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Oncocytic Adrenocortical Carcinoma: Case Report from a Level IV Clinic in Cali-Colombia

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1. Abstract

We present the case of a 37-year-old patient diagnosed with oncocytic adrenocortical carcinoma, a rare variant characterized by its variable clinical behavior and high potential for aggressiveness. The patient presented symptoms compatible with hypercortisolism, and a large, functional left adrenal mass with vascular extension was found. The definitive diagnosis was confirmed by histopathological and immunohistochemical studies. This case is relevant due to the rarity of the oncocytic variant, the advanced local involvement, and the importance of an interdisciplinary approach in surgical and oncological management. It also contributes to highlighting the need for clinical suspicion in the face of nonspecific endocrine symptoms and the value of diagnostic imaging and histological markers for timely detection and treatment.

2. Introduction

Adrenocortical carcinoma (ACC) is a rare and aggressive malignant neoplasm originating in the adrenal cortex. It has an estimated incidence of 0.7 to 2 cases per million per year and a bimodal age distribution, peaking in early childhood and between the fourth and fifth decades of life. It affects a higher proportion of females (55-60%) [1].

Most cases of ACC are sporadic; however, it can occur in the context of hereditary cancer syndromes such as Li-Fraumeni, Lynch, Beckwith-Wiedemann, MEN1, and familial adenomatous polyposis. The pathogenesis involves frequent somatic alterations in genes regulating the p53 pathway, WNT/β-catenin signaling, IGF2 overexpression, and other pathways involved in cell cycle control, chromatin remodeling, and telomere maintenance [2]. From a clinical point of view, approximately 60% of patients present with evidence of excess adrenal steroid hormones, most commonly hypercortisolism (Cushing's syndrome), associated with weight gain, weakness, hypertension, hirsutism, among others. Virilization, hyperaldosteronism, or more specific symptoms depending on sex and secretion of either androgens or estrogens may also occur [1]. Hormonally inactive tumors usually present with symptoms related to the mass effect, such as abdominal or back pain, early satiety, or weight loss [3].

Diagnosis is based on a combination of hormonal evaluation, cross-sectional imaging such as computed tomography, magnetic resonance imaging and 18F-fluorodeoxyglucose positron emission tomography (FDG-PET) and histopathological evaluation [4], including immunohistochemistry and determination of the Ki67 proliferation index as an important pathological prognostic factor [5]. The treatment of adrenocortical carcinoma (ACC) presents a number of distinctive challenges, frequently requiring the integration of oncologic and endocrine factors. The cornerstone of therapeutic intervention for ACC is surgical resection. For individuals classified as stage I or II, as well as for some stage III cases, radical surgical resection remains the only intervention that can offer a curative outcome. Before surgical removal, patients should undergo a comprehensive hormonal evaluation to determine whether the neoplasm produces cortisol

and to assess their susceptibility to postoperative adrenal insufficiency, for which replacement therapy is necessary [4]. Options for advanced disease include mitotane, combination chemotherapy, and, in selected cases, radiotherapy or locoregional treatments [4].

3. Clinical Case

A 37-year-old patient with no significant medical history presented to the emergency department with a 6-month history of weight gain, abdominal distension, fatigue, and spontaneous bruising. Upon admission, the patient presented elevated blood pressure (173/107); physical examination revealed tenderness on palpation in the left hemiabdomen, with no signs of peritoneal irritation, no masses, or megalia. Macular lesions were observed on the torso. Based on the symptoms, an abdominal computed tomography scan was ordered, revealing a 10-cm lesion in the left adrenal gland. Elevated cortisol levels were documented, suggesting autonomous cortisol secretion. An abdominal MRI confirmed a mass in the left adrenal gland measuring 9.3 x 9.0 x 9.8 cm (Figure I), suggestive of a pheochromocytoma. Additionally, thrombosis of the left renal vein extending to a portion of the inferior vena cava was evident. Therefore, an inferior vena cava filter was implanted on 12/30/2024, with the recommendation for postoperative removal and reinitiation of anticoagulation. Alpha blockade with prazosin was subsequently initiated, and hydrocortisone was prescribed during the procedure and postoperatively due to the risk of adrenal insufficiency. A laparoscopic resection of the retroperitoneal tumor and left adrenalectomy were performed on January 3, 2025, without intraoperative complications. The surgical finding was a retroperitoneal tumor measuring approximately 12 x 10 cm located in the territory of the left adrenal gland, vascularized, with displacement of the colon, spleen, and stomach, and with a thrombus in the adrenal and inferior diaphragmatic vein (Figure II).

