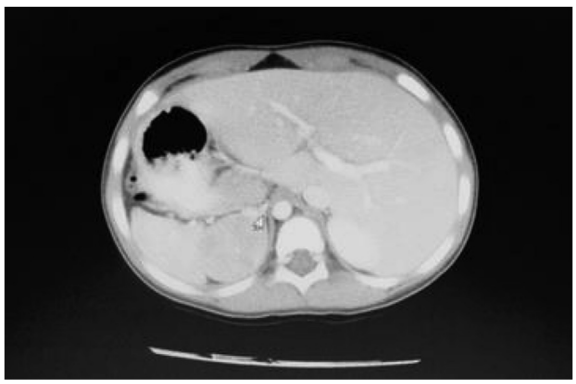

Fig. 2. The abdominal computed tomography (CT) scan showed a 4.8 × 4 cm sized well defined left adrenal mass showing internal calcification.

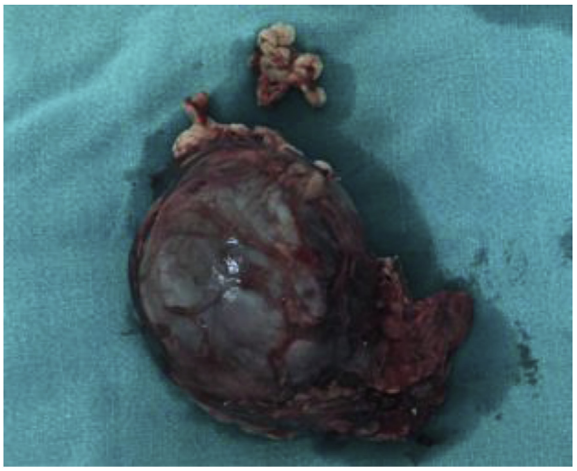

Fig. 3. Macroscopic view.

3. Discussion

Hirsutism is defined as excessive coarse (terminal) hair in a pattern not normal in the female. Virilization is defined as

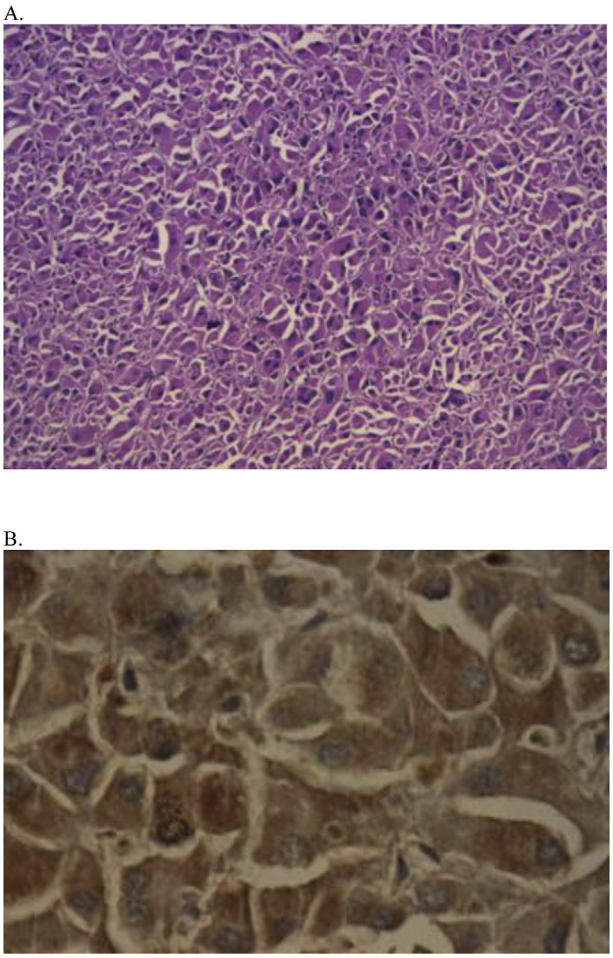

Fig. 4. Microscopic findings of resected tumor. (**A**) The tumor was composed of large cells with eosinophilic cytoplasm. The capsular or sinusoidal invasion was not noted and the subcapsular area showed a compressed normal adrenal cortex. (**B**) Atypical mitosis and necrosis were absent.