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Case Report

Functioning adrenocortical carcinoma causing virilisation: a case report

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ABSTRACT

Adrenocortical carticnoma (ACC) is an uncommon tumor with an incidence of 1-2 cases/million/year. It has two peak incidences; the first one in the first decade and the second one in the fourth decade. Most patients present with features of steroid hormone excess or abdominal mass effects, but about 15% of ACC are diagnosed incidentally. It is hormonally functional in 80 - 100% patients and the predisposing lesions include congenital adrenal hyperplasia and adenoma. ACC has significant syndromic and genetic association. Surgery offers the best chance of cure, especially in localized disease. Here, we present the case of virilization in a young female child secondary to a functioning ACC. The child had classical hormonal and imaging features of functioning ACC and underwent resection of the tumor (Adrenalectomy) with good outcome.

keywords: Adrenal adenoma, Adrenal tumor, Adrenalectomy, Adrenocortical carcinoma, Clitoromegaly virilisation

INTRODUCTION

The onset of virilisation in a child who was previously asymptomatic, is a significant event which requires detailed and systematic evaluation. A functioning Adrenocortical carcinoma (ACC) is one of the most important differential diagnosis, especially in a child. Adrenocortical carticnoma (ACC) is an uncommon tumor with an incidence of 1-2 cases/million/year.[1] It has two peak incidences; the first one in the first decade and the second one in the fourth decade.[2]

Most patients present with features of steroid hormone excess or abdominal mass effects, but about 15% of ACC are diagnosed incidentally.[2,3] It is hormonally functional in 80 - 100% patients and the predisposing lesions include congenital adrenal hyperplasia and adenoma.[3,4] ACC has significant syndromic and genetic association.

CASE REPORT

A 3yr old girl presented with precocious development of pubic hair since 2 years of age. There was no significant antenatal or postnatal history. The positive findings were hirsutism, muscular appearance and deep voice. Male pattern distribution of pubic hair was present and the external genitalia showed clitoromegaly with no palpable gonads. Vaginal and urethral orifices were normal (Figure 1).There was no hyperpigmentation or thelarche. BP was normal for age and there were no clinical features of Cushing syndrome. Abdominal examination revealed no palpable mass or organomegaly.

Investigations

Hemogram was normal. Serum Electrolytes and Renal function tests were normal. Urine 17 ketosteroids were elevated (29mgm /24hrs - Normal : 7-20). Urine VMA

was 2.3mg /24hr (Normal : 6 - 10.5). S. DHEAS was elevated (1740ng /dl ; Normal : 280 - 750). S.17-OH Progesterone was elevated - 320 ng /dl (Normal <100). S. Costisol was normal and Karyotyping showed 46 XX.

Figure 1: Clinical examination showing clitoromegaly and pubic hair. There were no palpable gonads.

USS abdomen showed an 8 x 8 cm mass lesion, possibly arising from Left Adrenal gland. CT abdomen showed a heterogenous soft tissue mass lesion located between the left kidney, spleen and tail of pancreas, arising from left Adrenal measuring 8 x 10 cm with no infiltration into surrounding structures and no para-aortic nodes (Figure 2).

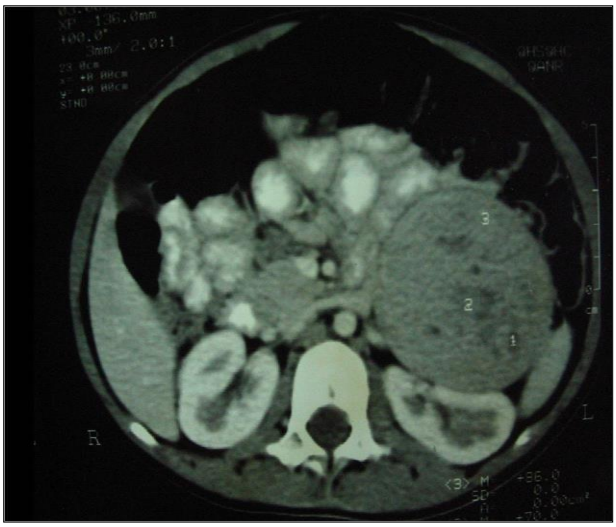

Figure 2: CT abdomen showing mass arising from left adrenal gland.

The patient underwent left adrenalectomy . At surgery, liver, both kidneys, small bowel, uterus and both ovaries were normal. There was no free fluid in the abdomen or retroperitoneal lymphadenopathy. The tumor of size 8 x 12 cm was arising from the left Adrenal gland. The tumor was well encapsulated with no infiltration into surrounding structures. There was no tumor spillage intraoperatively. The postoperative recovery was uneventful and the child kept well during follow up. The child received no postoperative adjuvant therapy. The histopathologic report showed a soft tissue mass measuring 7.5 x 6.4cm with features of ACC (Figure 3).

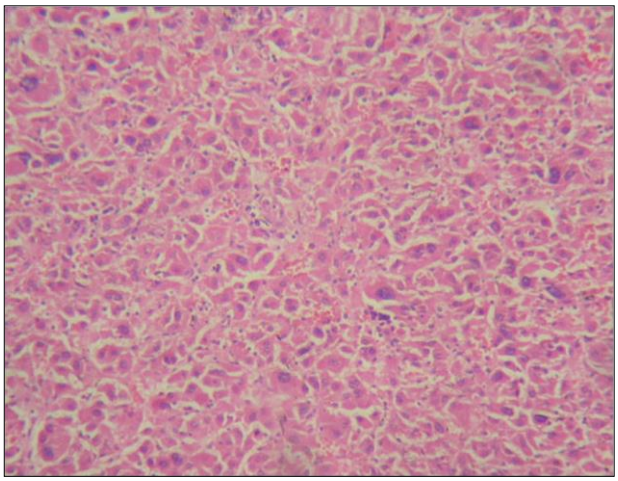

Figure 3: Histological picture of classical adrenocortical carcinoma.

DISCUSSION

ACC is a rare tumor (0.2% of all childhood malignancies) with incidence of 1-2 case /million/year population.[1] It is more common before 5 years of age and female to male ratio 2 :1. It is hormonally functional in 80-100% patients and the predisposing lesions include congenital adrenal hyperplasia and adenoma.[2] It may also arise de novo and arise from a single progenitor cell.[3]

Clinical presentation in children is usually associated with steroid overproduction; most commonly Virilization (66%) and the rest usually will have symptoms of Cushing's syndrome. Virilization is secondary to secretion of adrenal androgens. Feminization can occur in 2-25%, due to excess of oestrogens.[3,4] Nonfunctional tumors are rare in children (5%).[4,5]

Adrenalectomy offers the best chance for cure.[4,5] If extensive disease is found at operation, wide en bloc resection of tumor and involved organs with lymphadenectomy is recommended.[5] Surgery may be feasible for recurrent disease also.[6,7]

CONCLUSION

Though ACC is a rare tumor in children, many of these tumors are hormonally functional , with resultant changes of virilisation, features of Cushing's syndrome or feminization, according to the hormonal milieu. ACC has significant syndromic and genetic association. Surgery offers the best chance of cure, especially in localized disease.

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