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Endocrine

A Rare Case of Adrenocortical Carcinoma in an 8-Year-Old South Asian Male Presented with Hypertensive Encephalopathy and Precocious Puberty

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Abstract **Case Report**

Adrenocortical carcinoma (ACC) represents an inordinately rare malignancy, particularly among children, where its incidence is even more infrequent. The narrow number of firm instances presents challenges in pinpointing prognostic, biological, and therapeutic markers to adapt innovative targeted strategies. Despite the distinction in clinical presentations and pathology, the treatment protocol for childhood ACC typifies that established for adults, occasionally leading to more contentious outcomes. In regions with limited research frameworks and financial restraints interrupting access to modern diagnostic modalities, cases of rare malignancies like pediatric ACC serve as climactic academic and scientific focal territory for further investigation. Presented here is a case of an 8-year-old male from Bangladesh diagnosed with ACC. Surgical intervention was undertaken following imaging studies, and confirmation was received through histopathological examination. However, despite adherence to the protocol-based treatment regimen, the patient succumbed shortly thereafter.

Keywords: Adrenocortical Carcinoma, Precocious Puberty, Hypertensive Encephalopathy.

INTRODUCTION

Adrenocortical carcinoma (ACC) is an exceptionally uncommon form of malignancy observed in both adults and children, typically associated with a poor overall prognosis [1]. Geographical distribution of ACC displays significant variability, with elevated incidence rates noted in South and Southeast regions of Brazil [2]. According to data from the National Cancer Institute, carcinoma accounts for only 1.3% of all pediatric malignancies [2], among which ACC constitutes a mere 0.2% [2], translating to an incidence rate of 0.2-0.3 patients per million with a female predominance [1, 3–5]. Due to the scarcity of cases, comprehensive understanding of pediatric ACC including its demographics, clinical presentation, biological characteristics, prognosis, treatment strategies, and overall survival remains limited [2]. Although the establishment of registries has contributed to enhancing our knowledge [6, 7], the majority of available data predominantly stem from Brazilian patient cohorts [2]. Notably, pediatric ACC exhibits distinct clinical features, molecular profiles, histopathological characteristics, and prognostic markers compared to its adult counterpart [8–11]. However, treatment options for pediatric ACC, such as surgery, radiotherapy, or chemotherapy employing agents like Etoposide, doxorubicin, Cisplatin, and Mitotane, largely rely on protocols developed for adult ACC cases [1, 6, 12]. Research on alternative treatment modalities for pediatric ACC is scant, and consensus regarding protocols for managing advanced disease is lacking [13]. To address these knowledge gaps, there is a pressing need for international prospective studies drawing upon published clinical evidence, including case reports and case series, to effectively demonstrate the importance of discerning between pediatric and adult adrenocortical carcinoma, and to establish standardized clinical classifications and risk-based therapeutic strategies for pediatric ACC. Here, we present a rare case of an 8-year-old male from Bangladesh, the Southeast Asian zone diagnosed with Adrenocortical carcinoma who expired shortly after receiving the first cycle of post-operative chemotherapy.

CASE PRESENTATION

An 8-year-old Bangladeshi male child presented with complaints of dull aching, diffuse headache associated with blurred vision for 7 days, and noticeable pubertal growth over the last year with prominent appearance of mustache, axillary, pubic hair

and visible enlargement of phallus size, for last six months. In the ER, the patient developed multiple episodes of generalized tonic-clonic convulsions associated with unconsciousness, tongue biting, bowel and bladder incontinence. Each attack persisted for around 5 minutes despite administering IV Diazepam. The interval period between two convulsions was less than 30 minutes.

Vital signs demonstrated elevated blood pressure (~ 150/110 mmHg), recorded on several occasions in non-convulsive state with a standard pediatric blood pressure cuff. There was no R-R or R-F delay, and with a normal pulse rate. On examination, the patient was sedated, with a Glasgow Coma Scale (GCS) score of 9/15 (E2 V3 M4). Skin survey revealed acne on the face, the appearance of facial/axillary/pubic hair. Anthropometrically, his height and weight were above the 97th percentile. The patient had attained puberty, as evidenced by Tanner staging- the stretched penile length (SPL) measured 7 cm, presence of coarse pubic hair (Tanner stage 3), and bilateral testicular volume (TV) was observed 6 ml (Tanner stage 2). Notably, the testes were non-tender, with scrotal rugosity present. The rest of the physical examination was unremarkable.

