Sarcomatoid renal cell carcinoma: A case report and literature review

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ABSTRACT

Renal cell carcinoma (RCC) is the most common malignancy of the kidney and consists of multiple subtypes. The sarcomatoid variety, while previously considered a distinct histologic subtype, is now categorized as a form of dedifferentiated carcinoma. When present, it is associated with a significant decrease in patient survival due to its rapid growth and intrusive behavior. Preoperative knowledge of this diagnosis may be beneficial to clinicians in order to modify treatment options and follow-up protocols. This report describes a case of sarcomatoid renal cell carcinoma in which the patient initially presented with flank pain. We then discuss the clinical features of sarcomatoid renal cell carcinoma and its imaging appearance on computed tomography (CT), and succinctly review the subtypes of renal cell carcinoma and their imaging characteristics.

CASE REPORT

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An 87 year old woman presented with left flank pain. There were no pertinent physical exam findings and the only significant laboratory abnormality was microscopic hematuria. A computed tomography (CT) urogram was performed demonstrating a large heterogeneous mass measuring 6.0 x 5.2 x 5.5 cm within the inferior pole of the left kidney, with extention into the renal sinus. The lesion was isoattenuating with respect to the renal parenchyma on the noncontrast images and had a single thin curvilinear calcification anteriorly. The lesion also demonstrated multiple areas of low density centrally, suggesting necrosis. No macroscopic fat was noted in the mass. During the corticomedullary phase, there was enhancement of the more peripheral aspects of the mass, although the enhancement was less than the renal cortex. The solid enhancing component of the lesion remained mildly hypodense compared to the kidney during the nephrographic phase, while the previously described non-enhancing, low density portions became more conspicuous. Excretory phase imaging revealed questionable enhancing intraluminal soft tissue density within the left renal pelvis and proximal left ureter (Fig. 1). There was also mild pelvicaliectasis. There were several prominent left para-aortic lymph nodes, measuring up to 1.1 cm in short axis diameter. The left renal vein was patent and the adrenal glands were normal. The differential diagnosis at this point included renal cell carcinoma and subtypes, lipid poor angiomyolipoma, oncocytoma, transitional cell carcinoma, lymphoma and metastatic disease [1].

Given the possibility of urinary upper tract involvement, the patient was taken to surgery for left nephroureterectomy for potential transitional cell carcinoma. However, pathologic analysis revealed renal cell carcinoma of the chromophobe histologic subtype with significant sarcomatoid differentiation (98% of the total tumor volume), extending into the renal sinus (Fig. 2). The Fuhrman grade was designated as 4, and discrete areas of both hemorrhage and necrosis were identified. No tumor was identified within the ureter, making the filling defect within the ureter on imaging likely blood products.

The patient returned to the emergency department 5 months later with peritoneal signs. Other than a tender abdomen, physical examination and laboratory work up were non-contributory. CT of the abdomen and pelvis demonstrated

changes status post left nephroureterectomy, including a small fluid collection in the left renal fossa. A new 6.8 cm rounded and heterogeneous mass was present within the anterior left lower quadrant abutting the transverse colon, highly concerning for metastatic disease (Fig. 3). At surgical resection of this mass, there was colonic wall invasion without perforation. There were no other sites of involvement. Pathologic analysis confirmed the diagnosis of metastatic sarcomatoid renal cell carcinoma. The patient is currently undergoing conservative management with surveillance imaging.

