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Renal Cell Carcinoma with Interseptal Metastasis

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ABSTRACT

We report a single case of renal cell carcinoma (RCC) with renal vein, suprahepatic inferior vena cava, and right atrium thrombosis. The patient subsequently underwent radical nephrectomy and thrombectomy with intraoperative pulmonary artery exploration and tumor thrombectomy. During surveillance, the patient presented with an intracardiac interventricular septal disease recurrence 40 months after resection. The patient elected adjuvant therapy with oral sunitinib for the unresectable cardiac metastasis and was free from clinical progression for 8 months at last followup. This is the first known report in the literature of RCC with intraseptal cardiac metastatic recurrence.

KEYWORDS: Renal Cell Carcinoma; Metastasis; Cardiac recurrence; Inferior vena cava thrombectomy; Sunitinib

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INTRODUCTION

Renal cell carcinoma (RCC) accounts for 2-3% of adult malignancies. It carries a higher mortality rate than prostate cancer or bladder cancer. Incidence of RCC has increased over the past 30 years, at least part of which can be attributed to earlier detection with computed tomography (CT) scans. Unique to RCC is its ability to grow into the renal vein with possible extension into the vena cava. A high percentage of these cases can be successfully treated with nephrectomy and IVC thrombectomy. Recurrence, unfortunately, is still common.

The present report summarizes a case of renal cell carcinoma with vena caval involvement and tumor extension into the pulmonary veins. After initial surgical removal of the tumor, the patient was found to have recurrence involving the ventricular septum. To the authors' knowledge, this is the first report of this recurrence pattern.

CASE REPORT

The patient presented as a healthy 49-year-old man complaining of gross hematuria. A review of systems revealed intermittent parasternal chest pain over the previous 6 to 12 months. The hematuria evaluation included abdominal imaging, which ultimately led to additional testing. Magnetic resonance imaging of the abdomen and pelvis demonstrated a heterogeneously enhancing 8.5 x 4.6 x 3.0 cm mass in the upper pole of the right kidney.Tumor thrombus filled the right renal vein and suprahepatic inferior vena cava, with apparent extension above the diaphragm into the right atrium (Figure 1a transverse; Figure 1b coronal). A metastatic survey was negative. The patient underwent right radical nephrectomy with vena caval thrombectomy under cardiopulmonary arrest. An intraoperative transesophageal echocardiogram demonstrated embolization of the atrial thrombus to the pulmonary artery. Thus, right pulmonary artery thrombectomy was performed. Pathologic staging revealed a Fuhrman grade III, pT3cN0Mx clear cell RCC.

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original study

Figure 1a. MRI image demonstrating the inferior vena caval involvement of the RCC at the level of the heptic vein.

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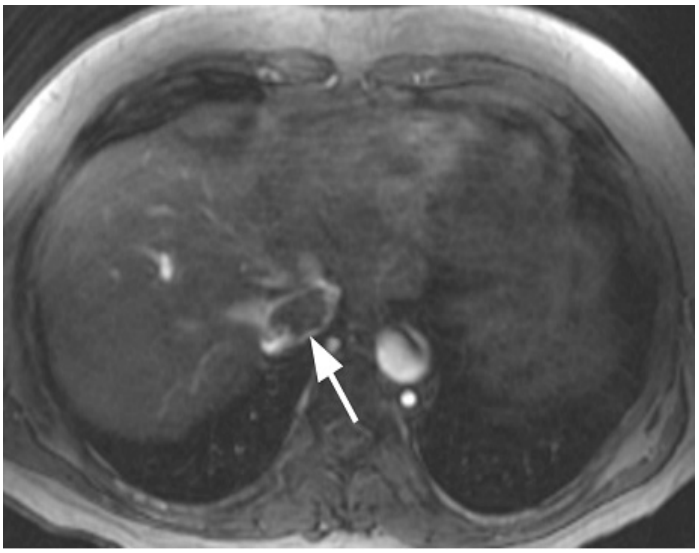

Figure 1b. Coronal MRI image demonstrating the cephalad extent of the RCC in the IVC (arrows)

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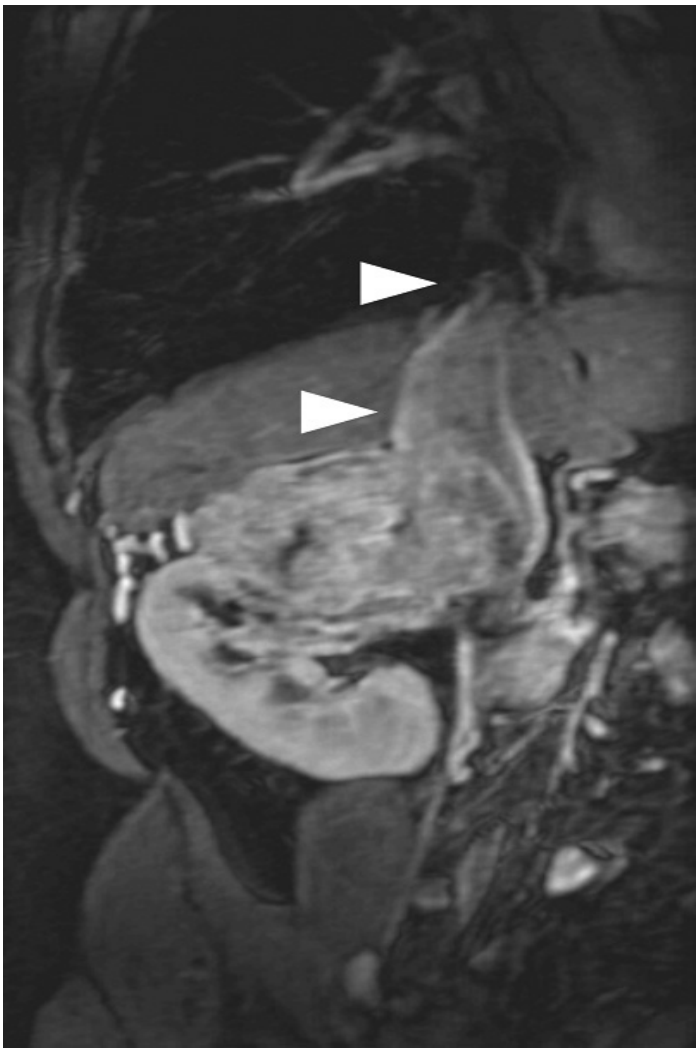

