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Original Research Paper

Pathology

ADRENAL CORTICAL ADENOMA PRESENTING AS PRECOCIOUS PUBERTY IN A 7-YEAR-OLD GIRL- A CASE REPORT

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ABSTRACT Childhood adrenocortical tumors (ACT) are rare entity. Because of the heterogeneity and rarity of the disease, prognostic factors have been difficult to establish in pediatric ACT. Most of the affected children are young girls with classic endocrine syndromes (virilizing and/or Cushing). The other presentation can be Conn syndrome, feminization and precocious puberty. Typical imaging findings of pediatric ACT consist of a large, well-defined suprarenal tumor containing calcifications with a thin capsule and no necrosis or hemorrhage. The pathologic classification of pediatric ACT is troublesome. Even an experienced pathologist can find it difficult to differentiate carcinoma from adenoma. In patients with localized and completely resected tumors, the size of the tumor has predictive value. Patients with large tumors (>500g) tend to behave aggressively with much higher relapse rate than those with small tumors. It is important for the clinicans and pathologists to be aware of these because of varied presentations.

KEYWORDS : Adrenocortical tumors, precocious puberty, adenoma,carcinoma, Feminization

Introduction

Adrenocortical tumors (ACT) are a heterogeneous group of benign neoplasms. Although, these are rare in children, they are important causes of inappropriate virilization and cushing's syndrome in childhood. The first case of childhood ACT was reported in 1865.[1] Childhood ACT has peculiar clinical and biological features that contrast with those observed in other pediatric neoplasms. The incidence of most childhood carcinomas increases with age, whereas 65% of ACT occur in children younger than 5 years of age.[2]

In fact, this age distribution resembles that of tumors of embryonic origin. Moreover, most children with ACT appear healthy and usually have normal development. Finally, smaller-sized ACT can cause exuberant endocrine syndromes that often mask or delay the diagnosis [2]. The presenting manifestations of ACT depend largely on the specific tumor's secretion of adrenocortical hormones. Hormone-secreting tumors and the associated classic endocrine syndromes (virilizing, feminizing, Cushing's and Conn's syndromes) represent the most common presentation in this age group. Although the clinical manifestations of one endocrine syndrome may predominate, ACT usually secrete several hormones and thus present signs and symptoms of multiple syndromes (mixed forms). Nonfunctional tumors, the most common type in adults, comprise approximately 10% of the pediatric/ adolescent cases.[3]

Levels of urinary 17-ketosteroids and plasma dehydroepiandrosterone sulfate (DHEA-S), which are abnormal in approximately 90% of the cases, provide the pivotal clue to a diagnosis of ACT. To avoid delaying diagnosis of ACT, any child less than 4 years with pubarche should be considered to have an ACT until proven otherwise. In addition, Cushing's syndrome is very rare in children, it should be considered highly indicative of ACT in children younger than 10 years of age.[4]

A rare case of adrenal adenoma is being reported in a 7-year-old girl.

Case Report

A 7-year-old girl presented with normal intelligence for her age presented with clinical features of pubic hair, Acne and rapid growth at pediatric OPD. Clinical diagnosis of precocious puberty was made. Initial work up did not reveal any biochemical / hormonal imbalances. On further CT scan whole body, a hypointense, encapsulated, marginated adrenal mass with uniform attenuation was noted. A diagnosis of an adrenal tumor was offered and the patient underwent adrenlectomy. Post operative period was uneventful.

A well circumscribed mass was received in the laboratory measuring 5 x 3 x 3 cm weighing 40 gm. Cut section revealed greyish yellowish homogenous surface. No areas of hemorrhage and necrosis noted. (Fig.1) Representative sections were taken and microscopy revealed an encapsulated tumour demarcated from the adjacent cortex by a pseodocapsule. Tumour displayed sheets of loosely cohesive atypical cells in small nests, vague alveolar pattern. Tumour cells had vacuolated (clear) cell morphology, having a large size than normal cortical cells, abundant clear eosinophilic cytoplasm, round, eccentric nuclei with vesicular chromatin and small distinct nucleoli. (Fig 2). No necrosis, venous/capsular invasion or increased mitosis seen. A differential diagnosis of adrenal cortical hyperplasia, adrenal cortical carcinoma & pheochromocytoma was entertained.

Fig 1: Gross Examination(A), showing a well circumscribed lesion measuring 5X3X3cm.& Cut Surface(B) with a homogenous surface.

