



Malignant Adrenocortical Carcinoma - Case Report

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Abstract

Adrenocortical carcinoma (ACC) is a rare endocrine cancer, with poor prognosis and very aggressive. It has a rare incidence and it's more prevalent in pediatric and geriatric patients and women. It's a malignant neoplasm that has diagnostic by complementary exams and needs surgical treatment. May be occur pulmonary, hepatic and bones metastasis during the clinical development.

Keywords: Adrenocortical carcinoma; Adrenal Glands; Adrenalectomy

Introduction

Malignant adrenocortical carcinoma (ACC) is an endocrine neoplasm of adrenal gland origin of undefined etiology. It is, in itself, a rare entity, having an incidence of 1-2 cases per million inhabitants/year [1-4], and commonly affects patients from 40 to 60 years old [2]. In southern Brazil, the incidence in children is, on average, 3.5 new cases per million inhabitants/year, while the worldwide pediatric incidence is 0.2 to 0.3 per million inhabitants/year [1]. Although diagnosis occurs between the fifth to sixth decades of life, commonly [1], it can be done at any age. Patients diagnosed with this neoplasm have poor prognosis, since it is a very aggressive tumor [2,3]. Risk factors include gender (female), age (under 5 years and 4th and 6th decades of life) and genetic inheritance [2,5-7].

This article aimed to discuss a clinical-surgical case of adrenocortical carcinoma at the Surgery, Urgency and Trauma Ser-

vice (SCUT) of the Celso Pierro Hospital and Maternity Hospital (HMCP), a teaching hospital of Pontifícia Universidade Católica de Campinas.

Case Report

An 62-year-old male patient, born in Pongaí, São Paulo, married, civil and rural worker, sought outpatient service of the HMCP by referral from the Basic Health Unit due to a suspected primary liver neoplasm. At admission, the patient reports progressive dyspnea to small and medium efforts for 8 months associated with dry cough and occasional wheezing, weight loss of 30kg in 2 months, and pain in right hypochondrium. He has tuberculosis of miliar pattern treated for 6 months, is hypertensive and diabetic, ex-smoker and former smoker. On physical examination, it is slightly deformed, lucid and oriented in time and space, without alterations to the examination

of the cardiovascular and pulmonary appliances. The abdomen was tympanic, with hepatic edge hepatomegaly at 7 cm from the costal rim, hardened and painful to deep palpation in the right lateral region, negative abrupt decompression. Complementary tests were requested – Computed tomography (CT) of the chest and total abdomen, upper digestive endoscopy, ultrasound and laboratory-guided biopsy – and pain analgesia. The patient was referred to the Pulmonology service for evaluation of the respiratory condition.

Chest CT showed centrilobular nodules, some with a “budding tree” (or “tree roots”) pattern, sparse through the pulmonary fields, especially in the middle, lower right and upper left lobes. There are also foci of consolidation (Figure 1) and cavities, diffuse thickening of the bronchial walls with content sums inside some bronchi (Figure 2) and diffuse mosaic attenuation areas in the pulmonary parenchyma, possibly related to ventilatory/perfusion disorder. Tomographic findings are compatible with previous infectious process.

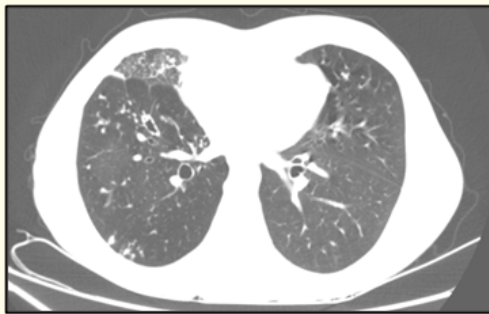


Figure 1: Contrast-enhanced chest tomographies.

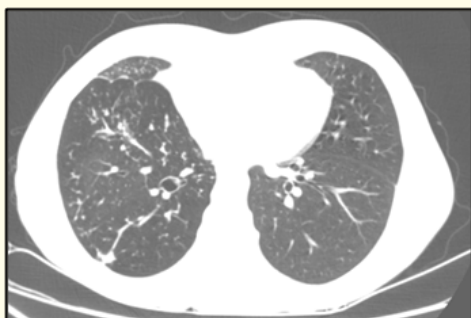


Figure 2: Contrast-enhanced chest tomographies.

Abdominal CT scan showed a large expansive lesion centered in the right adrenal store (Figures 3 and 4), which showed no suggestive signs of hepatic origin. Despite its large dimensions, it showed no characteristics of aggressiveness. There is also hepatic hemangiomas and expansive polypoid lesion in the sigmoid colon, with enhanced suspicion for neoplasia.



Figure 3: Computed tomography scans of total abdomen, identifying adrenal mass.



Figure 4: Computed tomography scans of total abdomen, identifying adrenal mass.

The anatomopathological of ultrasound guided biopsy demonstrated multifragmented material containing abundant necrosis, also showing proliferation of cells of intermediate size and pleomorphic aspect.

