

# Pediatric Adrenocortical Carcinoma: Case Report

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**Abstract:** Although adrenocortical tumors are found with some frequency, particularly in older patients, and more often in females, care must be taken to obtain an accurate diagnosis and course of treatment. Adrenocortical adenomas (ACA) and adrenocortical carcinoma (ACC) have very distinctly different potential outlooks for patients, and unless treatment is aligned with the correctly identified etiology, outcomes may be severe. The incidence of ACC in pediatric populations is much rarer, and the disease manifests itself with an array of symptoms that can be assessed and identified with treatment options. Unless the correct diagnosis and treatment are determined at an early stage of the disease, the prognosis for the young patient's status can be very unfortunate. This document will discuss a recent case involving pediatric ACC.

**Keywords:** pediatric adrenocortical carcinoma, virilizing tumors, pediatric oncology.

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## I. INTRODUCTION

Adrenocortical carcinoma (ACC) has different features in adult or senior populations and that of children. In pediatric cases, "ACC has a bimodal peak of incidence, the first in children less than 5 years old, and usually presents as virilization syndrome (84%) and less frequently as isolated Cushing syndrome (6%)".<sup>1</sup> There are different types of adrenocortical tumors, and it is important to distinguish between those that are actually symptomatic of ACC, rather than adrenocortical adenomas, as the former, if undiagnosed or incorrectly diagnosed, can be fatal.

While the Weiss system is most often utilized to determine the presence of ACC in adults, it is found not to be as helpful in pediatric cases. Assessment of this indicates that for children, "miR-483-3p was also upregulated in benign and malignant pediatric tumors, which is consistent with the finding that IGF2 is overexpressed in adrenocortical tumors independently of their malignancy"<sup>2</sup>, making it more difficult to distinguish between carcinoma and adenomas. ACC in children is extremely rare, and occurs at the rate of "0.3/million/year with a bimodal peak under the age of 5 and after 10 years and they also affect girls more frequently than boys."<sup>3</sup> Among the possible contributory factors is suspected malfunction of the adrenal glands, which have significant physical effects on the child's body, as well as often resulting in symptoms such as pain and abdominal discomfort. Research has indicated that congenital defects are linked to malignancies in pediatric cases in general.<sup>4</sup>

## II. CASE STUDY

A three years and seven months old boy presented to a pediatrician with progressive weight gain and increased appetite for 3 months associated with facial acne. The pediatrician prescribes topical treatment for acne which did not help. Later he developed pubic hair, enlarged penis and started to get adult sweat odor, so his pediatrician referred him to a tertiary hospital.

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<sup>1</sup> João Evangelista Bezerra and Ana Claudia Latronico, MicroRNA Era: The Importance for Diagnosis and Prognosis of Adrenocortical Tumors. BioMed Research International, 2014.

<sup>2</sup> Ibid.

<sup>3</sup> Enzo Lalli and Bonald C. Figueiredo, Pediatric adrenocortical tumors: what they can tell us on adrenal development and comparison with adult adrenal tumors. Frontiers in Endocrinology, 2015.

<sup>4</sup> Ibid.

On examination his height was 102 cm (90<sup>th</sup> centile), weight was 18.4 kg (97<sup>th</sup> centile), and blood pressure was 120 / 90 mmHg (normal < 95 / 48 mmHg). He had Cushinoid facies, signs of virilization in the form of facial acne (mainly frontal), hirsutism, presence of axillary hairs, puberache (Tanner stage 4), and an enlarged penis (stretched penile length 6.2 cm). There was a firm mass occupying the left hypochondrial, epigastric and right hypochondrial regions. There were no neurocutaneous markers. There was no family history of malignancy. Laboratory investigations revealed normal hematological and biochemical parameters. Morning (0800 hours) serum cortisol was 1102 nmol/L (repeated 1074 nmol/L). Other hormonal levels were as follows: serum adrenocorticotropic hormone (ACTH) 0.4 pmol/L, dihydro-epiandrosterone sulfate 54.28  $\mu\text{mol} / \text{L}$ , and testosterone 10.76 nmol / L (High for age). Computed tomography (CT) of the abdomen showed a heterochoic left suprarenal mass measuring 10 x 9 x 11 cm in anteroposterior, transverse and craniocaudal dimensions respectively, with streaks of calcifications noticed in the center. The left kidney is displaced inferiorly, and the lesion appear to be separable from the lower pole of the left kidney. There is no evidence of vascular encasement, nor intraspinal involvement. Liver, spleen, pancreas, right kidney, and right adrenal gland are unremarkable. Fine needle aspiration cytology (FNAC) from the mass was suggestive of an adrenocortical tumor.

