

A Rare Cause of Peripheral Precocious Puberty: Adrenocortical Tumor

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Background: Adrenocortical tumor is very rare in the pediatric age group. These tumors may cause peripheral precocious puberty, Cushing's syndrome or both. It is seen most commonly in children under 5 years of age and fourth decade. p53 mutation and other pathologies that may accompany should be investigated especially in young children.

Objective and hypotheses: A 18-month-old boy was brought with pubarche and phallic enlargement, and was noticed 6 months ago for the first time by parents. Parents defined growth spurt and erections. His medical records and family history were unremarkable. Chronological age 1.6 years, height: 86 cm (78 p+0.80 SDS), weight 14.5 kg (95 p), target height SDS -1.65, testicular volume 2 ml/2 ml, stretched penil lenght 6 cm (90-97 p), and pubarche grade 2. There was no other findings of hyperandrogenism. Laboratory results were consistent with peripheral precocious puberty FSH: <0.3 U/l, LH: 0.05 U/l, total testosterone: 473 ng/dl, 17-OH P: 0.5 ng/dl, DHEA-SO₄: 206 µg/dl, and androstenedione: 3.35 ng/ml. Thyroid functions, α-fetoprotein ve βHCG were normal. A mass with 22×17 mm was detected ultrasonographically in right adrenal gland and being 35 Hounsfield Unit, the mass was considered as nonadenomatosis. Tumor weighted 50 g with intact capsula after adrenalectomy. Pathological diagnosis was adrenocortical carcinoma and no metastasis was detected with PET-CT scanning. p53 mutation analysis is being tested. Androgen hormones decreased to prepubertal levels in second day postoperatively. Mass originated from soft tissue was detected third month after adrenalectomy and resected material was consistent with rhabdomyosarcoma histopathologically.

Conclusion: Simple radiologic methods should be selected in peripheral precocious puberty before further complicated investigations. Besides it is noteworthy to be careful for comorbidities such as other malignancy especially in young children.