Adrenal cortical carcinoma. A case report

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Case Report

Radiology

OPEN ACCESS

Background: Adrenocortical carcinoma (ACC) is a heterogeneous group of pathologies with different clinical behaviors among patients of different ages. It is the second most common malignant tumor of the endocrine organ after anaplastic thyroid carcinoma. Most tumors are metastatic or locally invasive at the time of diagnosis. Differentiation between ACC and other adrenal masses depends on clinical, biochemical, and imaging factors. The 2022 WHO classification of adrenocortical nodular disease now includes: (a) sporadic adrenocortical nodular disease, (b) bilateral micronodular adrenocortical disease, and (c) bilateral macronodular adrenocortical disease (previously known as primary bilateral macronodular adrenocortical hyperplasia). This article explores the clinical manifestations, diagnostic methods, and procedures for ACC.

Keywords: Adrenocortical carcinoma, adrenal carcinoma.

Adrenocortical carcinoma (ACC) is a rare cancer, but it is the most common primary cancer in the adrenal gland. It is the second most common malignant tumor of the endocrine organ after anaplastic thyroid carcinoma. In large pathology series, ACC accounts for 6.8% of all primary adrenal tumors. According to the most recent population-based database (based on approximately 2,000 cases) from the United States Surveillance, Epidemiology, and End Results (SEER) program published in 2018, covering patients diagnosed with ACC between 1973 and 2012, the annual incidence of the cancer was 1.02 per million.

Case report

A 33-year-old female patient, 24 weeks pregnant, in her fourth pregnancy and with no other significant medical history at the time of consultation, presented to the emergency department due to epigastric pain, accompanied by headache and tachycardia. Her blood pressure was 220/160 mmHg, heart rate was 114 bpm, and fetal heart rate was 140 bpm. She was admitted to the gynecology and obstetrics department of the Hospital Regional de Occidente. Complementary studies were performed with the following results: Hemoglobin (HGB): 14.7 g/dL, Hematocrit (HCT): 43.5%, White Blood Cell count (WBC): 9,950, Neutrophils: 76.2%, Platelets: 376,000, AST (TGO): 96.3 U/L, ALT (TGP): 115 U/L, Serum creatinine: 1.06, TSH: 3.16.

Antihypertensive treatment sequences were initiated, but no apparent improvement was observed. A cesarean section was performed based on maternal extraction criteria due to severe refractory systemic arterial hypertension. The newborn male had a birth weight of 500 grams, with Apgar scores of 0 at both the first and fifth minute, and a Ballard score corresponding to 24 weeks gestation. Given the patient's history of hypertension and current clinical condition, a brain CT scan was performed, showing findings suggestive of **posterior reversible encephalopathy**. An abdominal ultrasound also revealed an **occupying process in close contact with liver segment VI,** prompting further evaluation with a **triphasic abdominal CT,** which reported: A mass in the region of the adrenal gland, with **pheochromocytoma** considered the leading diagnostic possibility (based on clinical history), **hepatomegaly, bilateral duplicated renal collecting systems,** pneumoperitoneum and **subcutaneous emphysema** of the abdominal wall and bilateral basal atelectasis.

Complementary tests were conducted, producing the following results: cortisol: 18.16 µg/dL, urinary catecholamines: 98.6 mg/24 hrs, metanephrines: 0.77 mg/24 hrs, all within normal range, CK-MB: 57.1 U/L, troponin I: < 0.100 ng/mL, D-dimer: 1775.00 ng UEF/mL. The patient underwent surgery with the indication of right adrenal mass resection, revealing a perioperative finding of an adrenal mass approximately 6 cm in diameter with macroscopic features of malignancy. The surgical

From the Department of Radiology at Hospital Regional de Occidente. Quetzaltenango, Guatemala. Received on June 25, 2025. Accepted on July 1st , 2025. Published on July 2nd, 2025.

