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Department of Imaging Sciences, University of Rochester Medical Center, Rochester, USA

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CASE REPORT

Functioning Adrenocortical Carcinoma with Extension upto the Right Atrium Producing Cushing's Syndrome

Santosh Kumar, Gautam R. Choudhary, Arawat Pushkarna

Departments of Urology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Address for correspondence:
Dr. Santosh Kumar,
Department of Urology, Postgraduate Institute of Medical Education and Research, Chandigarh - 160 012, India.
E-mail: santoshsp1967@yahoo.co.in

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ABSTRACT

Adrenocortical carcinoma (ACC) is a rare malignancy with poor prognosis. Surgery is the only curative therapy available and overall 5-year survival for patients who undergo a complete resection is 32% to 48%. They are known to produce intravascular invasion and into the inferior vena cava (IVC) and in rare cases they may reach the right atrium. We report a case of functioning ACC extending into the inferior vena cava and right atrium in a female with Cushing's syndrome.

Key words: Adrenocortical carcinoma, cushing syndrome, inferior vena cava thrombus, right atrium thrombus

INTRODUCTION

Adrenocortical carcinoma (ACC) is a rare endocrine malignancy with poor prognosis causing up to 0.2% of all cancer deaths. Its annual incidence is 1 to 2 per million people. Women are afflicted more often than men, reported ratio being 1.5:1. Surgery is the only curative therapy available and overall 5-year survival for patients who undergo a complete resection is 32% to 48%.[1-3] Usual metastatic sites are the lung (71%), lymph node (68%), liver (42%), and bone (26%). They are also known to produce intravascular invasion and into the inferior vena cava (IVC) and in rare cases they may reach the right atrium.[1] This is however a rare phenomenon and only a few cases have been reported in the literature.

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CASE REPORT

A 40-year-old female presented with a 7-month history of intermittent right flank pain in the loin region and features of Cushing's syndrome such as weight gain, central obesity, bipedal edema, moon facies, hirsutism, oligomenorrhea, recent onset of hypertension and diabetes mellitus (treated with insulin). There was no history of persistent headache, vomiting, and focal neurological symptoms. Her electrolytes, renal and liver function tests were within normal limits. Hormonal profile testing revealed

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markedly elevated serum 8 AM cortisol (1177 nmol/L) with loss of diurnal variation (11 PM cortisol - 800 nmol/L) which was not suppressible by Dexamethasone (LDDST {low dose dexamethasone suppression test} - 917.1 nmol/L; HDDST {high dose dexamethasone suppression test}- 812.9 nmol/L). Adrenocorticotropic hormone (ACTH) was suppressed (1.89 pg/mL), serum testosterone was elevated (6.3 nmol/L) with suppressed luteinizing hormone (LH) 1.14 mIU/mL (normal in follicular phase 2.4-12.6mIU/L) and follicle-stimulating hormone (FSH) 1.15 mIU/mL (normal in follicular phase 3.3-12.5 mIU/L). Ultrasound abdomen revealed a hypoechoic lesion in the right lobe of liver of size 10.1 × 8.2 cm. On contrast enhanced computed tomography (CECT) abdomen there was a well-defined heterogeneously enhancing mass lesion 10 × 9 cm with necrotic areas in right suprarenal location with infiltration into the segment VI and V of the liver and tumor thrombus in the IVC reaching up to the right atrium [Figure 1]. Left adrenal gland was normal. positron emission tomography (PET CT) with computed tomography scan showed intense FDG uptake {standardized uptake values} (SUV) max 8.6) [Figure 2].

The patient underwent a bilateral subcostsal incision with midline sternotomy for excision of tumor mass with removal of intracaval and right atrial extension for which she required a cardiopulmonary bypass. Tumor planes with the inferior surface of the liver were lost and during resection there was liver parenchymal injury from where she had severe bleeding. Packing of the abdominal cavity and abdomen closure had to be done with plans for pack removal 48 hours later, by which time bleeding had subsided. Specimen excised revealed complete removal of tumor with the thrombus [Figure 3]. Post-operation the patient was kept on a ventilator and inotropic support, which was gradually withdrawn. Histopathology of the specimen confirmed the diagnosis of adrenocortical carcinoma with tumor thrombosis [Figure 4].