The patient was followed without recurrence until 40 months post-resection, when a surveillance computerized tomograpy scan of the chest revealed a 3.6 x 4.0 cm peripherally enhancing intracardiac mass, with central low attenuation consistent with necrosis (Figure 2). On echocardiogram, the lesion occupied the entire thickness of the proximal interventricular septum with extension into the left ventricular outflow tract causing mild associated flow acceleration (Figure 3). The mass also invaded an area at the base of the anterior aortic root and aortic valve right coronary cusp. The lesion was not free-floating or valve-based. Right and left ventricular biopsies obtained via right heart catheterization with transesophageal echocardiograpy and fluroscopic guidance were inconclusive. The patient ultimately required open cardiac biopsy of the interventricular septum, which revealed metastatic RCC. The lesion was deemed unresectable given the location. However, due to the cardiovascular risk associated with mass progression, the patient was treated with sunitinib.

DISCUSSION

In 2007 over 51,000 new cases of RCC were estimated, with nearly 13,000 patients dying of disease [1]. Survival is directly related to disease stage, tumor grade, and patient performance status [1]. Despite the poor survival associated with locally advanced disease, complete surgical resection provides long term cure in 40-50% of patients with pT3 tumors. Nonetheless, long-term outcomes are largely determined by the presence or absence of distant metastases. Renal cell carcinoma presents with solid metastases in approximately 20% of cases, yet cardiac involvement is rare [2,3]. Autopsy studies have reported the incidence of unrecognized cardiac metastasis as 2-3%, with approximately 8% of patients with known primaries harboring cardiac metastasis at the time of death [4,5]. In general, cardiac malignancies are rarely of primary cardiac origin (5%) and most cardiac tumors are secondary

Figure 2. Chest CT computerized tomograpy demonstrating 3.6 x 4.0 cm intracardiac necrotic mass.

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Figure 3. Echocardiography demonstrating lesion (arrows) occupying the interventricular septum with extension into the left ventricular outflow tract (LA = left atria, LV = left ventricle).

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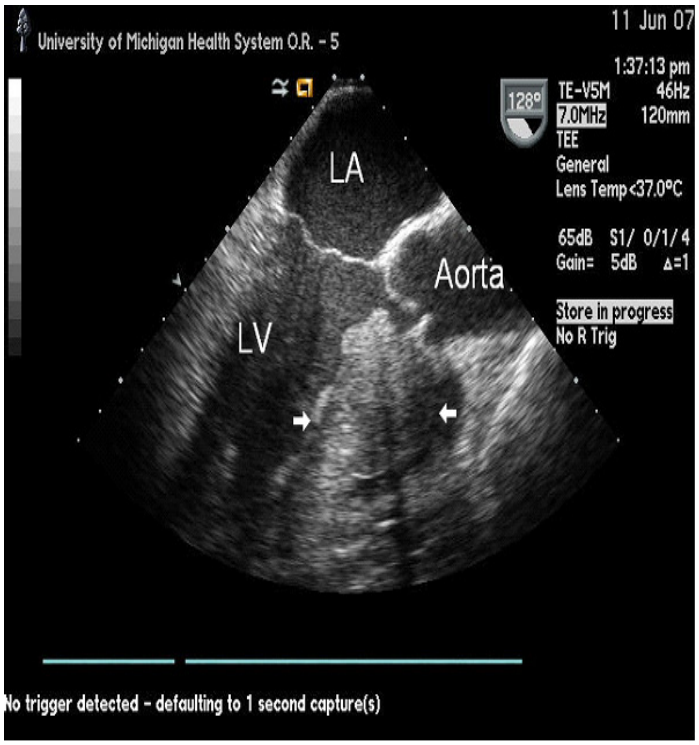

to carcinoma of the lung (32%), esophagus (30%), lymphoma (12%), and breast (7%) [6]. Intracardiac recurrences after surgical resection have rarely been described for breast, bronchus, and hepatocellular carcinoma, but when they occur, they are typically free-floating or valve-based implantations.

This patient is unique because the isolated metastatic disease to the interventricular septum could be related to tumor seeding during the pulmonary artery thrombectomy. Still, a remission of 40 months prior to recurrence seems unlikely. Although this patient remained asymptomatic during surveillance, most cases with cardiac metastasis to the ventricular myocardium report syncope or dyspnea as a presenting symptom [2,7,8]. Survival in the setting of metastatic RCC is limited, with the median survival approximating one year [9]. Although this patient's survival is poor due to metastasis, the long disease-free period after initial nephrectomy and his excellent performance status predict a longer duration of survival [9]. Adjuvant therapy with immune modulators or vascular endothelial growth factor (VEGF) inhibitors may delay progression, but complete and durable responses are infrequent and uncertain [10].

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