Routine lab work and hormonal assays showed

Clinical Parameters	Results Obtained	Normal Range
Erythrocyte sedimentation rate (ESR)	60 mm in 1st hour	Male — 0-15 mm/h Female — 0-20 mm/h
Serum Potassium	2.90 mmol/L	3.5-5 mmol/L
Serum Cortisol	27.10 µg/dL	4.5 – 24.0 µg/dL
Serum Aldosterone	433.30 pg/ml	20-180 pg/ml
Dehydroepiandrosterone sulfate (DHEA)	200.6 µg/dL	1.7-61.8 µg/dL
Serum Testosterone	21.1 nmol/L	<0.45 nmol/L
Serum Luteinizing hormone (LH)	0.6723 mIU/mL	2.8-6.8 mIU/mL
Serum Follicle Stimulating Hormone (FH)	<0.2250 mIU/mL	1.3-11.8 mIU/mL

CT and MRI Brain were negative. US of KUB demonstrated a well-defined mixed echogenic structure, measuring approximately 7.5 x 6.6 cm, detected in the right adrenal region, suggestive of a right adrenal mass. CT Abdomen with and without contrast revealed a heterogeneously enhancing large well defined hypodense lesion with internal coarse calcification noted in the right adrenal region measuring about (8.1 x 7 x 5.7) cm causing indentation over upper pole of the right kidney with a Hounsfield Unit (HU) >20. CT Chest with and without contrast was negative for metastasis.

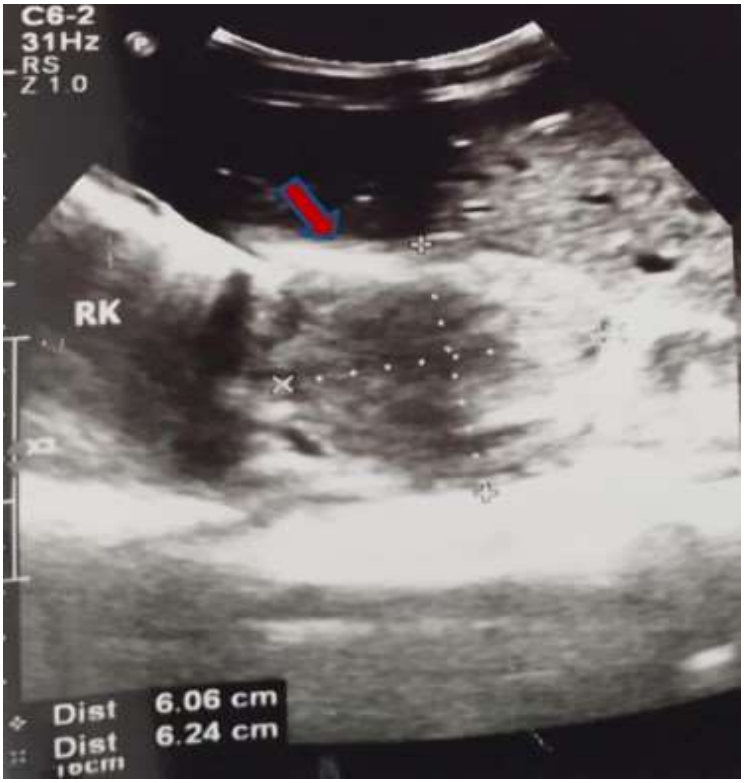
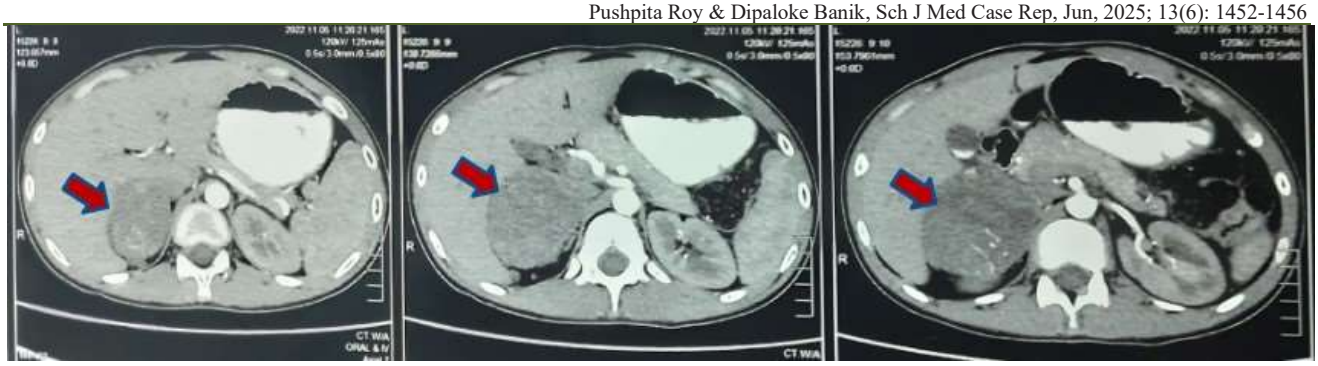


