A CASE REPORT OF ADRENOCORTICAL ADENOMA IN A YOUNG GIRL

Huyen Tran Thi Bich, Loan Huynh Thoai
Children Hospital 1, Vietnam

INTRODUCTION:

Adrenocortical tumors are rare childhood neoplasms. More than 95% are functional and present with virilization, Cushing's syndrome, hypertension, or hyperestrogenism. We present a exceptionally rare case of a patient with androgen- and cortisol-co-secreting adrenal adenoma.

CASE REPORT:

A 4-year-old girl presented with symptoms of virilization: moustache, pubic hair, gradual enlargement of clitoris for more than 1 year and her voice changed to male pattern deep voice.

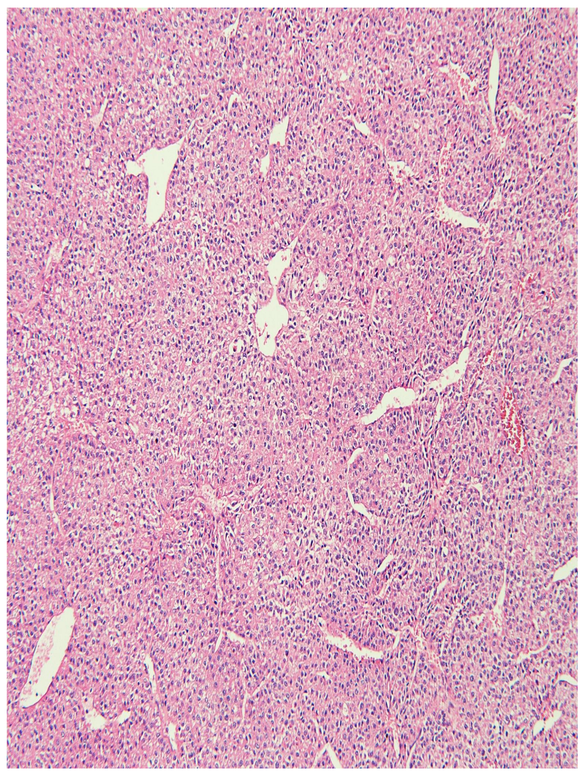
Physical examination revealed normal vital sign. She had no Cushingoid features, no hypertension. Laboratory evaluation was notable for elevated 24 hour urine free cortisol level- 362.7 µg/24h (21-143 µg/24h) - and serum testosteron - 286.5 ng/dl (14-76 ng/dl) - with suppressed serum ACTH - 1.58 pg/ml (7.2-63.3 pg/ml). Levels of DHEAS, 17OH(P), LH, FSH, Estrogen were in normal range. The bone-age was 11 years. Abdominal CT scan showed a left adrenal mass.

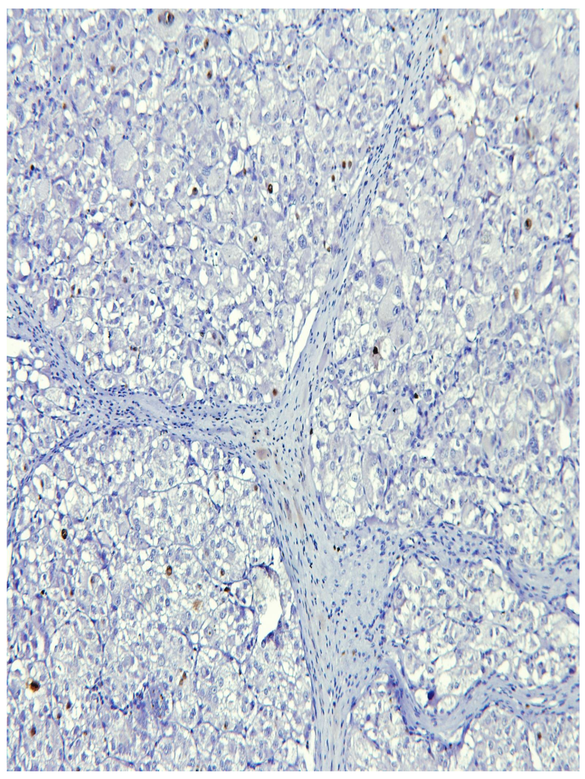
Subsequently left adrenalectomy was performed and histopathology study revealed a 5.5cm x 5cm x 3.5cm adrenal adenoma.

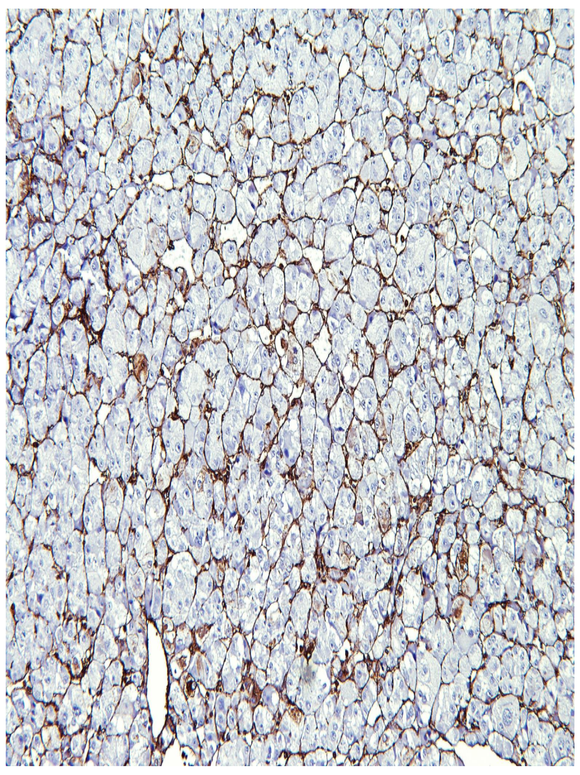
Signs of virilization were suppressed after surgery with serum testosterone levels dropped and remain normal.