According to inconclusive biopsy, right adrenalectomy was indicated associated with retroperitoneal lymphadenectomy. Retroperitoneal tumor exeresis was performed by median laparotomy. A voluminous retroperitoneal tumor was observed that rejected the

right kidney inferiorly and had adhesions anteriorly by the liver; in addition to localized lesion with hemangioma aspect in hepatic segment II and absence of free fluid or peritoneal metastases. During the intraoperative period, the colon was palpated and a lesion enlashed in polypoid sigmoid was identified measuring about 2 cm in diameter and dissection. Beyond the lesion, it was performed a release of ascending colon with retroperitoneal opening.

The encapsulated, solid-cystic lesion, which rejected the right kidney inferiorly was adhered to the vena cava and also encompassed the ipsilateral adrenal gland, with a cleavage plane between the structures. Tumor dissection was performed, which weighed 2,355g (Figure 5), with lower vena cava release and undone its adhesions to the right liver and kidney, preserving them (Figure 6). On the 2nd postoperative day, the patient underwent new laparorrhaia by evisceration. On the 3rd postoperative day due to laparorrhania, the patient was hemodynamically stable, with good acceptance of a water diet, but still with absent evacuation. On the 6th postoperative day, the patient had good acceptance of a soft lactose-free diet, evacuation and diuresis present and was walking in abdominal band use. It was then given discharge with orientations of caring.



Figure 5: Adrenal mass removed with 2355g and compared with a cold blade scalpel handle.

Discussion

Adrenal tumors are significantly common, since most are benign, non-functional and small adrenocortical adenomas, reaching about 7% of the population and, on the other hand, ACC are rare [1]. Although it does not have a precise etiology, studies suggest that ACC can originate from adrenal cells at rest, forms of accessory

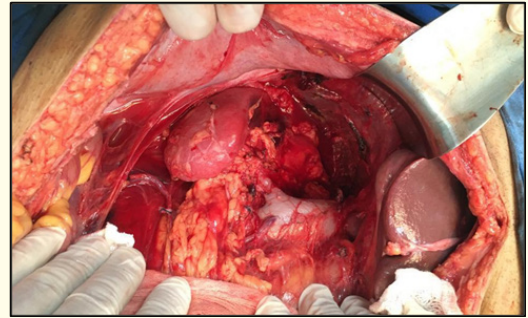


Figure 6: Cavity inventory after surgical removal of the mass.

or exuberant adrenocortical tissue [2]. In addition, a minority of cases are attributable to known hereditary predispositions, including Li-Fraumeni syndrome, Lynch syndrome, multiple endocrine neoplasia type 1 (MEN 1) and family polyposa adenomatosis [4].

The size and weight of an adrenal cortical neoplasm were initially considered useful parameters in the distinction of malignancy [8], especially for adenomas carcinomas to be differentiated [9]. Recent literature suggests a more complex evaluation, taking into account the serum dosage of hormones, blood count, electrolytes, coagulation tests, renal function, liver function, tumor markers, alkaline phosphatase and LDH, via laboratory tests; CT, Magnetic Resonance and Abdominal Ultrasound, through imaging and, finally, scintigraphic criteria for the diagnosis of malignancy [2,6,7,10-13]. The present study is in accordance with the literature and only one of the three diagnostic methods – imaging: abdominal CT – was necessary in order to confirm the hypothesis and to achieve surgical removal.

Surgical management is dominant, according to the literature, beyond bringing a better chance of cure to patients [14]. In this sense, the intervention may occur via laparoscopic adrenalectomy [15] or open adrenalectomy, by exploitative laparotomy, the latter being the choice in the resection of ACC [10,14], including by the SCUT team that handled the patient in the case.

Regarding evolution and clinical picture, the literature shows that, in patients with ACC with or without hormonal complaints, the manifestations of signs and symptoms occur within 6 months [16,17]. In addition, patients with ACC may have metastases throu-

ghout the diagnosis, mainly affecting lungs, retroperitoneal lymph nodes, liver and bones [16].

Despite having female gender as a risk factor [2,5-7], the present study goes against the literature, since it is a male patient, showing, once again, its academic importance and rarity.

Given the severity of the case, it is possible to say that the treatment did not end with surgical resection of the mass. Consultations and outpatient segment, in addition to adjuvant therapy, are fundamental for the management of patients with ACC. Adjuvant therapy, when necessary, is justified because there is a high chance of recurrence, 60-70%, despite surgical excision [1,14]. It is necessary, therefore, individualized treatment of the patient, taking into account his history and clinical picture.

Conclusion

ACC stands out as clinical entities not only because of its rarity and low incidence, but also because they are a challenge in the operating room given the difficult location and management of the patient. It is a cancer that the only potentially curative and good prognostic is surgical. The clinical history of the patient combined with the findings in the imaging tests were decisive points for the correct diagnosis and resulted in the procedure performed - adrenalectomy. Therefore, it is a pathology that requires multi - and interdisciplinary care, involving the Surgery, Radiology and Clinical teams for complete patient care

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