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Case Report

Right atrial and ventricular invasion by adrenal carcinoma: A case report

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ABSTRACT

Adrenocortical carcinomas (ACC) are rare but highly aggressive neoplasms. Intra-cardiac involvement with ACC is extremely rare and usually develops by intravascular invasion through the inferior vena cava (IVC). Complete surgical resection remains the most effective treatment. ACC prognosis is poor with a five-year overall survival rate of ~35%. The poor prognosis may be related to the advanced stage at which the majority of adrenal carcinomas are detected. We encountered a 52-year-old male patient with a huge right adrenal mass with a tumor thrombus invading the IVC towards the right atrium and another tumor mass present in the right ventricle below the tricuspid valve. A whole-body scan revealed metastases everywhere. The patient started palliative chemotherapy and radiotherapy. Later, during a debulking surgery of the tumor with cardiopulmonary support the patient died.

<**Learning objective:** Adrenocortical carcinomas (ACC) are rare highly aggressive neoplasms. Intra-cardiac involvement with ACC is extremely rare and usually develops by intravascular invasion through the inferior vena cava. The prognosis is usually poor due to delayed presentation and diagnosis and difficult management options.>

Introduction

Adrenocortical carcinomas (ACC) are rare highly aggressive neoplasms. Intra-cardiac involvement with ACC is extremely rare and usually develops by intravascular invasion through the inferior vena cava (IVC). The prognosis is usually poor due to delayed presentation and diagnosis and difficult management options.

Case report

A 52-year-old male patient with no past medical or surgical history presented to our medical facility complaining of constitutional symptoms, bone aches, loin pain, and abdominal distension and mild bilateral lower limb edema for 6 months. Abdominal ultrasonography (US) revealed a large right suprarenal mass invading the IVC. All his laboratory work-up including plasma free metanephrines, aldosterone, potassium, and testosterone came back negative. Multi-detector computed tomography (MDCT) of the abdomen and pelvis revealed a large heterogenous suprarenal mass measuring 11 × 10 cm (Fig. 1) with extension to the IVC. A transthoracic echocardiographic evaluation (TTE) revealed the presence of a solid mass in the right atrium extending through the IVC and another solid one in the right ventricle immediately below the tricuspid valve with minimal valvular regurgitation (Fig. 2). Transesophageal echocardiographic evaluation (TEE) confirmed the invasion of the right atrium with a tumor thrombus extending through the IVC with another solid mass just below the tricuspid valve (Fig. 3). The patient was referred to an oncologist who ordered a whole-body scan that revealed multiple bone metastases. So, he was diagnosed with stage VI adrenal carcinoma with TMN classification of T4N3M1.

The patient started palliative therapy including radiotherapy, chemotherapy with etoposide and cisplatin, and pain management with no clinical improvement or reduction in the size of tumor. Two months later, the patient went for a debulking surgery of the

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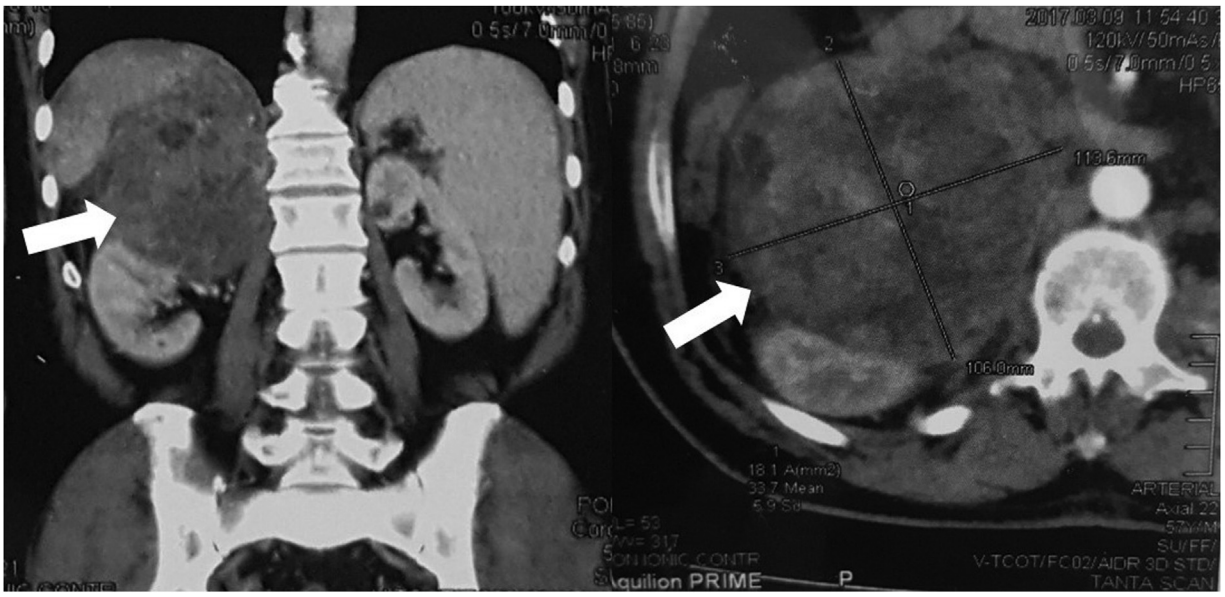