Thereafter the patient was surgically treated by upfront gross total resection of the tumor, however as it was inseparable from left kidney, he underwent left sided nephrectomy as well. As there was tumor spillage intraoperatively, the final stage was Stage 3. The patient tolerated the surgery well, and there were no post-operative complications. Following surgical excision of the tumor, a repeated dihydro-epiandrosterone sulfate level was 0.62  $\mu\text{mol} / \text{L}$  and, a computed tomography (CT) of the abdomen showed a well-defined enhancing soft tissue nodule abutting the left crus of the diaphragm which measures 8 x 12 x 15 mm (TS x AP x CC). The patient was commenced on a chemotherapy regimen comprising Mitotane, Cisplatin, Doxorubicin and Etoposide.

After completing four cycles, the interim assessment showed a rising dihydro-epiandrosterone sulfate level (1.08  $\mu\text{mol} / \text{L}$ ) and increasing in size of the above mentioned nodule (now measuring 2.3 x 3.9 x 3 cm (TS x AP x CC)). So, the patient was assessed to be not chemo-sensitive, and there were indications of renal toxicity. After discussion with parents and a recommendation that surgery provided the best potential removal of the carcinoma, the surgical team received approval from parents, for a second surgery, with a follow-up plan of oral Mitotane and local control radiotherapy. The patient tolerated this plan well and the clinical features of precocious puberty, namely pubic hair and acne, subsequently resolved. Due to challenges associated with drug availability, initially Mitotane was only available to our patient intermittently and eventually it become available and continued for six months.

Eight months after second surgery, follow-up computed tomography scan of the chest and abdomen confirmed relapsed disease with multiple pulmonary masses, as well as pelvic and mesenteric nodal masses. The liver was infiltrated with multiple enhancing lesions. The dihydro-epiandrosterone sulfate level was 18.4  $\mu\text{mol} / \text{L}$ .

Palliative care was commenced in view of the disease relapse with extensive multisystem involvement. The patient is currently on supportive care only.

### III. DISCUSSION

As with any confirmed or suspected cancer, diagnosis and a treatment plan should be effected as soon and as early in the progression of the disease as possible. For pediatric ACC, this is particularly critical, as the symptoms and outcomes are generally very different from those of adults. The child's age, the onset of symptoms and the status of the ACC will determine much of the course of treatment and survival. ACC should be considered among patients with a history of rapid weight gain and growth failure, even in the absence of virilization. For very young children, "completely resected tumors weighing less than 200 g and without metastasis, the overall 5-year event free survival rate for infants was 91%"<sup>5</sup>. This information holds some hope that identifiers and classifiers for pediatric ACC may be developed to reduce the time period for obtaining the very specific results indicating the nature of the tumor/s.

### IV. CONCLUSION

Adrenocortical carcinoma is considered to be extremely rare in children, and because it has specialized features that may elude standard testing, there is a need for both development of new diagnostic techniques and greater awareness among oncologists and other physicians about this disease. Parents who recognize significant medical and other issues with their

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<sup>5</sup> Sonali Malhotra, Apoorva R. Waikarb, Prabhsimranjot Singhc, Ludovico Guarinid, Elka Jacobson-Dickmana, Roja Motaghedia, and Irina Kazachkova Rare Presentation of Adrenocortical Carcinoma in a 4-Month-Old Boy. World Journal of Oncology, 2017.

children would be most likely to take the child to a family physician or primary care physician, who may or may not be versed in the specific manifestations of pediatric tumors in general, or ACC. In addition to development and dissemination of literature and training information for physicians and other medical professionals, it would benefit the field of oncology to implement ongoing courses with case study practicums to assure that there is better awareness of the symptoms of ACC, as well as the need for prompt medical intervention.

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