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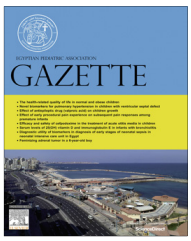

Feminizing adrenal tumor in a 6-year-old boy

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KEYWORDS

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Abstract A 6-year-old boy presented with bilateral gynecomastia and breast tenderness. The condition started since 1 year before medical consultation with gradual onset and progressive course of breast enlargement and accelerated growth with no history of any nipple discharge. There was no history of drug intake or chronic diseases. Investigations showed picture of pseudo precocious puberty with advanced bone age, suppressed LH, FSH and high estradiol (E2). Adrenal precursors (17 OHP, Δ4 A, DHEA, DHEAS) were normal with normal testosterone, cortisol, ACTH. Scrotal U/S was normal while, abdominal U/S revealed right sided hypo echoic supra renal rounded solid mass. Abdominal multi-slice CT with contrast was done and revealed a well-defined hypo dense right adrenal mass with heterogeneous enhancement in the post contrast study showing attenuation of about 25 HU in precontrast and 111 HU in post contrast which makes the mass suspicious of malignancy. Chest X-ray was normal with no lymphadenopathy or pulmonary infiltrates. The diagnosis of feminizing adrenal neoplasm was confirmed and laparoscopic right adrenalectomy was done. Microscopic examination was done after excision and revealed a picture of adrenocortical adenoma with distinct cell borders with no vascular or capsular invasion. The hormonal profile was repeated after 2 weeks of adrenalectomy and revealed normal levels of estradiol (E2), adrenal precursors, FSH and LH. Adrenal tumors can be functional presenting with virilization, Feminization, or Cushing's syndrome. Feminizing adrenal tumors are rare tumors especially in pediatrics, but should be excluded in cases presented with gynecomastia. The differentiation between benign and malignant tumors may be difficult.

Introduction

Feminizing adrenal tumors (FAT) are very rare neoplasms especially in children.[1–3]

Adrenocortical tumors can be functional with varied endocrine syndromes; rapidly progressing Cushing's syndrome, virilization or gynecomastia. When associated with gynecomastia and overproduction of estrogen in men, the term feminizing adrenal tumor is used.[1,3]

They can present with gynecomastia or breast tenderness with or without other manifestations of gonadal deficiency. A huge abdominal tumor may be observed. In children, they induce pseudo precocious puberty (isosexual in females and heterosexual in males).[2]

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In these neoplasms, the differentiation between adenoma and carcinoma is usually difficult.[1,4]

Case presentation

History

A six year old boy presented with bilateral gynecomastia and breast tenderness. The condition started one year before medical consultation by gradual onset and progressive course of breast enlargement and accelerated growth (in the form of height gain mainly). There was no history of galactorrhea or any nipple discharge. There was no history of drug intake or chronic diseases. There were no similar conditions or obesity in the family. Only he had a female sibling with Down syndrome.

Examination

On examination, gynecomastia was the most prominent feature on examination: bilateral, symmetrical, around 10 cm diameter with no focal lesions or palpable masses (Fig. 1a) with no galactorrhea, and no axillary lymph nodes enlargement.

He had pubic hair (Tanner stage P2), prepubertal testis (T3) bilaterally (Fig. 1b) with no axillary hair.

Apart from high BMI, he had no other clinical manifestations of Cushing's syndrome (no hirsutism, no striae with normal blood pressure).

Anthropometric measures: height: 125.6 cm (+2 SDS), weight: 31 kg (+3.5 SDS), and BMI: 19.7 (+2.4 SDS).

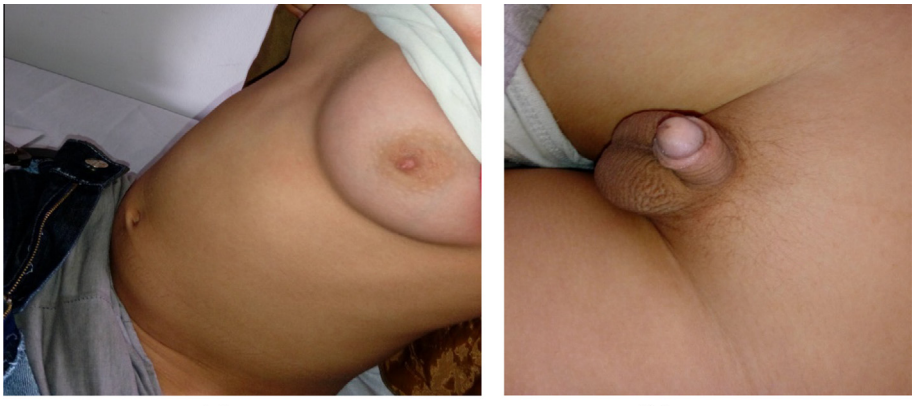
Investigations

The diagnosis of pseudo precocious puberty was suspected, for which the following investigations were done:

- X-ray left hand and wrist for bone age revealed bone age of 8 years (advanced).
- Abdominal and breast U/S showed:
  - Right sided supra renal rounded solid mass; 23 × 18 - mms, hypo echoic related to the upper pole of the right kidney and lies posterior to the liver.
  - Right breast disc 24 × 13 mms.
  - Left breast disc 22 × 14 mms.
- Scrotal U/S (Fig. 2):
  - Both testis of average sizes with homogenous even echotexture.
  - Right testis: 23 × 11 mms.
  - Left testis: 22 × 11 mms.
- Hormonal profile was done to exclude central and adrenal causes of precocious puberty (Table 1).
- LHRH stimulation test revealed suppressed gonadotropins suggesting pseudo precocious puberty:
  - FSH (basal): < 0.1 ml U/ml.
  - FSH (after 4 h): < 0.1 ml U/ml.
  - LH (basal): 0.1 ml U/ml.
  - LH (after 4 h): 0.2 ml U/ml.
- Cushing's syndrome couldn't be ruled out as however, 24 h urinary cortisol was normal (147 μ/d), with normal HbA1c (5.1%), and normal fasting and post prandial blood glucose, serum cortisol was not suppressed by single dose dexamethasone suppression (serum cortisol: 12.3 μ/dl).
- Other investigations revealed normal serum sodium (139 meq/l), normal serum potassium (4.2 meq/l) with normal renal and liver functions.
- Chest X-ray was done to exclude metastasis and was normal with no lymphadenopathy or pulmonary infiltrates.
- Abdominal multi-slice C.T with oral and IV contrast administration was done and revealed: A well-defined hypo dense right adrenal mass measuring 2.3 × 1.7 × 2 cm with heterogeneous enhancement in the post contrast study showing attenuation of about 25 Hounsfield units (HU) in precontrast and 111 HU in post contrast (Fig. 3).

Intervention

The diagnosis of feminizing adrenal neoplasm was confirmed and the decision to do laparoscopic right adrenalectomy was



(a) (b)

Figure 1 It shows bilateral gynecomastia which was the presenting symptom (a), and the pubic hair, tanner stage 2 (b).