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CASE REPORT

Functional Adrenocortical Carcinoma in Adolescence: A Case Report

Carcinoma Adrenocortical funcional en la adolescencia: A propósito de un caso

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ABSTRACT

Introduction: Pediatric adrenocortical carcinoma is a rare and aggressive endocrine neoplasm, with an incidence of less than 0.5 cases per million children. Most are functional tumors, associated with endocrine manifestations such as Cushing's syndrome or virilization. Radical surgical treatment is the mainstay of therapy and the primary prognostic factor. Case report: We present the case of a 13-year-old male adolescent with headache, lower extremity edema, and severe hypertension. Physical examination revealed Cushingoid facies, central obesity, and pitting edema. Hormonal studies showed hypercortisolism and elevated DHEA-S. Contrast-enhanced computed tomography showed a heterogeneous left adrenal mass measuring 16.7 × 13.7 × 11.6 cm, with necrosis and calcifications, consistent with functional adrenocortical carcinoma. Complete surgical resection was performed via open laparotomy, achieving total excision of a 910 g mass, with areas of extensive necrosis and focal capsular rupture on histopathological analysis. Discussion: Early diagnosis of pediatric adrenocortical carcinoma remains a challenge due to its low frequency and variable clinical presentation. Tumors larger than 6 cm require an open approach to reduce the risk of capsular rupture and tumor dissemination. Factors such as extensive necrosis, large tumor size, and capsular rupture are associated with a worse prognosis and the need for adjuvant treatment. Conclusions: Functional adrenocortical carcinoma should be suspected in pediatric patients with Cushing's syndrome and large adrenal tumors. Complete oncologic surgical resection via laparotomy remains the treatment of choice and the main prognostic determinant.

Keywords: Adrenocortical carcinoma; pediatrics; Cushing's syndrome; adrenal tumor; laparotomy hypercortisolism; cancer surgery; adolescence.

RESUMEN

Introducción: El carcinoma adrenocortical pediátrico es una neoplasia endocrina infrecuente y agresiva, con una incidencia menor a 0.5 casos por millón de niños. La mayoría son tumores funcionales, asociados a manifestaciones endocrinas como síndrome de Cushing o virilización. El tratamiento quirúrgico radical constituye el pilar terapéutico y principal factor pronóstico. Caso clínico: Se presenta el caso de un adolescente masculino de 13 años con cuadro de cefalea, edema de miembros inferiores e hipertensión severa. Al examen físico presentó facies cushingoide, obesidad central y edema con signo de Godet. Los estudios hormonales evidenciaron hipercortisolismo y elevación de DHEA-S. La tomografía contrastada mostró una masa suprarrenal izquierda heterogénea de 16.7 × 13.7 × 11.6 cm, con necrosis y calcificaciones, compatible con carcinoma adrenocortical funcional. Se realizó resección quirúrgica completa mediante laparotomía abierta, logrando exéresis total de una masa de 910 g, con áreas de necrosis extensa y ruptura capsular focal en el análisis anatomopatológico. Discusión: El diagnóstico precoz del carcinoma adrenocortical pediátrico continúa siendo un desafío debido a su baja frecuencia y presentación clínica variable. Los tumores mayores de 6 cm requieren abordaje abierto para disminuir el riesgo de ruptura capsular y diseminación tumoral. Factores como necrosis extensa, gran tamaño tumoral y ruptura capsular se asocian a peor pronóstico y necesidad de tratamiento adyuvante. Conclusiones: El carcinoma adrenocortical funcional debe sospecharse en pacientes pediátricos con síndrome de Cushing e imágenes suprarrenales de gran tamaño. La resección quirúrgica oncológica completa mediante laparotomía continúa siendo el tratamiento de elección y el principal determinante pronóstico.

Palabras clave: Carcinoma adrenocortical; pediatría; síndrome de Cushing; tumor suprarrenal; laparotomía; hipercortisolismo; cirugía oncológica; adolescencia.

