

ADRENOCORTICAL TUMORS IN CHILDRENKannepalli Srinivas¹**HOW TO CITE THIS ARTICLE:**

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ABSTRACT: Adrenocortical tumors rare in children which are 0.2% of all Pediatric neoplasms. These are mostly Occurs less than 10 yrs. Most of pediatric (Endogenous) Cushing's syndrome cases are caused by adrenocortical carcinomas and 20-40% of adrenocortical tumors have Cushing syndrome in addition to virilization. In present study we have treated 5 patients of adrenocortical tumors. All are left sided functional benign adrenocortical adenoma except one (Carcinoma). After resection of Tumors the clinical Features of Cushing's & Hypertension were subsided.

KEYWORDS: Adrenocortical Tumors, Cushing syndrome Virilization.

INTRODUCTION: Adrenocortical tumors rare in children .0.2% of all Pediatric neoplasms, Occurs mostly in less than 10yrs of age. Most of pediatric (Endogenous) Cushing's syndrome cases are caused by Adrenocortical carcinomas.^(1,2) 20-40% of Adrenocortical tumors have Cushing syndrome in addition to virilization.⁽³⁾ The tumors mostly occur in girls. The tumors present with benign nature in most cases and surgical excision of tumor is the mainstay of treatment which relieve all the symptoms & signs.

MATERIALS AND METHODS: This is a prospective study for a period of Five years from 2009 to 2014 conducted in the department of Pediatric Surgery at a tertiary care Centre. Five children with functional adrenocortical tumors have been detected. All the patients presented with Cushing's features with hypertension. Out of Five three patients had virilisation symptoms.

Serum testosterone, estradiol, 17(OH) progesterone, cortisol and 24-hour urine cortisol levels were measured using commercial kits. Ultrasonography and computed tomography (CT) were used for localization of the tumor. Diagnosis was based on clinical manifestations and evaluation of steroids in serum or in 24-hour urine samples.

All patients underwent open surgery except one. During surgery, a blood sample was collected from the effluent adrenal vein for evaluation of testosterone, estradiol, 17(OH) progesterone, and cortisol levels in all cases. In addition, a simultaneously collected peripheral vein sample was evaluated for the same hormonal profile. Histopathologic evaluation was done. Written informed consent was obtained from the parents.

RESULTS: Clinical characteristics of the patients are Cushing's Features, Virilization, Precocious puberty, Hypertension, Non-palpable Abdominal mass. The subjects were 3 girls and 2 boys aged 5months to 8 years. Duration of disease ranged from 3 months to 5 months. One patient died preoperatively. The patient follow-up ranged between 1 and 6 years. Signs and symptoms of virilization including temporal hair loss, hirsutism, pubic hair growth, cliteromegaly, phallic growth, voice deepening and acne were the prevailing manifestations seen in 3 of the patients. Five patients presented with Cushing's syndrome. All patients had advanced bone age and hypertension.

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Results of laboratory evaluation of the patients at diagnosis are serum testosterone, dehydroepiandrosterone sulfate (DHEA-S), estradiol, 17(OH) progesterone and cortisol levels. All were elevated.

The tumors weighed 40-90 grams and were located in the left adrenal gland in all cases except one. The tumors were encapsulated and a rim of compressed atrophic adrenal tissue surrounded the tumors. Microscopic evaluation, using hematoxylin-eosin staining, showed hyper vascular tumors composed of lipid laden, various sized cells.

DISCUSSION: In this paper, we attempted to portray a comprehensive clinical and laboratory picture in children with functioning adrenal tumors.

Adrenocortical tumors comprise 0.3-0.5% of the neoplasms detected in patients under the age of 10.^(1,2) In two previous large reports on 209 and on 254 pediatric and adolescent cases, the most common presentation was virilization followed by a mixed picture of virilization and Cushing's syndrome.^(4,5) The disease is more prevalent in girls, as seen in our report as well.

Tumors are functionally active in 95% of cases with overproduction of androgens, corticosteroids, aldosterone and estradiol in decreasing order of frequency. Sometimes, the tumors are polyhormone secretors or their secretion pattern changes.

In our series, most of the tumors were located in the left adrenal. It seems that adrenocortical tumors are more prevalent in the left side,⁽⁶⁾ Histopathologic diagnosis was benign adrenocortical adenoma in all cases except one.

Our findings are in accordance with data reported by other investigators who showed that in childhood functioning adrenocortical tumors, the size and weight of the tumors are more important factors in determining the benign or malignant nature of the tumor. Indeed, tumors weighing less than 90grams have a benign nature and a favourable prognosis.⁽⁷⁾

In some studies, functioning adrenocortical tumors are reported to have a good prognosis.⁽⁶⁾ This finding may be just due to earlier diagnosis because of earlier clinical manifestations secondary to hormone over secretion.⁽⁸⁾

In conclusion, this study supports that the childhood adrenocortical tumors commonly present with clinical features that should be immediately diagnosed and surgically treated to avoid untoward complications. Thorough evaluation of hormonal profile, irrespective of hormone-related clinical features, will reveal the usually multi-hormonal secreting nature of the tumors.

The assessment of adrenal venous effluent hormones has shown that hormones are directly secreted from the tumor and peripheral conversion has little contribution to peripheral hormone levels. Features of Cushing's & Hypertension subsided after resection. Pre, intra, postoperative management is crucial for survival.

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