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Precocious Puberty Associated with an Adrenal Tumor: A Case Report

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Abstract Precocious puberty is a common complaint in pediatric medicine these days with numerous causative factors. Though adrenocortical tumors are extremely rare in children, the differential diagnosis is broad with androgen producing adrenal adenoma. Pediatric adrenocortical tumors are therapeutic and diagnostic challenges because histological criteria for distinguishing benign from malignant tumors seen in adult are not always reliable in children. Here, we report a 2 year old toddler with precocious puberty with hyperglucocortism and hypertension. His plasma testosterone, dehydroepiandrosrerone and cortisol were elevated. An abdominal MRI and CT scan identified well defined left –sided adrenal mass. Left adrenalectomy was performed and histopathological examination revealed adrenal adenoma. This case emphasizes the importance of early consideration of an adrenocortical tumor causing precocious puberty in toddler.

Keywords: *adrenal tumor, adrenocortical carcinoma, precocious puberty, virilization*

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1. Introduction

Assessment of precocious puberty is complex and cancer is often underestimated. Primary organ lesion included brain tumor (pineal or optic pathway/hypothalamic tumor), adrenocortical tumor, hepatoblastoma and ovarian tumor [1,2]. Adrenocortical tumor is an extremely rare neoplasm in childhood with an annual incidence of approximately 0.3-0.5 cases per million children under 15 years [3]. Adrenocortical tumors in children are frequently hormone-secreting tumors, and precocious puberty alone or in combination with sign of overproduction of adrenocortical hormones is the most common endocrine syndrome [4], on other hand in adult they are silent and manifested in form of pressure symptoms and signs. The presentation of an adrenal adenoma can vary, with isolated androgen production causing precocious puberty being an uncommon presentation. We report a case of precocious puberty due to adrenal adenoma.

2. Case Report

A 2 year old male toddler presented to OPD with early development of pubic hair and increasement penile length & size. He had history of excessive hair on face and other body part (Figure 1). He was born at 38 weeks of gestation, second of birth order through vaginal delivery with birth weight 2800gms without any h/o antenatal maternal drug intake or radiation exposure, with uncomplicated postnatal period. There was no history of radiation exposure, exposure to drugs and herbal products, vision problem, headache, head trauma, cold or heat intolerance without past surgical history. None of his family members had endocrine tumors or genetic diseases. On physical examination, his height &weight was more than expected for his age. His vitals were stable except hypertension. He had hepatosplenomegaly without dysfunction and Tanner stage was III.

The patient was subjected to serum and urinary laboratory test. Random serum cortisol, early morning serum cortisol & urinary cortisol were elevated, serum ACTH was normal. The serum testosterone and serum dehydroepiandrosterone sulphate were elevated. Follicle stimulating hormone (FSH) and luteinizing hormone (LH) were below the limit of detection. Thyroid function test, lipid profile, serum ionized calcium, kidney function test, liver function test and hemogram were within normal limit except he had "AS" pattern on hemoglobin electrophoresis.

His bone age was 4 years based on the Greulich-Pyle method. Abdominal ultrasound, MRI and computed tomography demonstrated a left well defined heterogeneous mass measuring 3.6cm×2.4cm×3.7cm without enhancement, calcification or hemorrhages (Figure 2A, 2B & 2C). Few small mesenteric, precaval and aortocaval lymph nodes were noted. The brain, lungs, bones, liver and spleen were free from metastases on magnetic resonance imaging and high resolution CT. A provisional diagnosis of adrenocortical tumor was made and patient was posted for surgical removal after hypertension was in control. Grossly, there was no metastasis to regional lymph nodes or vessels detected during the operation. The adrenalectomy specimen was partially encapsulated and soft, measuring 4.5cm×4cm×2cm and weighing about 25gms. The cut

surface was homogenous, solid, smooth grey white & boaselated with no evidence of cystic changes and necrosis but tiny hemorrhagic area were noted. On microscopy, the capsule was thick, fibrocollagenous and beneath the capsule solid, diffuse sheets of tumor cells were noted. The individual cells were large, polyhedral with well defined cell margin, cytoplasm was abundant, dense eosinophilic, nucleus was large centric with vesicular chromatin and prominent nucleoli. Bizarre cells were noted. There were no evidence of mitosis and no evidence of capsular or vascular invasion (Figure 3A & 3B).

