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CLINICAL CASE

Adrenal carcinoma: Clinical case report.

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Abstract

Introduction: Adrenal carcinoma is a malignant neoplasm representing 0.2% of malignant tumors. Its etiology is unknown, although the existence of a predisposing effect due to high concentrations of ACTH is suspected.

Clinical case: This is a 5-year-old female patient with seizures and high blood pressure symptoms. On physical examination of the abdomen, a mass was palpated at the level of the left hemiabdomen that exceeded the midline, non-mobile, non-painful, with irregular edges, virilization of the external genitalia.

Diagnostic workshop: Laboratory tests determined hypercortisolism and hyperandrogenism. Abdominal tomography reports left adrenal region: space-occupying mass with defined borders, liver of homogeneous shape and size, in segment II: defined hypodense lesion of 2 cm.

Evolution: Once the arterial hypertension was controlled, left adrenalectomy was performed, plus lymph node dissection, and the diagnosis of adrenal carcinoma was confirmed with the histopathological study. After the patient's poor prognosis, chemotherapy was started, decomposing and presenting multisystem failure. Patient dies.

Conclusions: Adrenal carcinoma has a poor prognosis in advanced cases, as in the present case.

Keywords:

MESH: Adrenocortical carcinoma, Pediatric adrenocortical tumors, Adrenalectomy, Child, Pediatrics.

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Introduction

Adrenal carcinoma (AC) accounts for 0.2% of childhood and adolescent malignancies, with an annual worldwide incidence of 0.2 to 0.3 cases per million subjects [1]. Due to the clinical presentation, the diagnosis is made by performing complementary hormonal profile tests, abdominal tomography, or magnetic resonance imaging. The treatment is surgical resection and chemotherapy, and its stratification gives us the prognosis.

Clinical case

A 5-year-old female patient with no relevant history suffered a fall from a bicycle, receiving a blow to the abdomen and skull; this event's context and companions are unknown. After four days, the patient was taken to a second-level public hospital due to disorientation, rambling speech, ataxic gait, and progressive deterioration of the state of consciousness during her stay in the hospital; the Glasgow Coma Scale was 9/15. The picture worsened with tonic-clonic seizures, reaching status epilepticus, arterial hypertension, and a new Glasgow scale assessment of 6/15, so it was decided to intubate the patient with assisted mechanical ventilation, and the referral was decided. To the Baca Ortiz Pediatric Hospital in Quito-Ecuador.

Upon admission, the patient was hemodynamically unstable, so she was hospitalized in the pediatric intensive care unit. His blood pressure was 145/80 mm/Hg, his heart rate was 100 beats per minute, and his weight was 45 kilograms. On physical examination: normocephalic head, hyperpigmented face, hirsutism, and facial hair. Pinpoint pupil eyes. Soft, depressible abdomen, a mass is palpated at the level of the left hemiabdomen that exceeds the midline, nonmobile, nonpainful, with irregular edges. Tanner IV female genitalia in the urinogenital region with clitoral hypertrophy.

Diagnostic workshop

In the laboratory tests, leukocytes were observed: 6,400 u/uL; Hemoglobin 19.7 g/dl; hematocrit 59%; Platelets 244,000 u/ul; neutrophils 72%; Creatinine 0.57 mg/dL; Urea: 72.3 mg/dL; Lactate dehydrogenase 2,790 U/L; Testosterone 5,005 ng/dL (Reference value 15 to 70 ng/dL); Follicle - stimulating hormone < 10 mIU/ml; Cortisol AM 41.37 mcg/dl (Normal value 6.02-18.4 mcg/dl); Metanephrines 4.2 mcg/dl (reference value <2.2 mcg/dl); Adrenaline 186 pg/ml (reference value <100 pg/ml); Norepinephrine 986 pg/ml (Reference value < 750 pg/ml).

No space-occupying lesions were found in the skull tomography. In the abdominal tomography, it was reported that in the left adrenal region, there was a space-occupying mass with defined edges, lobulated with a heterogeneous appearance due to the presence of calcifications, measuring 13 x 10 x 12 cm, with an adequate cleavage plane with the left kidney displacing it toward the caudal area. In segment II, the liver had a homogeneous shape and size: a defined hypodense lesion of 2 cm without postcontrast enhancement was described. The kidneys were usual in size, shape, and location, with a preserved corticomedullary relationship and without pelvicalyceal ectasia (Figure 1).