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INTRODUCTION

Adrenocortical carcinoma is a rare and highly aggressive neoplasm of the adrenal glands, with an annual global incidence of 1 to 2 cases per million inhabitants. In pediatrics, it is even rarer, affecting 0.2–0.38 cases per million in those under 20 years of age; its clinical presentation is highly variable and can mimic other pathologies. Approximately 60-90% of pediatric cases are functional, commonly manifesting through virilization or Cushing's syndrome; other symptoms include abdominal pain; however, up to 80% of cases may be asymptomatic at the time of diagnosis (1). Due to its aggressive behavior, it represents a diagnostic challenge that requires a multidisciplinary and comprehensive approach, in addition to complementary laboratory and imaging studies and confirmation by pathological anatomy. Open surgery is the treatment of choice. Adrenocortical carcinoma (CAC) is a rare and highly aggressive endocrine neoplasm originating in the adrenal cortex, with an estimated global incidence of 1–2 cases per million inhabitants per year. In the pediatric population, it is an exceptional entity, with an approximate incidence of 0.2–0.38 cases per million in those under 20 years of age, presenting a bimodal distribution with greater frequency in children under 5 years and a second peak during adolescence. Unlike adults, between 60% and 90% of pediatric cases correspond to functional tumors, characterized by hormonal hypersecretion, mainly of cortisol and androgens, which leads to clinical manifestations such as Cushing's syndrome, virilization, arterial hypertension, central obesity, and metabolic alterations. (1)

Various international reports and clinical series agree that the clinical presentation of pediatric CAC is heterogeneous and frequently nonspecific, and may mimic benign endocrine, metabolic, or abdominal pathologies, which delays diagnosis and favors tumor progression. Although abdominal pain and the presence of a palpable mass are frequent findings in advanced tumors, some patients remain asymptomatic until late stages. From a radiological point of view, lesions usually present as large, heterogeneous adrenal masses with areas of necrosis, hemorrhage, and calcifications, characteristics associated with malignant behavior and worse prognosis. The management of adrenocortical carcinoma requires a multidisciplinary approach that integrates clinical evaluation, hormonal profile, advanced imaging studies, and histopathological confirmation. Currently, complete surgical resection via open surgery constitutes the treatment of choice and the main prognostic factor for survival, especially in tumors larger than 6 cm or with suspected local invasion, where the laparoscopic approach increases the risk of capsular rupture and peritoneal dissemination. In this context, we present the case of an adolescent with functional adrenocortical carcinoma, highlighting its clinical manifestations, imaging findings, surgical management, and prognostic considerations, with the aim of contributing to early recognition and adequate therapeutic management of this rare entity in pediatric age.

CASE REPORT

History and Physical Examination: A 13-year-old male patient from Achacachi, La Paz, referred for a 2-month clinical picture of headache and swelling of the lower limbs. On admission, he presented a weight of 78.3 kg, height of 162 cm, and severe hypertension (180/115 mmHg). On physical examination, a puffy face (full face), central obesity, and edema with Godet sign (+++) in the lower extremities stood out, findings compatible with a cushingoid syndrome. Complementary Tests: Laboratory tests showed polycythemia (Hb 180 g/L), hyperglycemia (134 mg/dL), and hypoalbuminemia (3.1 g/dL)4. The hormonal profile revealed elevated levels of cortisol (27.3 ug/dL) and DHEA-S (428 ug/dL). Immunology: AND determination by chemiluminescence method, 72.5 ng/mL; hormones: Co am determination by chemiluminescence method 27.3 ug/dL; ACTH am 21.7 pg/mL; testosterone 0.6 nmol/L; DHEA – S 428 ug/dL. On admission, a renal and urinary tract ultrasound was performed, as well as of the adrenal glands; the former showed no alterations, and in the latter, a solid, lobulated lesion was observed in the left adrenal topography, with locoregional mass effect, predominantly hypoechoic, displacing the left kidney laterally, measuring 12.8 x 13.5 x 9.8 cm with a volume of 893 cc.

To complement that study, an abdominopelvic tomography was performed in the contrast-enhanced phase, observing in the left adrenal topography a mass with lobulated but well-defined borders, with irregular hyperdense images (calcifications) of up to 154 UH attenua-tion and others less than 74 UH, in addition to scattered irregular hypodense images of up to 35 UH. There is enhancement of fine septa within it. It exerts a locoregional mass effect dis-placing the pancreatic tail cranially and the left kidney laterally and caudally.

