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Infantile Virilization Secondary to Malignant Etiology

Nagesh Dasarwar*, Ramakrishnan Santosh and Sravya Datla

Consultant Pediatrician and Neonatologist, GMR Varalaxmi Care Hospital, Andhra Pradesh, India

Abstract

Adrenocortical tumor is a rare malignancy (1-2/million) in children with a heterogenous presentation and generally poor prognosis. We report two cases of adrenocortical carcinoma who presented primarily with virilization along with cushingoid features and hypertension. Both the Children were managed with surgical resection of tumor, steroid replacement and antihypertensives.

Keywords:Virilization; Clitoromegaly; Cushing syndrome; Adrenocortical carcinoma

Introduction

Adrenocortical carcinoma is a rare but highly aggressive malignancy with an estimated annual incidence of 1.5-2 per million population [1]. Females are more commonly affected. There is bimodal age distribution with peaks occurring before age 5 years and a second peak in the fourth to fifth decade [2]. The prognosis is poor with a significant prpoportion (21%-39%) of patients having distant metastasis at the time of presentation [2] and a 5 year and overall survival ranges between 38%-60% [1]. Even after curative resection, the majority of patients develop early tumour recurrence or distant metastasis [1,2]. Adrenocortical tumors may occur sporadically or as a component of certain hereditary syndromes such as the Li-fraumeni syndrome, Beckwith weidemann syndrome, Multiple endocrine neoplasia-1, Careny complex,Congenital adrenal hyperplasia [3,4].

Case Report

Case 1

An eight month old girl presented with excessive weight gain, predominantly in the truncal region and face, hirsutism, pubic hair growth. On examination cushingoid phenotype was observed with hirsutism, facial and pubic hair, clitoromegaly and hypertension, BP 142/90 mmhg (Figures 1a-1c). Biochemical evaluation showed normal plasma ACTH (Adrenocorticotropic hormone), serum cortisol 42.1 mcg/dl, testosterone 6.91 g/dl, DHEA-S(Dehydroepiandrosterone sulfate) 28.8 mmol/l and androsterdione >12.0 ng/ml. Complete blood profile showed Hb (Hemoglobin) 11.3 gm%, TC (Total cell count) 11400 cells/cumm, platelets 6.2 lakhs, ESR (Erythrocyte sedimentation rate) 62 mm at the end of one hour, serum electrolytes, calcium, renal and liver functioning tests were with in normal limits. CT (computed tomography) abdomen with Intravenous contrast showed 5.1 cm*5.2 cm mass lesion of soft tissue density attenuation in right suprarenal region showing mild contrast enhancement with no evidence of calcification or liver involvement, suggestive of adrenocortical carcinoma (Figures 2 and 3). Right adrenal tumor resection was performed and histopathological examination was reported as adrenocortical carcinoma with no extracapsular extension. Post-surgery outcome was favourable with decreased cushingoid appearance and decreased clinical virilization (Figure 2). Blood pressure returned to normal. Testosterone, androstendione, DHEA-S levels returned to normal over a period of next 2-3 months, oral hydrocortisone and enalapril were tapered and stopped.

Case 2

A seven months old female child presented with rapid weight gain and hirsutism. On detailed physical examination she was having excessive pubic hair, clitoromegaly. Hypertension [BP-148/88] and cushingoid features. Abdominal palpation showed lump in left side of abdomen. Biochemical evaluation showed normal plasma ACTH, serum cortisol 84.0 mcg/dl, testosterone 9.4 g/dl, DHEA-S 42.8 mmol/l and androsterdione >17.0 ng/ml. Complete blood profile showed Hb 9.3 gm%, TC 9200 cells/cumm, platelets 4.2 lakhs, ESR 40 mm at the end of one hour, serum electrolytes calcium, renal and liver functioning tests were within normal limits. MRI (Magnetic resonance imaging) abdomen showed heterogenous 7.7*6.4 cm, mass in left suprarenal region suggestive of adrenal adenocarcinoma (Figure 4). Left adrenal tumor resection was performed and histopathological examination was reported as adrenocortical carcinoma with no extracapsular extension. Post-operative CT abdomen was reported to be normal with no residual

Figure 1a/1b/1c: Cushingoid habitus, buffalo hump, truncal obesity, moon facies, pubic hair growth.

***Corresponding author:** Nagesh Dasarwar, Consultant Pediatrician and Neonatologist, GMR Varalaxmi Care Hospital, Andhra Pradesh, India, Tel: 80042 51834; E-mail: dsrwr@yahoo.co.in

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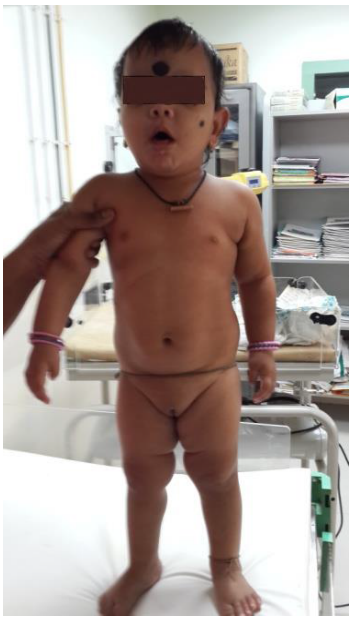

Figure 2: Clinical picture of child 3 months post resection of ACC (showing complete recovery).

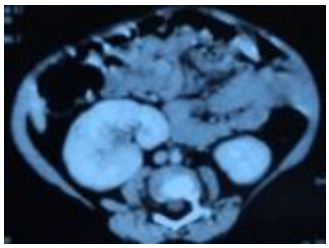

Figure 3: CT Abdomen showing right adrenal cell carcinoma.

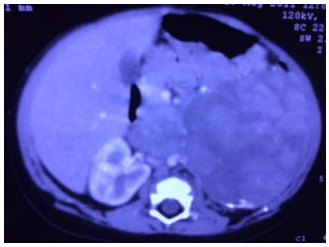

Figure 4: MRI abdomen showing left adrenal cell carcinoma.

lesion. Post-surgery outcome was favourable with total disappearance of cushingoid features, hypertension and clinical virilization.

Discussion

Adrenocortical carcinoma presenting as cushingoid features in early infancy is extremely rare [1.5-2/million] [1]. Most studies show adrenocortical carcinoma having female preponderance [2]. Adrenal carcinoma presents with evidence of adrenal steroid hormone excess in approximately 60% of cases. Most tumors in children are functional and virilization is by far the most common presenting symptom followed by cushing syndrome and precocious puberty. Rapidly progressing cushing syndrome with or without virilization is the most frequent presentation. Androgen secreting adrenocortical carcinoma in females presents with deepening of voice, male pattern baldness and oligomenorrhea. Although benign adrenocortical tumors tend to secrete a single class of steroid, ACC can secrete various types; cosecretion of cortisol with androgens is a frequent combination and is highly suggestive of malignancy [5,6]. Using a logistic regression model, Hussain et al. [7], found tumor size of greater than 4 cm and heterogenous enhancement to be the most important discriminators of malignancy. Regarding prognosis, the only clinical parameters that are invariably associated with a poor prognosis are tumor size and resectability [5,8]. The average survival rate of children with adrenocortical carcinoma is estimated around 50% which increases to 70% when resection is achieved full [9]. The hormonal normalization occurs in the week following surgery and clinical virilization take weeks to months to disappear. Adrenocortical cell carcinoma generally carries poor prognosis and is unlike other tumors of the adrenal cortex which are benign (adenomas) and only occasionally cause cushings syndrome [10-12].

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