Fig. 1. Multidetector computed tomography of the abdomen and pelvis (sagittal and coronal views) showing large suprarenal mass measuring (11 × 10 cm) (arrows).

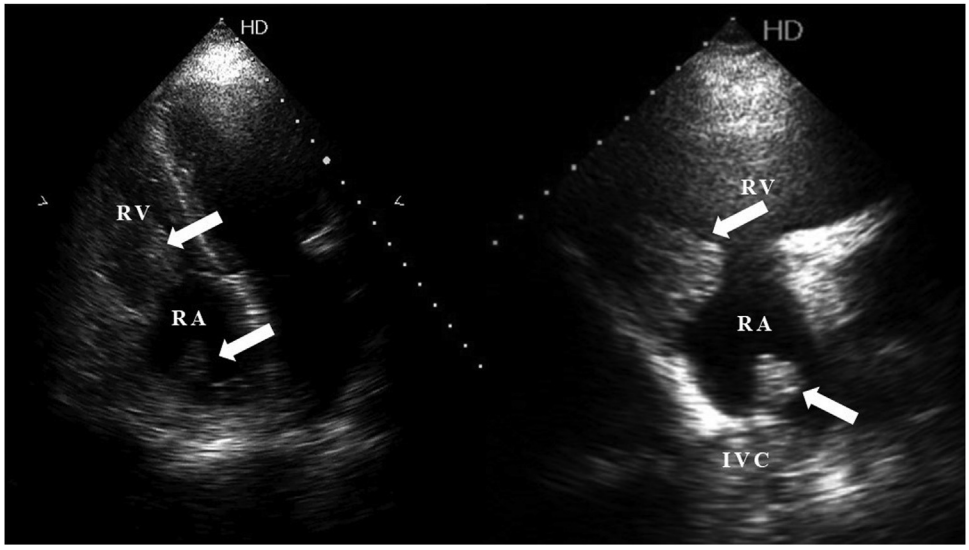

Fig. 2. Transthoracic echocardiography (apical 4-chamber and short-axis views) showing two masses, one extending from the inferior vena cava (IVC) to the right atrium (RA) and another in the right ventricle (RV) just below the tricuspid valve (arrows).

tumor for symptomatic relief of severe abdominal distension and compressive symptoms with a cardiopulmonary bypass support, but he died during the surgery due to major bleeding.

Discussion

ACC is a rare, but highly aggressive type of tumor with an annual report of 1–2 cases per million [1]. ACC has two peaks of age distribution: in children in the first decade of life and in adults in the fourth to fifth decades of life [2], with a higher reported incidence in females [3]. Patients can present with a variety of signs and symptoms, depending on the extent of the tumor. Up to 40% of adrenal tumors are nonfunctional with a very late clinical manifestation during the course of disease as a large mass causing compressive symptoms such as early satiety, weight loss, and abdominal pain or distension [4]. There have been some reports of functional adrenal tumors commonly in the form of mixed hormonal syndrome of hypercortisolism and virilization, isolated virilization, and “pure” Cushing syndrome, or rarely as isolated primary hypermineralocorticism [5]. IVC tumor thrombus extension is a rare presentation of this pathology. IVC tumor thrombus involvement can occur by either direct invasion or, more commonly, by intraluminal extension of the tumor thrombus via the adrenal or renal veins [6]. US of the adrenal gland has proven to be an effective technique for screening of adrenal masses but depends substantially on operator skills. Obesity, overlying gaseous distention, or lack of patient’s cooperation remain major drawbacks for US [5]. Multidetector computed tomography (MDCT) of the chest and the abdomen is representing to date, the gold standard for ACC diagnosis and staging. With regards to the tumor thrombus, magnetic resonance imaging represents the better imaging modality in order to assess the extension and