In chest tomography, vascular structures of the mediastinum and cardiac silhouette were reported without alteration, with no enlarged lymph nodes or mediastinal masses. At the level of the lung parenchyma, bilateral basal consolidations were described with slight pleural effusion and multiple bilateral diffuse nodular lesions, the largest of which measured 12 millimeters.

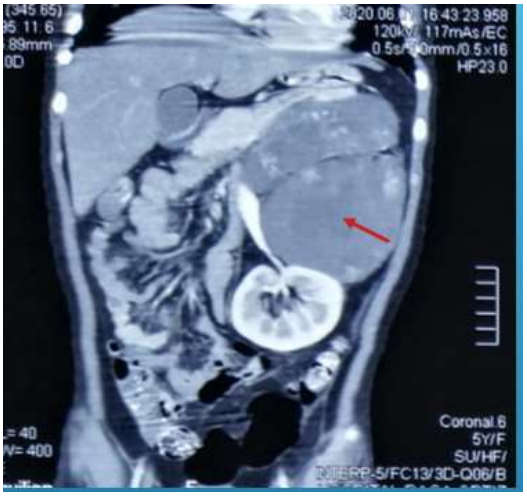

Figure 1. Abdominal tomography: adrenal mass.

Evolution

The patient remained in the intensive care unit for 12 days with a diagnosis of hypertensive urgency; it was managed with nitroprusside, hydralazine, and carvedilol. Once the hypertension was controlled, the patient was extubated and transferred to hospitalization at the General Clinic service. Multidisciplinary oncology, pediatrics, cardiology, endocrinology, nephrology, and urology management are performed to manage and control blood pressure prior to surgery.

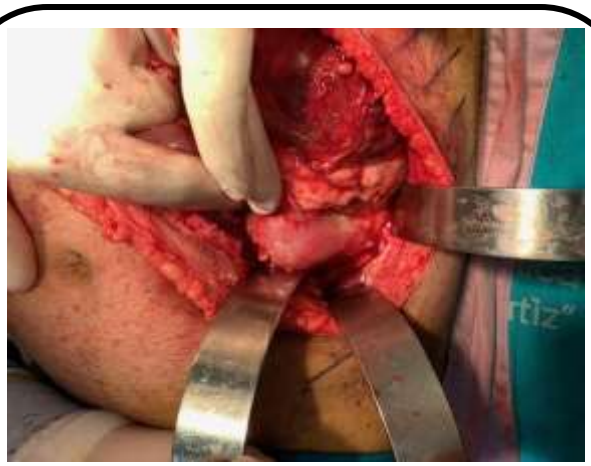

Figure 2. Transsurgical approach to adrenalectomy.

Figure 3. Surgical piece of left adrenalectomy.

At 29 days of hospitalization with hydralazine 20 mg IV every 4 hours, carvedilol 10 mg every 12 hours, doxazosin 1 mg orally every day, and amlodipine 5 mg orally every 12 hours are hemodynamically compensated by maintaining blood pressure in the 90-99th percentile. The patient underwent tumor resection with left adrenalectomy and lymph node dissection.

Among the intraoperatory findings, a mass dependent on the left adrenal gland displaces the kidney and with a size of 14x10x6.5 cm, with abundant neovascularization described; in the left kidney, there was no evidence of tumor infiltration. Postoperative bleeding was approximately 300 milliliters (Figure 2-3). Postoperatively, he remained in the Pediatric Intensive Care Unit for three days, after which he went to the General Clinic floor. His pressures remained stable.

Pathology report: tumor lesion formed with cells with ample cytoplasm with an oncocytic appearance and a central nucleus with an evident nucleus, histological type oncocytic adrenal cortical carcinoma, lymphovascular invasion present, tumor invades the capsule, compromised resection margins, diagnosis Adrenal gland carcinoma stage IV.

Patients with poor evolution after starting chemotherapy go to the intensive care unit (ICU) despite diagnostic and therapeutic measures with an increase in failures and the development of multiorgan dysfunction. Patient death.

Discussion

Adrenal cortical carcinoma (ACC) is a heterogeneous group of plasmatic neoplasms arising from the adrenal cortex. Adrenal cortex tumors (ACTs) have an incidence of 0.3 cases per million per year in children under 15 years of age. The age of the patient in this report was five years. They represent less than 0.5% of all pediatric solid tumors and usually only affect one of the glands but are occasionally bilateral (2% to 10%) [2].