DOI 10.5281/zenodo.15792960

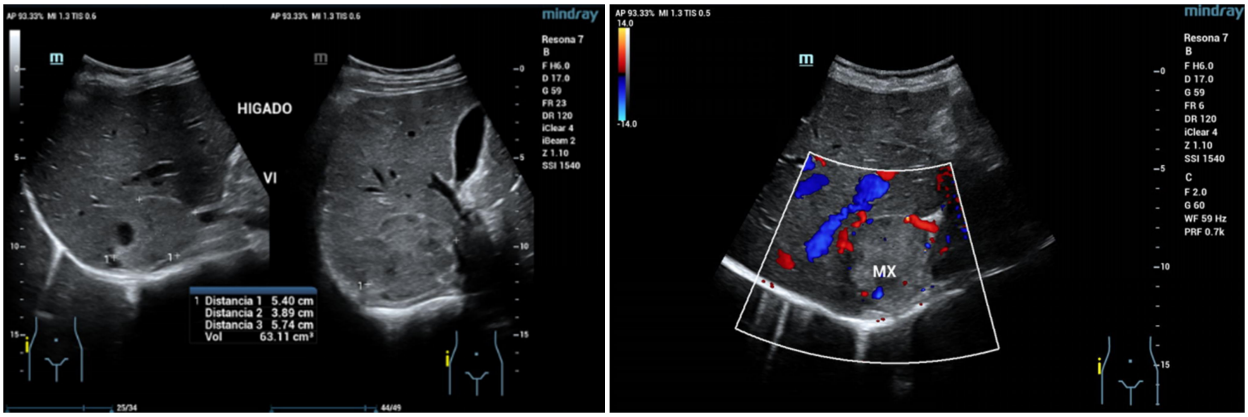

FIGURE 1. Images obtained via ultrasound: (a) in B-mode and (b) with color Doppler application. A solid-appearing mass is visualized superior to the right renal pole and in close contact with liver segment VI, according to Couinaud's anatomical classification. The mass appears isoechoic relative to the hepatic parenchyma, with well-defined margins and some hypoechoic areas within, suggesting necrosis, and displays peripheral vascular flow.

specimen was sent to pathology for histopathological and immunohistochemical study, which reported adrenocortical carcinoma.

Discussion

ACC (Adrenocortical Carcinoma) can produce excessive levels of cortisol, androgens, estrogens, or aldosterone, although hormonal excess may not always be present. Most adrenal tumors are benign—primarily adenomas—accounting for up to 80% of cases. Malignant tumors are rare, with adrenocortical carcinoma being of primary concern due to its aggressiveness. Prognosis largely depends on the stage at diagnosis, with five-year survival rates of 60–80% for stages I and II, 35–50% for stage III or locally advanced disease, and 13% for stage IV.

CLINICAL MANIFESTATIONS

Approximately half of ACC is functioning with signs and symptoms of hormone secretion. Patients with functioning ACC were younger, more likely to be females and present with metastatic disease. Of functioning tumours, almost half present with signs or symptoms of cortisol excess (Cushing syndrome). The second most common functional presentation was sex hormone secretion accounting for 20% of cases. These sex hormone-producing ACCs are mainly androgens. Rarely, estrogen secreting/feminizing ACC were reported. Aldosterone secreting ACC is uncommon and accounted for approximately 8% of cases. In addition, mixed hormone production was seen in 15% to 25% of functioning ACCs. Non-functioning ACCs commonly present with abdominal mass, abdominal pain as well as general symptoms of malignancy. Paraneoplastic manifestations of having ACC such as hypoglycaemia (due to insulin growth factor 2 [IGF2] production) and clinical manifestations related to adrenocorticotropic hormone ACTH production have been reported. Rare manifestations of patients with ACC include cancer-related thrombotic microangiopathy and tumour rupture with retroperitoneal haemorrhage. ACC could be a cancer in the setting of several hereditary syndromes. These syndromes could occur in high as 5–10% of patients with ACC. In adult patients with ACC, these

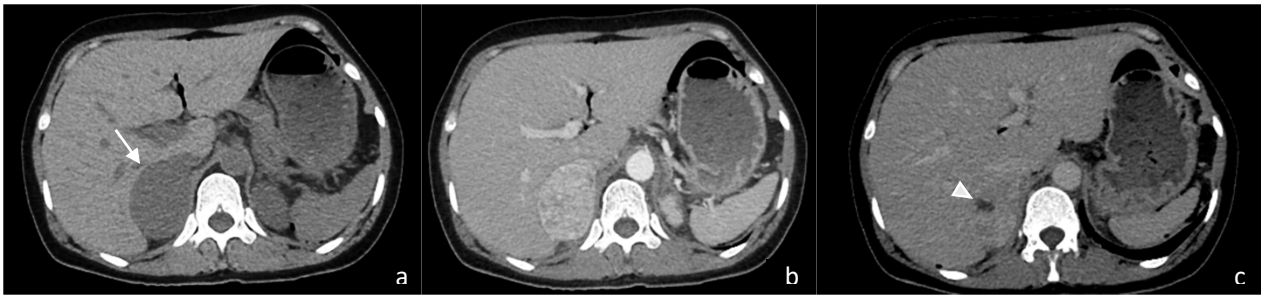

FIGURE 2. Axial slices from an abdominal CT scan. (a) Non-contrast phase showing an occupying lesion at the level of the right adrenal gland (thin arrow), which appears hypodense relative to the liver tissue, homogeneous, with well-defined borders, and measuring approximately 5.9 × 4.3 × 5.4 cm, with attenuation values up to +52 Hounsfield units (HU). (b) Arterial contrast phase showing the same mass with heterogeneous enhancement, with attenuation values reaching +144 HU. Some internal hypodense areas are seen that do not enhance, corresponding to necrotic regions and suggesting a malignant component. (c) Venous phase showing washout of the intravenous contrast medium, with previously described necrotic areas still visible (arrowhead).