It displaces the vascular hilum (vein and artery) but does not compromise them. It measures 16.7 x 13.7 x 11.6 cm (volume of 1397 cc). (Fig. 1)

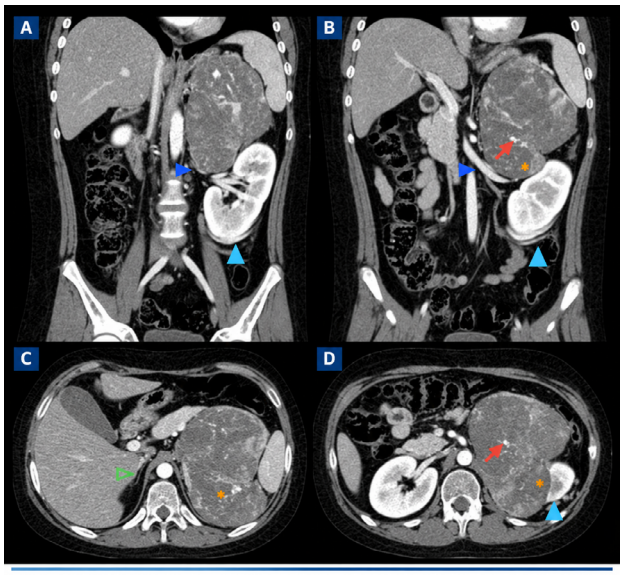
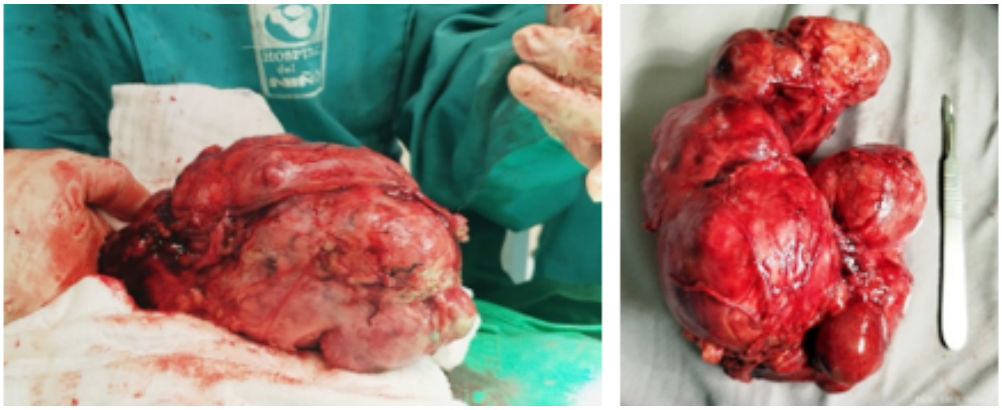


Figure 1. Contrast-enhanced computed tomography (**CT**) images of the abdomen in coronal (A, B) and axial (C, D) planes.

- Enhancing solid component of the splenic mass.
- Area of necrosis within the splenic mass.
- Splenic artery.
- Perisplenic extension.
- Pancreas.

The surgical management of the patient was performed electively through a total surgical resection, considered the main diagnostic and therapeutic step given the high suspicion of malignancy and the size of the mass. A laparotomy was chosen through a wide midline supra- and infraumbilical incision. The choice of open surgery over laparoscopy is essential in tumors larger than 6 cm to minimize the risk of capsular rupture and peritoneal dissemination. A giant, solid, lobulated, and irregular-looking left adrenal mass was identified, with a network of neovascularization and the omentum adhered to its structure. A partial omentectomy and adhesiolysis were performed, followed by meticulous separation of the mass from neighboring structures; the mass was exteriorized and totally resected. During the surgical procedure, significant oozing bleeding from the surgical bed occurred, which was satisfactorily controlled using hemostasis techniques, allowing the procedure to be completed without further complications.

Figure 2. a. Macroscopically, a giant encapsulated, lobulated tumor mass is observed.



Description of the Surgical Specimen. After surgery, macroscopic and histopathological examination revealed critical technical details about the extent of the lesion: dimensions and weight; the excised mass measured 18.1 x 13.5 x 9.5 cm and weighed 910 gr. Although the mass was apparently encapsulated, a sloughed area and capsular rupture were identified in a sector of 5.5 x 2.5 cm3. The surface was heterogeneous, with extensive areas of necrosis (40%), hemorrhage, and dystrophic calcifications.