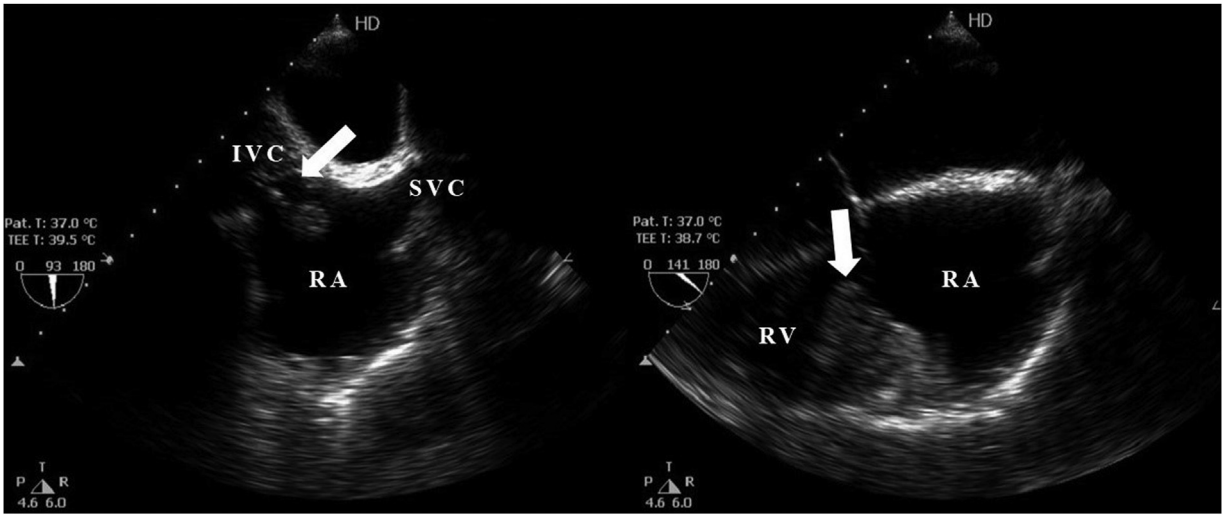

Fig. 3. Transesophageal echocardiography showing two masses, one extending from the inferior vena cava (IVC) to the right atrium (RA) and another in the right ventricle (RV) just below the tricuspid valve (arrows).
SVC, superior vena cava.

features of tumor thrombus [3]. TTE and TEE have been the main tools for diagnosis and follow up of all intracardiac masses. Recent advances in cardiac imaging including MDCT, cardiac magnetic resonance, and positron emission tomography scan can help in further tissue characterization of different cardiac masses as well as detection of any complications [7]. Complete surgical resection with a safety margin is the only curative option for localized disease [2]. The presence of a thrombus represents an additional risk factor due to the additional complexity of the surgery. So, for tumors extending into the atrium, the institution of a cardiopulmonary bypass as well as a cardiothoracic surgical assistance is mandatory [8]. The role of surgery in patients with recurrent and metastatic disease remains controversial [9]. Although a debulking surgery (R2 resection), did not significantly improve progression-free survival, but aggressive surgery should be considered even in the setting of most advanced ACCs probably offering the only chance of survival for patients with this aggressive tumor [3]. Even with complete resection, 50–80% of patients develop relapse and/or progression to metastatic disease. The prognosis is poor with a five-year overall survival rate of ~35% [3].

Conclusion

ACC is a highly aggressive malignancy with rare cardiac invasion through the IVC tumor thrombi with low survival rates which are related to the advanced stage of ACC detection. Complete surgical resection remains the most effective treatment and, along with an early staging, is among the strongest predictors of overall survival.

Disclosure

The authors declare no conflict of interest.

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