DOI 10.5281/zenodo.15792960

FIGURE 3. Coronal slices from abdominal CT scan in the arterial phase (a) and delayed venous phase (b), showing the previously described findings.

syndromes include multiple endocrine neoplasia type 1 (MEN1, approximately 20 cases reported), Lynch syndrome (mismatch repair genes) (approximately 10 cases), Li-Fraumeni syndrome (TP53, more than 10 cases) and neurofibromato sis type 1 (NF1, approximately 10 cases). Rarely, ACC can occur in patients with Carney complex (protein kinase A regulatory subunit 1A [PRKARIA]), Gardner's syndrome and familial adenomatous polyposis (adenomatous polyposis coli [APC]). Although extremely rare, ACCs have been reported in patients with congenital adrenal hyperplasia and myelolipoma. It is likely these are collision tumours rather than having been aetiologically related.

DIAGNOSIS BY CT SCAN

ACC (Adrenocortical carcinoma) typically presents as a large, well-circumscribed mass with heterogeneous attenuation due to cystic and/or necrotic changes, hemorrhage, and calcification. Smaller ACCs

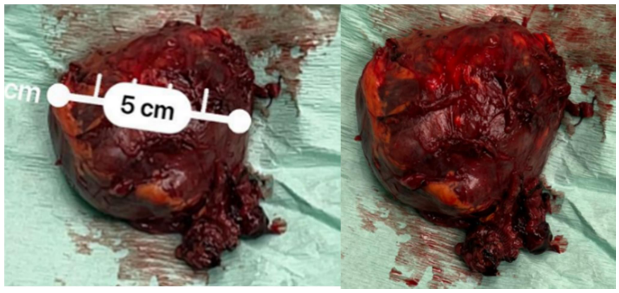

FIGURE 4. Surgical specimen of the adrenal occupying lesion following excision, showing a mass measuring approximately 5 × 6 cm.

can appear homogeneous and may be difficult to distinguish from adenomas, especially if they measure less than 4 cm. On non-contrast CT imaging, ACC rarely shows attenuation below 20 Hounsfield units (HU), which helps differentiate it from adenomas. ACC enhances heterogeneously, with central non-enhancing areas due to necrosis or hemorrhage. Although data are limited, ACC often shows an absolute washout below 60%, but it can sometimes exceed washout thresholds typically associated with adenomas.

Imaging studies play a key role in the diagnostic approach, as they provide information that can indicate the behavior of malignant tumors. It's important to remember that adrenocortical carcinomas (ACC) typically present as unilateral tumors, measuring more than 6 cm in 90% of cases, and often exhibit necrotic areas. On computed tomography (CT), these tumors are characterized by densities greater than 10 Hounsfield units (HU), with a sensitivity of 93–100% and a specificity of 71–72%, and show slow contrast washout (< 50%).

In the diagnostic approach, a metabolic panel should be performed that includes mineralocorticoids, glucocorticoids, androgens, and catecholamines, which will help differentiate between adrenocortical carcinoma (ACC) and other tumors such as pheochromocytoma. In ACC, elevation of androgens and cortisol is commonly observed.

CLASSIFICATION

The World Health Organization classifies adrenocortical nodular disease into three subtypes: (a) sporadic nodular, (b) bilateral micronodular, and (c) bilateral macronodular. The latter two entities are associated with germline mutations and are distinguished according to nodule size (<1 cm for micronodular, ≥1 cm for macronodular) and affected patient population (children and young adults for micronodular, adults for macronodular). Bilateral macronodular adrenocortical disease is a rare cause of bilateral adrenal nodules that can result in clinical hypercortisolism or mild autonomous cortisol

DOI 10.5281/zenodo.15792960

secretion (previously subclinical hypercortisolism. Differential considerations include bilateral metastases or adenomas. Other less common causes of bilateral nodules include pheochromocytoma, congenital adrenal hyperplasia, Cushing disease, and secondary adrenal hyperplasia from ectopic ACTH production.

TREATMENT

Treatment varies depending on the stage of the disease. Whenever possible, the initial treatment is surgery to remove the affected adrenal gland. In some cases, surgery is also performed to remove nearby lymph nodes and other tissues where the cancer may have spread. Mitotane is a drug that works by destroying adrenal cells and is used after surgery in cases with moderate to high risk of recurrence. Chemotherapy is a complementary treatment to surgery in cases of high recurrence risk or when the cancer has already returned. It typically involves intravenous medications given in cycles, aiming to stop cancer cell formation by destroying them or preventing their multiplication. Radiotherapy may be used as an additional treatment to surgery in some cases to prevent tumor recurrence. New types of treatments are currently being tested in clinical trials, such as immunotherapy and targeted therapy.

Conclusion

Computed tomography (CT) is the method of choice for diagnosing adrenal masses, and key findings are highly important for distinguishing adrenocortical carcinoma (ACC) from other adrenal lesions such as pheochromocytoma, adrenal adenoma, among others. Findings such as size greater than 6 cm, heterogeneous enhancement following intravenous contrast administration, presence of necrotic areas, cystic degeneration, or calcifications within the mass are indicative of malignancy. Surgical resection is the standardized treatment method for ACC.

Conflicts of interests

None declared by the authors.

Acknowledgements

To the administrative and medical personnel of Hospital Regional de Occidente for providing us with the necessary supplies and conditions in the radiology department to help in the diagnosis of this and other patients with poor economical background.

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 DOI 10.5281/zenodo.15792960