DISCUSSION

Pediatric CAC has incidence peaks before 5 years of age and after 10 years of age, this case being representative of the second age group 2, 10. Hormonal symptoms, such as those observed in this patient, are key for early diagnosis, since most of these tumors in children are hormone-producing 9.

The fundamental pillar of treatment is surgery with complete resection. The open approach (laparotomy) is strongly recommended over laparoscopy for tumors larger than 6 cm or with suspected invasion, since intraoperative capsular rupture drastically increases the risk of recurrence and decreases survival 7. In this case, the size of 16.7 cm and the partial rupture of the capsule observed in pathology underscore the complexity of management and the need for close follow-up 7,8.

Factors such as tumor size, local invasion, and extensive necrosis (40% in this patient) are indicators of a prognosis that requires evaluation for adjuvant treatment with mitotane or chemotherapy 11. Likewise, in specific regional contexts, evaluation of the TP53 germline mutation is relevant due to its association with more aggressive behavior 11. Epidemiology and Incidence Adrenocortical carcinoma (CAC) is an extremely rare neoplasm with a global annual incidence of 0.5 to 2 cases per million inhabitants1. In the pediatric group (under 20 years), this figure drops drastically to 0.2 – 0.38 cases per million1. It is noteworthy that, although there is a female predominance (ratio 2.5–3.1 to 1) and 75% of cases usually occur before 5 years of age, our patient is in the second reported peak of incidence: adolescence12. Clinical Presentation and Hormonal Diagnosis Unlike adults, where many tumors are nonfunctional, between 80% and 90% of pediatric cases are functional3. Virilization (precocious pubic hair, acne, penile growth) and Cushing's syndrome are the most common initial manifestations3. In this case, the combination of severe hypertension (180/115 mmHg), central obesity, and elevated levels of cortisol (27.3 ug/dL) and DHEA-S (428 ug/dL) confirmed endogenous hypercortisolism that facilitated early diagnostic suspicion45.

Imaging Suspicion Criteria Imaging diagnosis (CT or MRI) is vital. Malignancy should be suspected in masses that present heterogeneity, necrosis, hemorrhage, or calcifications3. Current criteria suggest that CAC should be seriously considered if the patient is younger than 5 years or older than 10 years, and if tumor size exceeds 8.5 cm2. The mass in our patient, 16.7 cm and 1397 cc, far exceeded these parameters, in addition to showing calcifications and areas of necrosis of up to 35 UH on tomography6.

Surgical Management: The Importance of the Open Approach. Surgery with complete resection is the fundamental pillar of treatment7. There is a critical consensus on the technique: laparotomy (open surgery) should be preferred over laparoscopy in tumors larger than 6 cm or with suspected invasion7. This is because capsular rupture during surgery is associated with early recurrences and a drastic decline in survival7. In the case presented, a total resection was performed through a midline incision, prioritizing the integrity of the mass to avoid peritoneal seeding8.

Prognostic factors and pathology. The 5-year survival prognosis in localized stages (I and II) may exceed 85%, provided that surgery is successful and without rupture7. However, the presence of factors such as extensive tumor necrosis (40% in this patient), massive size, lymphatic invasion, and capsular rupture (observed in the area of 5.5 x 2.5 cm in the histological study) are high-risk indicators that make it necessary to consider adjuvant treatment910. In these scenarios, the use of mitotane and chemotherapy regimens (cisplatin, etoposide, doxorubicin) is essential according to international protocols such as ARAR033210.