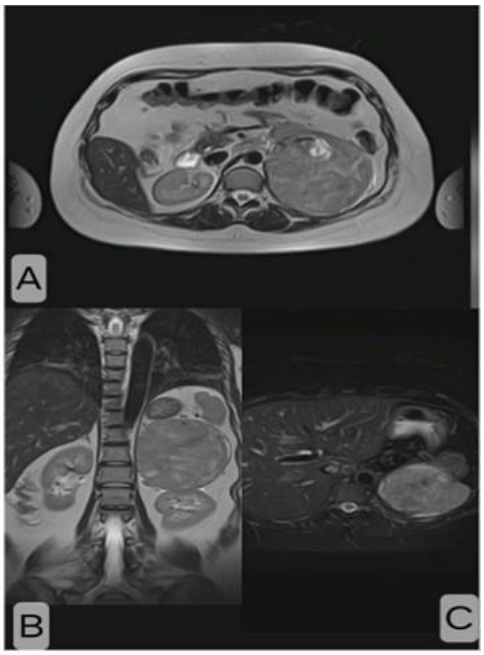

Figure I: Axial section (A-C), coronal section (B), . A heterogeneous mass with cystic lesions within the left adrenal gland is observed.

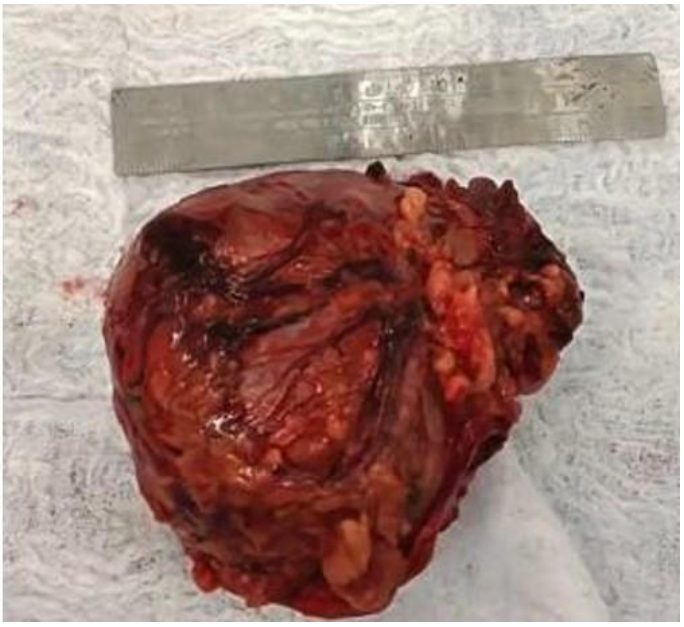

Figure II: Surgical specimen after laparoscopic adrenalectomy of retroperitoneal tumor.

During the hospital stay, postoperative cortisol levels were documented (2.93 µg/dL), confirming adrenal insufficiency, and hydrocortisone therapy was continued. Anticoagulation was restarted, and the vena cava filter was successfully removed on January 8, 2025. The patient was discharged 6 days postoperatively in good clinical condition. The final pathology report showed high-grade oncocytic adrenal cortical carcinoma, T4N-0MX, with tumor stage (ENSAT): III. The immunohistochemistry report was positive for AE1/AE3 receptors, Melana a, inhibin, calretinin, synaptophysin and S100. Ki67 80% (Figure III).