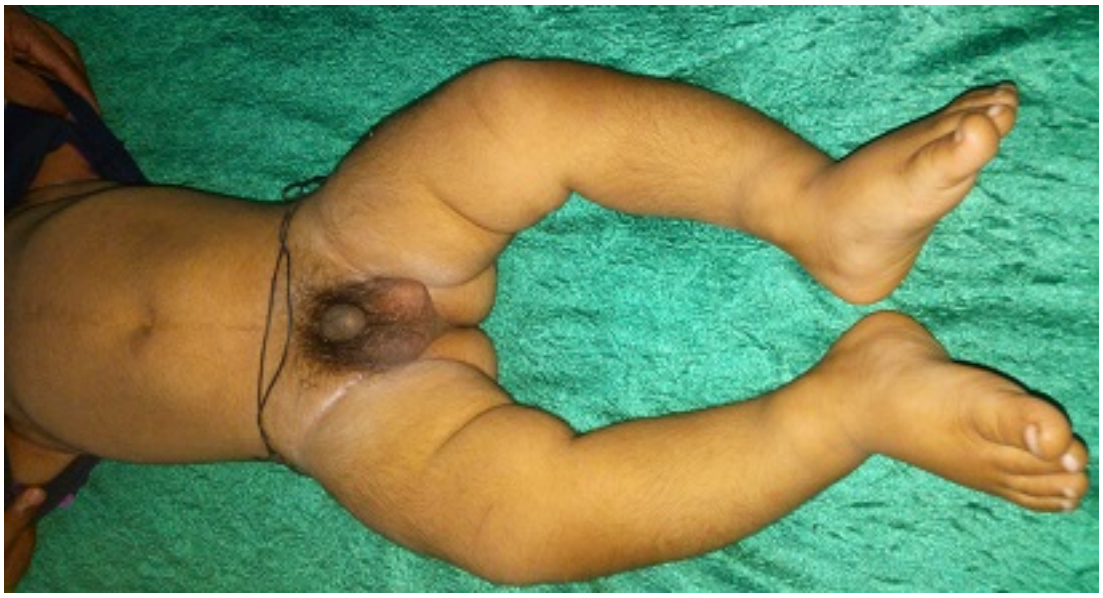

Figure 1. Shows external genitalia shows Tanner stage III & excessive hair on thighs