DISCUSSION

ACC is a rare malignant neoplasm with propensity for vena caval tumor extension and metastasizes to the lung, lymph node, liver, and bone. Small non-functioning ACC patients usually have no symptoms but as their tumors reach a large size they cause symptoms due to mass effect and compression of nearby structures. Usually these symptoms are vague and include abdominal fullness, nausea, obstipation, or early satiety, weight loss, weakness, fatigue, or fever. About 40-60% patients have functioning ACCs of which 75% show occult hypercortisolism demonstrable only with hormone testing. A lesser proportion of patients will present with overt Cushing syndrome.[1] Ultrasonography can detect the presence of mass and combined with Doppler study can delineate the extent of venal caval thrombus extension. Transesophageal ultrasonography is especially useful intra-operatively when the upper end of the tumor thrombus extends beyond and above the diaphragm.[4,5] On cross-sectional imaging, adrenal cortical carcinomas tend to be larger than benign adrenal tumors, with an average size of 10 to 12 cm on presentation. Indeed over 90% of ACCs are greater than 5 cms, four to 5% of tumors are less than 4 cm, 10% of tumors are larger than 4 cm, and 25% of tumors are greater than 6 cm.[6] Given the relationship between adrenal tumor size and malignancy, it is currently recommended that incidentally detected

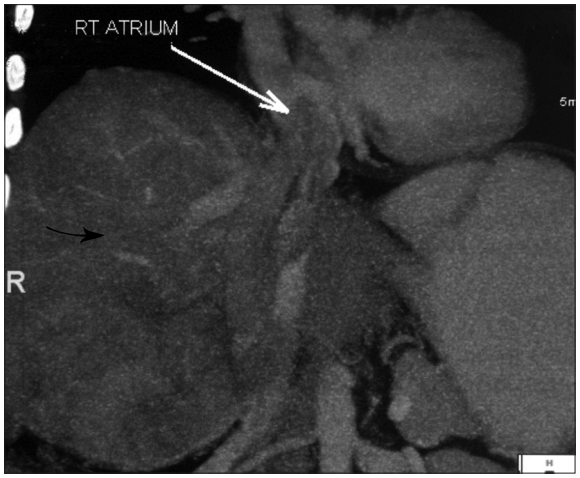

Figure 1: 40-year-old female with history of intermittent right flank pain and features of Cushing's syndrome diagnosed as having adrenocortical carcinoma with tumor thrombosis. CECT Abdomen shows a well-defined heterogeneously enhancing mass lesion in right suprarenal location (black arrow) with tumor thrombus in the IVC reaching up to the right atrium (white arrow).

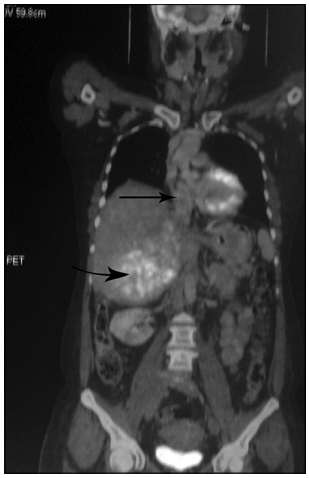

Figure 2: 40-year-old female with history of intermittent right flank pain and features of Cushing's syndrome diagnosed as having adrenocortical carcinoma with tumor thrombosis. PET CT scan shows intense FDG uptake (SUV max 8.6) in the right suprarenal tumor (solid curved arrow) and the intracaval tumor thrombus (solid straight arrow).

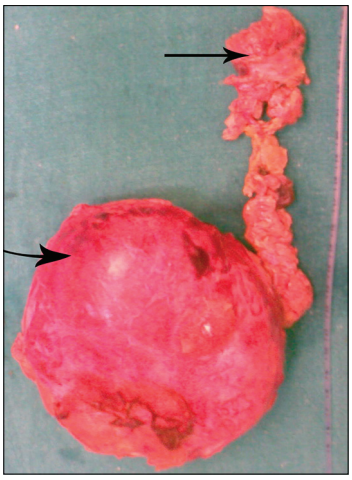

Figure 3: 40-year-old female with history of intermittent right flank pain and features of Cushing's syndrome diagnosed as having adrenocortical carcinoma with tumor thrombosis. Entire surgical specimen shows the adrenal tumor (solid twisted arrow) along with the tumor thrombus (solid straight arrow).