Fig. 1: US of KUB demonstrates a well-defined mixed echogenic structure measuring approximately 7.5 x 6.6 cm, detected in the right adrenal region, suggestive of a right adrenal mass



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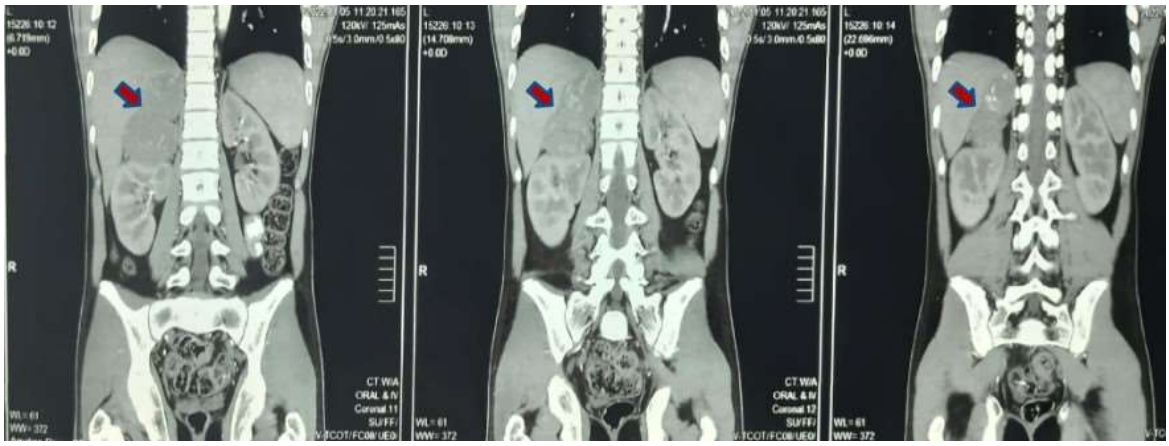

B

Fig. 2: Abdominal CT Scan (a) axial section and (b) coronal section reveal Heterogeneously enhancing large well defined hypodense lesion with internal coarse calcification noted in the right adrenal region causing indentation over the upper pole of the right kidney

The patient was initially managed conservatively with a combination of general and supportive treatments, including counselling of parents. Nutritional support was provided through NG tube feeding alongside pharmacological interventions for elevated BP and recurrent seizure episodes. Endocrinology, Oncology, and Urology were consulted. Ultimately, the patient underwent surgical resection of the tumor as part of the specific treatment plan.

The patient underwent a right adrenalectomy under general anesthesia, revealing intraoperative findings of an enlarged adrenal mass located above the right kidney, which had invaded the lower part of the liver and the inferior vena cava. An engorged vein was observed over the mass. Following excision, the specimen was sent for histopathological analysis. Histopathology revealed a tumor with solid, diffuse, trabecular, or glandular growth patterns comprising pleomorphic polygonal cells with abundant granular and deeply eosinophilic cytoplasm. The tumor exhibited large nuclei, prominent nucleoli, occasional binucleated giant cells, and a prominent trabecular growth pattern of relatively small-sized cells. Necrosis and frequent mitotic figures were also noted. Histologically, it was identified as an adrenal cortical carcinoma, Oncocytic type, categorized as high grade, with a tumor size of (13 x 7 x 6) cm. The tumor invaded into or through the internal capsule, exhibited hemorrhagic and necrotic areas, with the margin involvement.

The patient started on adjuvant chemotherapy following surgical resection of the tumor w/ Mitotone, Cisplatin, Doxorubicin, and Etopside w/ the plan to give 6 chemo cycles. However, patient could not tolerate the chemo, developed adrenal crisis (repeated vomiting, diarrhea and disorientation) after 1st cycle, and eventually expired despite all measures.

DISCUSSION

Our patient underwent the standard treatment regimen currently established for adult patients. However, the surgical intervention did not yield a favorable prognosis, ultimately resulting in the patient's death shortly after receiving post-operative chemotherapy. There is a crucial need for more available case reports focusing on childhood adrenocortical carcinoma. Such reports are imperative for doing further research, tailoring the development of targeted therapies, establishing prognostic markers, and facilitating post-surgical efficacy optimization.

Consent: We extend our sincere gratitude to the patient's parents for consenting verbally to the publication of the case for scholarly purposes.

Declaration of Interest: There is no conflict of interest to declare. We appreciate the contributions of all individuals involved in the entire process, from case diagnosis to treatment. PR, DB, gathered and structured the information, drafted the manuscript, and conducted the literature review.

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