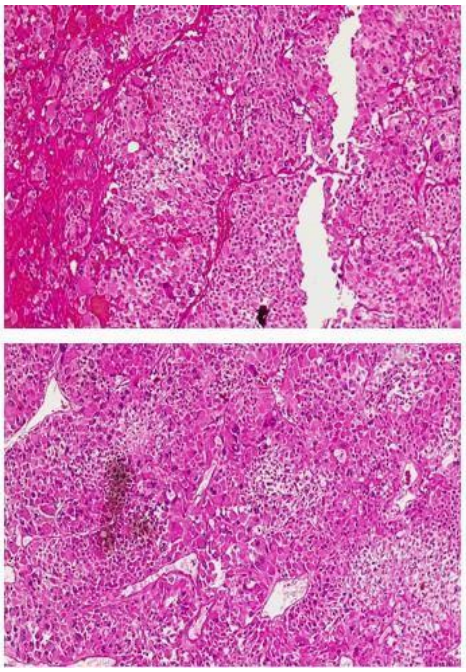

Figure III: Images compatible with an oncocytic adrenocortical tumor, nuclear pleomorphism, with cytological features suggestive of malignant neoplasia.

4. Discussion

Adrenocortical carcinoma is a rare neoplasm, with an estimated incidence of less than 2 cases per million people per year. Its oncocytic variant is rare in reported cases [6]. The oncocytic variant behaves heterogeneously, ranging from mildly aggressive lesions to highly aggressive tumors. Histological criteria for malignancy are used, such as the Lin-Weiss-Bisceglia system, which measures tumor size, local spread, and other morphological characteristics. The Ki-67 proliferative index is also used as a marker of tumor aggressiveness to determine the patient's actual prognosis. In the present case, it is a functional tumor with symptoms consistent with hypercortisolism confirmed by hormonal studies, additionally large in size of 10 cm, with local invasion (ENSAT T4) due to thrombosis in the renal vein and inferior vena cava and a Ki-67 index of 80%, which confers a high risk of recurrence and mortality [7,8]. Access to diagnostic images as a requirement for extension studies focused on initial staging and evaluation of the possibility of tumor resectability is part of the procedures that benefit the patient if done in a timely manner and as soon as possible according to the natural history of the disease, since the 5-year survival rate is less than 15% in patients with metastatic disease [9]. Scientific evidence recommends the diagnosis as the gold standard histopathology, since all adrenal tumors that cannot be easily classified and must be evaluated by a pathologist [10], in this case the patient is performed, together with the staging that results in high grade, which guides the treatment according to the clinical context and improves the probability of survival. Regarding the definitive management of adrenocortical carcinoma, it should undoubtedly be surgical, since it is evident that the recurrence rate is lower and survival increases [11]. In addition, recent studies have demonstrated the safety of laparoscopic resection in large tumors and in the absence of significant local invasion [12]. In this case, laparoscopic adrenalectomy allowed an efficient resection, with minimal invasion and a favorable postoperative period, as well as pharmacological treatment with corticosteroids and control of the adrenal thrombosis presented, with an inferior vena cava filter reflected a strategic planning to minimize thromboembolic risks. On the other hand, scientific evidence demonstrates that adjuvant mitotane therapy is important in patients with complete resection but at high risk of recurrence. Adjuvant mitotane has been shown to improve recurrence-free and overall survival, especially when initiated early. Furthermore, multidisciplinary follow-up with radiological and hormonal monitoring is essential to detect early recurrences or late metastases [13,14]. Current research is directed toward new therapeutic alternatives such as immunotherapy and therapies targeting specific molecular pathways associated with adrenocortical carcinoma, such as PD-1/PD-L1 inhibitors. Although their application has not yet been standardized, recent studies indicate positive results for the treatment of recurrent or metastatic disease resistant to conventional therapies [15,16]. In conclusion, given the high-risk characteristics identified in this case, this type of neoplasia requires a comprehensive oncological approach, close monitoring of diagnostic images, with evaluation of hormonal levels and proliferative markers, with recommendations for genetic evaluation to rule out other germline syndromes, additionally because this is a young patient with a high-histological grade oncocytic adrenocortical carcinoma, these studies are directed due to the high risk of aggressiveness and tumor recurrence.

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