At third month after resection, she was noticed to have significant symptoms of adrenal insufficiency, which serum AM cortisol and ACTH levels were low on several occasions. She was treated with hydrocortisone 10mg/m^2/day, withdrew gradually and stop after 3 months.

At sixth month, she had gradual enlargement of breast (Tanner B2), which positive with tests of diagnosis for central precocious puberty. She was given 3.75mg of Diphereline a month.







Stained with HE, Ki67, vimentin, some of mitosis

Laboratory test before and after left adrenalectomy.

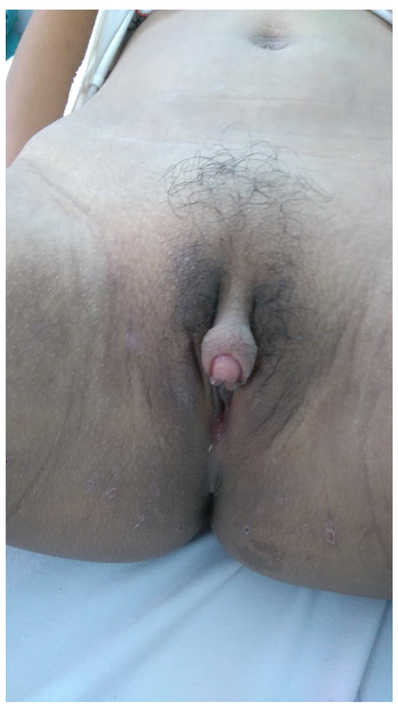
Test	Before	After 3m	After 6m
Plasma cortisol µg/dl	10.72	0.233	2.22
Urine cortisol in 24h µg/24h	362.7	10.25	12.46
ACTH (pg/ml)	1.58	28.72	38.22
Testosteron ng/dl	286.5	7.35	7.25
FSH mUI/ml	0.34	2.1	5.23
LH mUI/ml	0.01	10	10.53
Estradiol pg/ml	39	40.2	50.75
Abdominal ultrasound	Left adrenal mass	No mass	No mass

DISCUSSION:

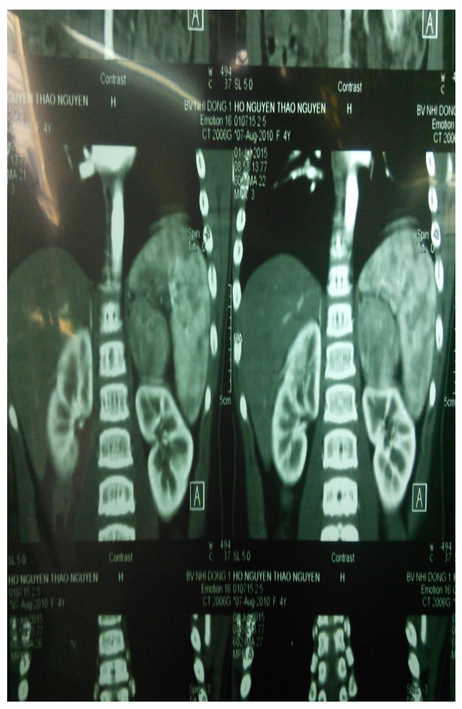
- Pediatric adrenal cortical neoplasm is rare, mostly hormonal hyperfunction.
- Prognostic depend on age and tumor characteristics. There has not yet been specific test for differentiating from benign to malignant. Only in case of metastasis, neoplasm is diagnosed definitely malignant.
- Resection is the main management in order to control.
- Long term follow up is needed for discovering recurrent neoplasm, adrenal insufficiency, central precocious puberty or metastatic.
- Image and hormonal re-evaluation after every 3 months for the first 2 year and prolongs at least 10 years later.

CONCLUSION:

This is a rare case of pediatric adrenal cortical neoplasm causing central precocious puberty. Our case highlights the role of discovery and surgical resection as soon as possible to prevent the untoward effects of virilization or corticosteroid excess. Although the girl in the present study seems to have been cured, long-term follow-up is warranted.



Enlarged clitoris and pubic hair



Left adrenal mass on abdominal CT scan

REFERENCES:

1. R.C. Ribeiro et al (2000): Adrenocortical tumors in children. Brazilian Journal of medical and biological research. 33: 1225-1234.
2. Cristina Eller-Vainicher et al (2010): Post-surgical hypocortisolism after removal of an adrenal incidentaloma: is it predictable by an accurate endocrinological work-up before surgery? European Journal of Endocrinology 162 91–99.
3. Ora Hirsch Pescovitz et al (1985): Central precocious puberty complicating a virilizing adrenal tumor. Treatment with a long-acting LHRH. The Journal of Pediatrics (106): 612-614.

55ESPE 88-P2
Adrenal
Huyen Tran
DOI: 10.3252/pso.eu.55ESPE.2016
Poster presented at: ESPE 2016 PARIS