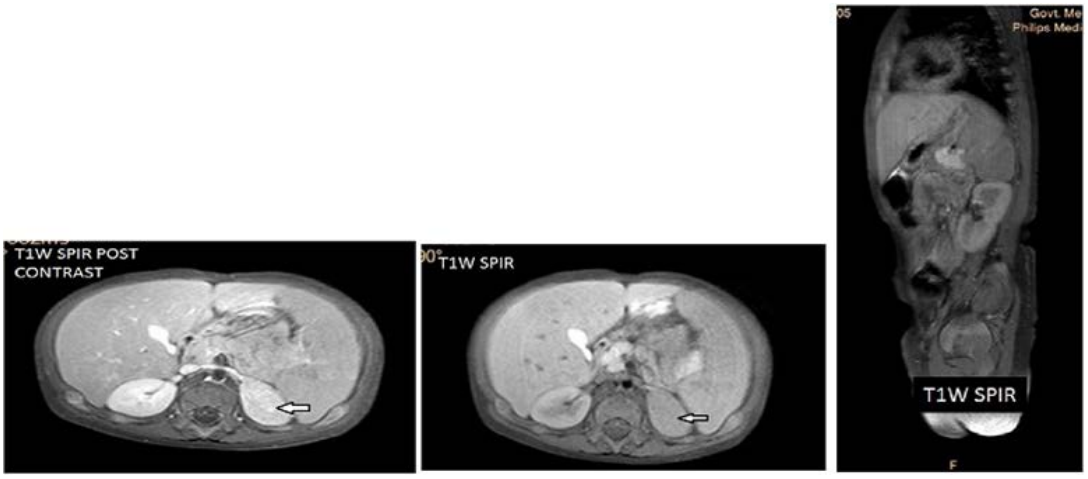

Figure 2. MRI showing tumor mass (arrow) with mixed signal intensity

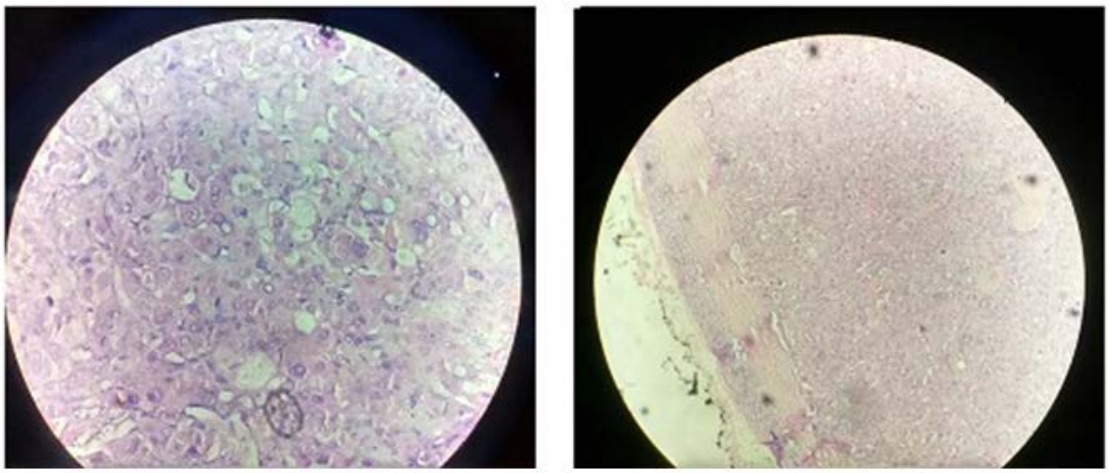

Figure 3A. Shows high power field diffuse sheets of polyhedral tumor cells, B. Shows microscopic pictures of adrenocortical adenoma

3. Discussion

Adrenocortical tumor has peculiar clinical and biological features that contrast with those observed in other pediatric carcinomas. Only about 25 new cases in patients younger than 20 years are diagnosed per year in the USA. Higher incidences of pediatric adrenocortical tumors occur in Brazil and are slightly more common in females [5]. There is paucity of such data in our country.

In contrast to adrenocortical tumors in adults, majority of tumors (95%) in children are hormonally active and secretes varying amount of glucocoticoides, minerolocorticoides and sex steroids or an excess of any one of these. Our patient have had also signs of hyperglucocortism, hypertension and hirsutisim Upadhya et al reported 64% patient presented with signs of hyperglucocortism, 36% with adrenogenital syndrome 71.4% with hypertension [6]. Virilization is an important clue to the diagnosis of ACT [7]. This was the features in present case and his hormonal assay confirmed functioning adrenocortical tumor. This is consistent with other reports in the literature [6,7]. Razavi et al reported 10 months old male infant with appearance of sexual pubic hair, hirsutisim, acne and seborrheic dermatitis [4]. Shenge et al reported 4.5 years old girl of adrenocortical adenoma mimicking congenital adrenal hyperplasia with appearance of pubic hair & deep voice [8]. Maji et al also reported 40 months old female with virilizing adrenal adenoma causing precocious puberty and masculine features [9]. Additionally, the histological criteria for distinguishing benign from malignant tumors used in adults are not always reliable in children. Based on previous related studies, tumor size is the most important prognostic factor [5,10].

Appropriate treatment remains a continuing challenge in pediatric ACT. Radical excision with enbloc resection of any local invasion offers the best chance for cure and long term survival. Adjuvant therapy, both radiation as well as chemotherapy have been disappointing [9]. The overall prognosis of adrenal adenoma is excellent, contrast to adrenal carcinoma [3].

4. Summery and Conclusion

Our patient was a rare case of adrenal adenoma with precocious puberty, hyperglucocortism and hypertension. Awareness about the occurrence of this uncommon tumor often with a dramatic onset, early diagnosis and prompt surgical intervention are the only means to salvage such patients with probable malignant potential.

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