CASE REPORT OF ADRENOCORTICAL ADENOMA

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INTRUDUCTION: Ardrenocortical neoplasms are rare in children, about 15-20 new cases per year at the age under 20 in America. This rate is 15 folds higher in Brazil population due to mutation in P53 gene. This tumor is common in girl, at 1-4 years old and almost hormone overproduction with virilization, Cushing syndrome and hypertension. Differentiating from benign to malignancy lesion still remains a big obstacle for the clinicians and histopathologist. We report two cases of adrenal cortical tumor in children two different signs and symptoms and review their clinical presentations, pathology and follow-up data.

CASE 1:

Girl, 25 months old, presented with Cushing appearance.

Physical exam revealed abdominal mass and hypertension (140/90mmHg).

Urine cortisol in 24h elevated (727.5pg), plasma ACTH reduced (<1.00 pg/ml).

Right adrenal mass on abdominal CT Scan (Figure 1) and ultrasound. Hypertrophy of her left ventricle on ECG.

Macroscopic anatomy: measured 4,5cm*3cm*2 cm, soft, intacted capsule, no necrosis, no sign of surround invasion. Microscopic anatomy: adrenocortical adenoma (Figure 2).

Three months after tumor resection, she remained hypertension, diffused distinct bounds nodes on chest CT Scan (Figure 3) =>observed for metastasis to the lung. Unfortunately, her family refused to treatment and we lost their contact.

CASE 2:

Girl, 4 years old, presented signs of virilization for more than 1 year.

Physical exam showed enlarged clitoris (Figure 4), men-like pubarche, mustache, dark skin, deep voice, no sign of Cushing, no hypertension.

Elevated in plasma testosterone (286.5 ng/ml), urine cortisol in 24h(362.7 pg) and decreased in plasma ACTH (1.58 pg/ml).

Her bone age was 11.

Left adrenal mass on abdomina CTscan (Figure 5) and ultrasound.

Macroscopic anatomy: measured 5.5cm*5cm*3.5 cm, soft, intacted capsule, no necrosis, no sign of surround invasion.

Microscopic anatomy: adrenocortical adenoma (Figure 6).

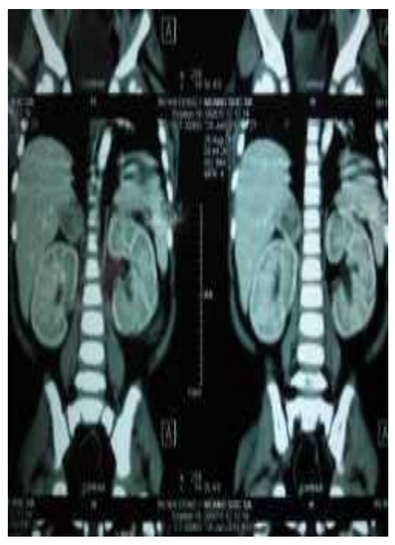

Figure 1: Right adrenal mass on abdominal CT Scan in case 1

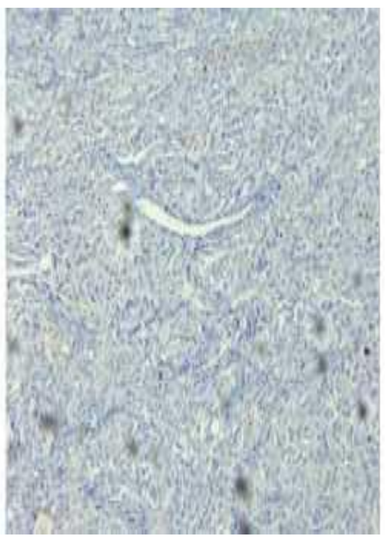

Figure 2: Case 1, stained with Ki67, some of mitosis

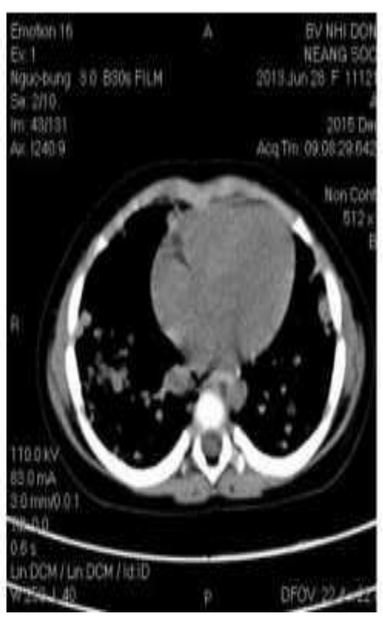

Figure 3: Diffused distinct bound nodes on chest CTscan in case 1

Figure 4: enlarged clitoris and pubarche in case 2.

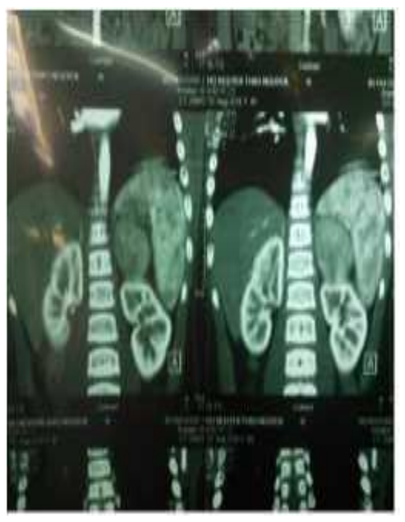

Figure 5: Left adrenal mass on abdomal CTscan in case 2.

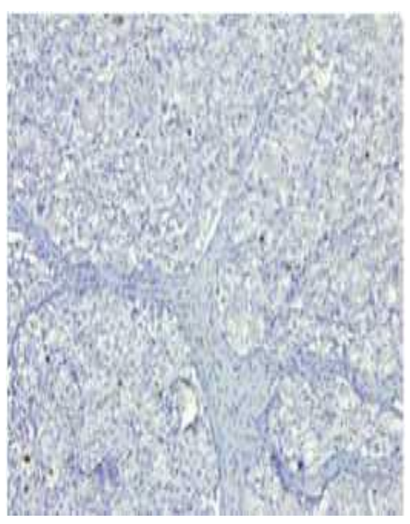

Figure 6: Case 2 stained whh Ki67, some of mitosis..

CASE 2 (cont.):

One month after tumor resection, she suffered from adrenal insufficiency, was treated with hydrocortisone 10 mg/m^2/day, withdrew gradually and stop after 3 months.

Sign of virilization reduced but she had thelarche, positive with central precocious puberty and was treated with Diphereline 3,75mg/month.

Laboratory test before and after unilateral adrenalectomy in Case2

Test	Before	After 3m	After 6m
Plasma cortisol	10.72	0.233	2.22
~~Urine cortisol in~~ 24h	362.7	10.25	12.46
ACTH	1.58	28.72	38.22
Testosterone	286.5	7.35	7.25
FSH	0.34	2.1	5.23
LH	0.01	10	10.53
Estradiol	39	40.2	50.75
Abdominal Ultrasound	Left adrenal mass	No mass	No mass

CONCLUSION:

-Pediatric adrenocortical adenoma is rare, mostly hormonal hyperfunction.

-Prognosis 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... -Image and hormonal re-evaluation after every 3 months for the first 2 year and prolongs at least 10 years later.