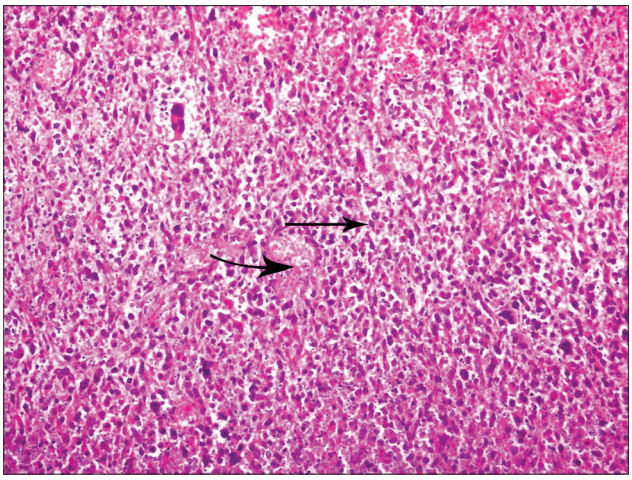

Figure 4: 40-year-old female with history of intermittent right flank pain and features of Cushing's syndrome diagnosed as having adrenocortical carcinoma with tumor thrombosis. Photomicrograph of the adrenal tumor shows moderately differentiated tumor cells in sheet with bizarre nuclei (solid straight arrow) and intervening vessels (solid twisted arrow) (H and E, x100).

adrenal tumors greater than 4 to 6 cm be surgically excised. All functioning adrenal tumors however, must be removed regardless of their size. CT images show the presence of irregular borders, irregular enhancement, calcifications, and necrotic areas with cystic degeneration. Mean attenuation on non-contrast CT scan in ACC is significantly higher (39 HU) compared with adenomas (8 HU).[5] Moreover, they will not demonstrate the contrast washout characteristics of benign adrenal adenomas. Adrenal adenomas are by definition benign and the vast majority are metabolically silent. Lesions that "washout" more than 40% to 60% of gained enhancement can be identified as adenomas with specificity that approaches 100%. MRI provides valuable information when evaluating adrenal tumors. ACCs appear isointense relative to the liver or spleen on T1-weighted images and demonstrate intermediate to increased intensity on T2-weighted images. On gadolinium-enhanced images, adrenal carcinomas demonstrate marked contrast uptake. In cases of suspicion for venous tumor thrombus, MRI can be an essential tool in detecting the presence of a tumor clot and delineating its extent. Other differential diagnoses of adrenal masses include angiomyoplipomas which can typically be differentiated on computed tomography and reveals a well-circumscribed adrenal lesion with varying amounts of mature adipose tissue (30 HU) interdigitated with higher-density myeloid components, which enhance upon contrast administration.[4-6]

Intracaval tumor thrombus extension is a special property of adrenocortical carcinomas which, however, is a rare phenomenon and only a few cases have been reported in world literature. However, almost all the cases reported in literature have been of non-functioning tumor. Hedican and Marshall reported 3 cases and reviewed 26 previously reported cases of ACC. According to their report, about half of cases showed direct invasion of IVC and extension upto right atrium.[2] Kim KH reported a case of large ACC extending up to the right atrium, where the patient refused surgery but the tumor showed spontaneous regression during follow-up.[7] Okazumi et al., also reported a case of ACC with extension up to the right ventricle.

Functioning tumor generally get detected early because of the various phenotypic and functional changes they produce. Our patient had classical Cushing's syndrome produced by a highly extensive adrenocortical carcinoma invading up to the right atrium. Approximately, 60% of ACC cases presents with symptoms and signs of adrenal steroid hormone excess.[1] Because chemotherapy has limited value in the treatment of ACC, complete surgical resection is the treatment of choice.[3] Several reports suggest that complete surgical removal of primary tumor with cavoatrial mass using a cardiopulmonary bypass is the treatment of choice.[2] However, our patient succumbed to overwhelming post-operative sepsis. Patients with Cushing's syndrome are known to be immunosupressed with propensity for rapid development of sepsis refractory to usual treatment modalities. Post operatively our patient developed pneumonia and ultimately expired secondary to post operative complications.

CONCLUSION

Functioning adrenocortical carcinoma are usually detected early because of wide spectrum of clinical manifestation. Our case exemplifies the fact that even these functional tumors can present at an advanced stage with tumor

thrombus extending up to right atrium making surgical resection a challenge. Imaging (US, CT, and MRI) plays an important role in differential diagnosis and in delineating the extent of the tumor thrombus.

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