Table 1. Staging of adrenocortical carcinoma proposed by MacFarlane and Sullivan

staging	Size	Nodes- Invasion	Metastasis
I	Small tumor less than or equal to 5 cm	No nodes – local invasion	No distant metastasis
II	Large tumor greater than 5 cm	No nodes – local invasion	No distant metastasis
III	Any size	nodes – local invasion	No distant metastasis
IV	Any size	nodes – local invasion	Distant metastasis

Adrenocortical carcinomas (ACC) in childhood and adolescence are rare, with aggressive heterogeneous biological behavior and poor prognosis [3]. ACC in the pediatric group is seen more frequently in girls, with a male to female ratio of 1:4.

Most patients exhibit virilization (pubic hair, accelerated growth and maturation, penile or clitoral enlargement, hirsutism, and acne) due to excess androgen secretion alone or in combination with hypercortisolism in more than 80% of patients [5]. The common hormonal expression is related to sex hormones. The sex hormones produced by pediatric ACC are primarily androgens (virilizing effects) with some estrogen-secreting ACCs. Approximately one-third of patients present with hypertension, which is usually caused by excess production of glucocorticoids; our patient presented arterial hypertension in addition to signs of virilization. In the present case, the neurological compromise with status epilepticus and decreased Glasgow was probably explained by hypertensive encephalopathy secondary to the levels of catecholamines produced by the adrenal tumor.

Some pediatric ACCs occur in the context of hereditary syndromes. They were found in Li-Fraumeni syndrome, multiple endocrine neoplasia type 1, neurofibromatosis type 1, and polyposis familial adenomatous. Furthermore, pediatric ACCs were found in Beckwith-Wiedemann syndrome [6].

The diagnosis is based on the biochemical characterization of the hormonal syndrome. Due to its known peak incidence, all patients under five years of age presenting with virilization should be evaluated for TCA. Imaging studies often cannot define the benignity of the lesion, and only the presence of metastases determines malignancy. Ninety percent of childhood ACCs cause clinically apparent hormone excess syndromes, typically hypercortisolism and hyperandrogenism. [7]

Sandrini et al. proposed a disease staging system modified by Michalkiewicz et al. It is related to the best cure rates when surgical resection is complete and with free margins [6]. Our patient was characterized by meeting stage IV (Table 1).

Complete tumor resection is currently the only documented effective treatment method; therefore, we recommend open surgery for all tumors with suspicious radiological findings for malignancy and local invasion evidence. Laparoscopic adrenalectomy (respecting the principle of oncological surgery) [8] has been proposed for tumors <6 cm without evidence of local invasion, and for children with advanced disease, chemotherapy and mitotane have been proposed [9]. In our patient, complete resection of the tumor was performed.

Pathological diagnoses are composed of neuroblastic cells and Schwann stroma, the types and quantity of which are crucial to proper classification. The neuroblastic component of a pNT can show a wide range of differentiation from primitive small round blue cells to mature ganglion cells [10]. Although immunohistochemistry may help differentiate between adrenal neoplasms and other tumors, there is still no consensus on its importance as a prognostic indicator in CA28 [7].

The use of adjuvant mitotane for 24 to 36 months in some stage II and III patients, alone or combined with standard chemotherapy, is instrumental 56-59 in improving overall survival and event-free survival [11], starting with adjuvant chemotherapy.

Conclusions

Hypertensive encephalopathy was the profound clinical debut in a girl patient with advanced-stage ACC. This picture was accompanied by Tanner IV sexual maturation. Despite excisional surgical treatment, the prognosis was poor.

Abbreviations

ACC: adrenocortical carcinomas.

Supplementary information

Supplementary materials are not declared.

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Author contributions

María Fernanda Vicuña Pozo: Conceptualization, Data Retention, Fundraising, Research, Resources, Software, Writing - original draft.
Jenny Elizabeth Arboleda Bustán: Conceptualization, Data Conservation, Supervision, Funding, Research, Resources, Writing: review and editing.
Juan Felipe Alarcón Salvador: Curation of data, research, acquisition of funds, supervision, methodology.
Jorge García Andrade: Conceptualization, Data Conservation, Supervision, Funding, Research, Resources, Writing: review and editing.
All authors read and approved the final version of the manuscript.

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Statements

Ethics committee approval and consent to participate

It was not needed.

Publication consent

Written permission for publication was obtained from the patient's parents.

Conflicts of interest

The authors declare no conflicts of interest.

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