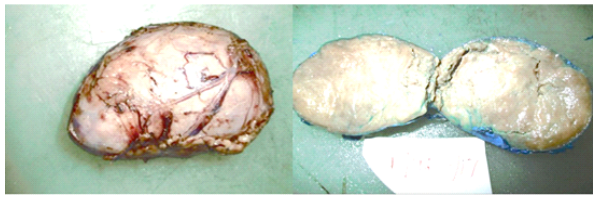
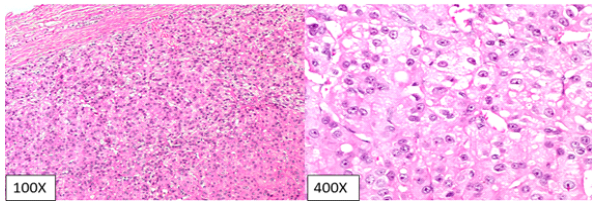


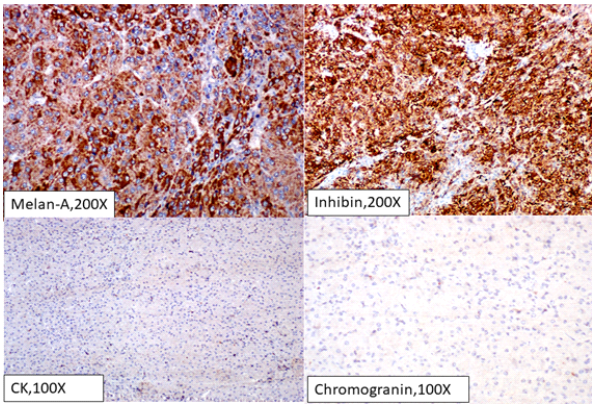
Fig 2 A& B : Microscopic Examination H &E, showing a well circumscribed tumour. B(400X), reveals presence of cells in nests and alveolar pattern. C



Immunohistochemistry revealed tumour cells to be positive for Melan A and Inhibin A. Tumour cells were negative for CK and chromogranin (ruling out Pheochromocytoma/ Paraganglioma). (Fig 3). The diagnosis of adrenal cortical adenoma was made based

upon histomorphological features and immunohistochemistry.

Fig 3 : Immunohistochemistry showing tumour cells positive for Melan A, Inhibin, and negative for CK & Chromogranin.



Discussion

Adrenocortical tumors are divided into benign and malignant groups. Either can be hormonally silent or hormone secreting. The vast majority of adrenocortical tumors are benign and hormonally silent. The frequency of small benign adrenocortical tumors gradually increases with age, ranging between 3–7% in adults over 50 yrs. Usually they are discovered incidentally, in the context of abdominal computed tomographic (CT) or magnetic resonance imaging (MRI) scans performed for various unrelated purposes. Hormone-secreting benign adrenal adenomas are rare; equally rare are hormonally silent or hormone-secreting adrenocortical carcinomas. Adrenocortical tumors can be sporadic or hereditary, with the former representing the overwhelming majority.[1,2]

Patients with hormone-secreting adrenocortical neoplasms have associated endocrine syndromes that result from the secretion of cortisol or aldosterone and their precursors, adrenal androgens and their precursors, and/or estrogens. The most common syndromes associated with adrenocortical tumors in adults are hyperaldosteronism and Cushing's syndrome.[5]

Children and adolescents with a functioning ACT are subject to growth disturbances.[2] Virilization occurs in 20–30% of adults with functional adrenocortical carcinoma, whereas it is the most common hormonal syndrome in children with adrenocortical cancer.[6]Virilization is secondary to hypersecretion of adrenal androgens, including dehydroepiandrosterone and its sulfate derivative,Δ^5-androstenediol, and Δ^4-androstenedione, all of which may be converted finally to testosterone and 5α-dihydrotestosterone. Feminization and hyperaldosteronism, as pure hormonal syndromes, are quite rare manifestations of malignant adrenocortical neoplasms. A few incidences has been reported in the literature wherein functional adrenal cortical adenomas has been linked to the precocious puberty in young child mostly females.[6] The signs and symptoms in adult females include oligoamenorrhea, hirsutism, cystic acne, excessive muscle mass, deepening of the voice, temporal balding, increased libido, and clitoromegaly. In young girls, heterosexual precocious puberty occurs. Puberty that begins before age 8 in girls and before age 9 in boys is considered precocious puberty [1].

This case is presented of a 7 year old girl with a functioning cortical adenoma weighing 50 gm who presented with rare clinical presentation of precocious puberty.

Conclusions

Adrenal Cortical tumours are rare and have varied presentations in children. They are important causes of inappropriate virilization and cushing's syndrome in childhood. Clinicians and pathologists need to be aware so as to diagnose them early. Immunohistochemistry plays an important role in diagnosing these tumours.

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