Genetic Considerations Finally, the genetic component should not be overlooked, particularly the TP53 germline mutation, which confers a more aggressive prognosis10. Due to the diagnostic relevance and familial implications, genetic evaluation is recommended in these patients for early detection in close relatives10. Adrenal carcinomas are rare, with an incidence of 0.5 to 2 cases per million inhabitants per year (2). Childhood adrenocortical carcinoma is a rare malignant tumor, with an estimated annual incidence of 0.2 – 0.38 cases per million children under 20 years of age (3). Although its incidence is low, its aggressive behavior requires an early and multidisciplinary therapeutic approach. The ratio between women and men is 2.5 -3.1. Approximately 75 % of pediatric cases are observed in the first 5 years. (2) Between 80 % and 90 % of pediatric adrenocortical carcinoma cases are functional, that is, they produce hormones. Most children present with virilization symptoms (such as precocious pubic hair growth, enlargement of the clitoris or penis, acne, and accelerated growth). Signs of Cushing's syndrome may also coexist. These hormonal symptoms are, in most cases, the first clinical manifestation, which is key for early diagnosis. (4)(5)

Recommended imaging studies for diagnosis include TC or RM of the abdomen and TC of the chest. TC findings include a well-defined mass with invasive borders, usually unilateral (bilateral in 10% of cases), with areas of necrosis, hemorrhage, calcifications, variable enhancement, venous

invasion, and distant metastases (2). Surgical management of adrenocortical carcinoma in pediatrics represents the fundamental pillar of treatment, with surgery involving complete resection, preferably via an open approach to minimize the risk of tumor rupture and peritoneal dissemination. When performed adequately and without tumor rupture, surgery can guarantee 5-year survival rates higher than 85% in patients with localized disease (stages I and II) (6). From a technical standpoint, the open surgical approach (laparotomy) is recommended over laparoscopy because of the increased risk of capsular rupture and peritoneal dissemination associated with minimally invasive techniques, especially in tumors >6 cm or in the presence of local invasion. Intraoperative tumor rupture is related to early relapses and a significant decrease in overall survival; therefore, its prevention is critical (7).

Another fundamental aspect to consider is the dissection of retroperitoneal lymph nodes, recommended in all cases with suspected lymph node extension. This technique allows more accurate staging and improves overall survival, as observed in ARAR0332, where its inclusion in surgical management was associated with a decreased risk of recurrence (6). Invasion of the adrenal vein, vena cava, adjacent kidney, and retroperitoneum is frequent. The most common sites of metastatic disease are the liver (60%), regional lymph nodes (40%), lungs (40%), pleura, bones, and skin (2). However, the prognosis deteriorates significantly in the presence of adverse factors: age > 4 years, larger tumor size, local invasion, positive margins, elevated Ki-67 (>15 %), Weiss score ≥ 6, or high Wieneke (8). In these cases, adjuvant treatment with mitotane and chemotherapy (usually combinations of cisplatin, etoposide, doxorubicin) is essential, as evidenced in the GPOH-MET 97 and ARAR0332 protocols (9). Genetics, particularly the TP53 germline mutation, is another pillar in diagnosis and management. Its presence, frequent in regions such as southern Brazil, is associated with specific tumor patterns and worse prognosis. Therefore, familial genetic evaluation is suggested, both for therapeutic implications and for early detection in relatives (10). From an imaging point of view, adrenal tumors are the least frequent; however, adrenocortical carcinoma should be suspected in the presence of parameters such as: age younger than 5 years or older than 10 years, size larger than 8.5 cm, lesion heterogeneity with areas of necrosis, hemorrhage, and calcifications, and, among the most severe, metastases or local invasion. As with any tumor in pediatrics, age and its characteristics are extremely important at the time of diagnosis.

CONCLUSIONS

Pediatric adrenocortical carcinoma represents a rare, highly aggressive neoplasm that is difficult to diagnose early, and clinical suspicion should be strengthened in the presence of suggestive endocrinological signs, such as Cushing's syndrome and severe arterial hypertension associated with large adrenal masses. This case demonstrates the importance of integrating clinical, hormonal, and imaging findings to establish a timely diagnosis and guide appropriate therapeutic management. Complete surgical resection through an open approach remains the therapeutic standard for adrenal tumors larger than 6 cm, as it allows better oncological control and reduces the risk of capsular rupture and tumor dissemination. Likewise, this case highlights the need to strengthen the early recognition of pediatric endocrine tumors in settings with limited resources, where diagnostic delay can lead to advanced stages and worse outcomes. The experience presented reaffirms that successful management of pediatric adrenocortical carcinoma depends on a comprehensive and coordinated approach among pediatric surgery, endocrinology, oncology, radiology, and pathology, aimed not only at improving survival but also at optimizing quality of life and long-term follow-up of these patients.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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