



Case Report

Adrenocortical Carcinoma in a 3 yrs Girl—A Case Report

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Abstract

Adrenocortical carcinoma is an uncommon tumour in the pediatric population. Account for only a small fraction of pediatric adrenal tumour. Most tumors in children are functional, and virilization is by far the most common presenting symptom, followed by Cushing's syndrome and precocious puberty. All patients with suspected adrenocortical carcinoma should be carefully evaluated for signs and symptoms of hormonal syndromes.

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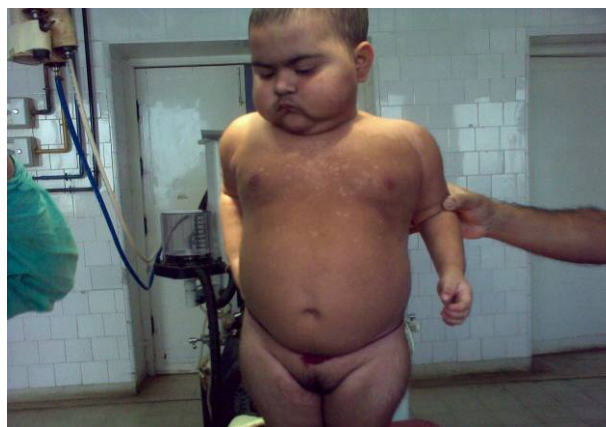
Introduction

Adrenocortical carcinoma may present differently in children and adults. For Cushing's syndrome (glucocorticoid excess) these include weight gain, muscle wasting, purple lines on the abdomen, a fatty "buffalo hump" on the neck, a "moonlike" face, and thinning, fragile skin. Virilism (androgen excess) is most obvious in women, and may produce excess facial and body hair, acne, enlargement of the clitoris, deepening of the voice, coarsening of facial features. The main etiologic factor of adrenocortical cancer is unknown. Families with Li-Fraumeni syndrome have increased risk. It may be due to abnormality in chromosome.

Case Report

A three years old girl presented to department of pediatric surgery with symptoms and signs of Cushing's syndrome with virilism. First it is the mother who felt the mass in the left upper abdomen for about six months back. On ultrasound revealed inhomogeneous mass noted in

the left upper abdomen in the region of left adrenal gland extends downwards anterior to the left kidney. Multiple axial non-contrast and contrast CT-scan were performed. Which suggest malignant left adrenal mass. Hormonal study revealed high cortisol level to cause of Cushing's syndrome. So we decide to do laparotomy and excision of the mass is done. After histopathology it reveals adrenocortical carcinoma.



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Discussion

Adrenocortical carcinoma is a rare malignant tumor with an estimated incidence of 1 in 1.5 million people.¹ Its etiology is unknown, but it may occur from de novo neoplastic changes or arise from preexisting adrenal pathologies, such as congenital adrenal hyperplasia and neoplastic transformation from chronic adrenocorticotropic hormone (ACTH) stimulation.² Although it is unknown whether there is a definite familial predisposition, an unexpectedly high incidence of congenital anomalies, hamartomas and secondary tumors has been reported in those with adrenocortical carcinomas. Adrenocortical carcinoma can occur at any age but is most common in children before the age of 5 years (mean age, 4.63 years) and in the fifth to seventh decades of life. In an effort to define the epidemiology of adrenocortical carcinoma, Kufe and Benedict² found 1891 pediatric and adult cases among 87 studies in the English literature (from 1952 to 1992) and added 8 additional patients. Females were affected slightly more often (4:3) and presented at a lower median age (median, 30 years) than did males (median, 39 years). The left adrenal was involved 52.8% of the time, and few cases (2.4%) were bilateral. Fifty-nine percent were functional tumors (i.e., cancers producing adrenal hormones that manifest clinical symptoms). Functional tumors were more common in females (7:3), whereas nonfunctional

tumors were more often found in males (3:2). Nonfunctional tumors were seen with much greater frequency in older patients, regardless of sex. Three percent were stage I (smaller than 5 cm and confined to the adrenal); 29%, stage II (larger than 5 cm and confined to the adrenal); 19%, stage III (loco regional disease with positive nodes and/or local invasion); and 49%, stage IV (distant metastasis).

Functional adrenocortical carcinomas are more common in children is correct. Based on whether adrenocortical carcinomas are hormonally active and manifest clinically, they can be classified as either functional or nonfunctional. It is found that fewer than 50% were overtly functional in adults, as opposed to approximately 95% in children. Kasper⁴ also found that functional tumors were more common in the pediatric population, regardless of gender. Strictly speaking, functional tumors produce abnormal amounts of adrenal hormones on routine tests and nonfunctional tumors do not, but the literature is often not definitive on what constitutes "routine" tests. Nonetheless, with more sensitive tests, nonfunctional tumors may demonstrate production of steroid precursors that generate minimal or no hormonal activity.⁴

References

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