DISCUSSION

Renal cell carcinoma (RCC) is the sixth most common neoplasm in adults with 56,000 new cases expected in 2011. It accounts for 3% of adult malignancies and more than 90% of renal cancers [2]. A history of cigarette smoking and obesity are risk factors that have the strongest correlation with RCC. According to the 2004 histologic classification of RCC by the World Health Organization (WHO), the clear cell histologic subtype is by far the most common, comprising 70% of all cases. Other less common histologic subtypes include papillary, chromophobe and collecting duct, which represent 10%, 5% and less than 1% of all renal cell carcinomas, respectively [3]. Histologic differentiation of these subtypes of RCC is clinically important, as both the papillary and chromophobe subtypes have a less aggressive clinical course and better prognosis than the clear cell variety. While initially felt to represent a primary renal sarcoma, sarcomatoid RCC is now considered a form of dedifferentiated carcinoma and is therefore not a distinct histologic entity. It is defined as any subtype containing foci of pleomorphic spindle cells and is seen in high grade RCC, at the end stages of disease progression. Sarcomatoid differentiation is reported to occur in approximately 1-8% of RCCs [4], and when present is indicative of an aggressive tumor as demonstrated by their rapid growth and poor prognosis. Several small series have shown a median survival of less than one year in patients with sarcomatoid differentiation [5]. The aggressive behavior of these tumors also results in nonspecific symptoms in 89% of patients, most commonly pain, as was present in this case [6]. While sarcomatoid transformation has been identified with virtually all histologic subtypes of RCCs, the chromophobe subtype is most frequently associated with a sarcomatoid component [7]. However, the associated RCC subtype does not seem to affect the patient's prognosis when sarcomatoid dedifferentiation is present given its aggressive behavior and high mortality. Surgical management has been the mainstay of treatment for RCC but does not improve the prognosis in those with sarcomatoid differentiation. This has prompted the use of chemotherapy in combination with nephrectomy in these patients and has seen promise in prolonging survival [8].

Recent studies suggests that differentiation of clear cell RCC from the other less frequent non-clear cell subtypes can be diagnosed by imaging. Both the papillary and chromophobe subtypes typically enhance to a lesser degree, when compared to the renal cortex, than clear cell on all three phases of post-contrast imaging (corticomedullary, nephrographic and excretory phases), which is postulated to be due to differences in the intratumoral vascularity. Typically, both papillary and chromophobe subtypes appear hypovascular when compared to renal cortex. The papillary subtype can occasionally show enhancement differences of less than 10 HU, mimicking a cyst [9]. Similar findings have been demonstrated on MRI. One series differentiated the three most common subtypes of RCC with statistical significance based on corticomedullary phase enhancement patterns showing that clear cell carcinomas enhanced avidly, chromophobe carcinomas enhanced moderately, while papillary carcinomas had the least enhancement [10]. Furthermore, diffusion weighted MR imaging may play a role in the differentiation of RCC subtypes [11]. The pattern of enhancement, in addition to the degree of enhancement, can also be useful in distinguishing between subtypes. Clear cell carcinomas frequently demonstrate a heterogeneous appearance with internal enhancing soft tissue and lower density regions which represent either necrotic or cystic changes. Clear cell carcinomas also may demonstrate loss of signal on out of phase MR imaging due to presence of microscopic fat which may be confounding. Non-clear cell carcinomas are more often associated with a homogeneous or peripheral enhancement pattern [12]. A spoke wheel pattern of enhancement has been described in some chromophobe carcinomas which is similar to the classic appearance of oncocytomas [13].

Investigation into the imaging features of sarcomatoid dedifferentiation of RCC has been limited. However, the importance of this histologic diagnosis cannot be understated given the high Fuhrman nuclear grade and the associated poor clinical outcome. The current trend is to report any sarcomatoid component seen at pathologic analysis, regardless of what percentage of the total lesion it comprises, as treatment and follow-up protocols may be affected. Therefore, it would be beneficial if the presence of sarcomatoid dedifferentiation could be suggested based on imaging characteristics, potentially with MRI. There is only one case series, to the authors' knowledge, describing the MRI appearance of sarcomatoid RCC [4]. Owing to its aggressive nature, necrosis is an expected feature of sarcomatoid dedifferentiation, and all nine cases demonstrated necrosis in this series. This is likely not a highly specific characteristic, however, as a significant number of conventional RCCs demonstrate this finding as well. Other findings which were seen in the majority of cases and indicate aggressive local behavior included renal sinus invasion and perirenal fat invasion. Further research into more specific imaging features of sarcomatoid transformation may be beneficial so that patient care can be modified accordingly.

TEACHING POINT

The presence of sarcomatoid differentiation in a patient with renal cell carcinoma portends a much worse prognosis than those without it. Awareness of the imaging characteristics of sarcomatoid renal cell carcinoma is important as this diagnosis may alter treatment plans and follow-up protocols.

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FIGURES

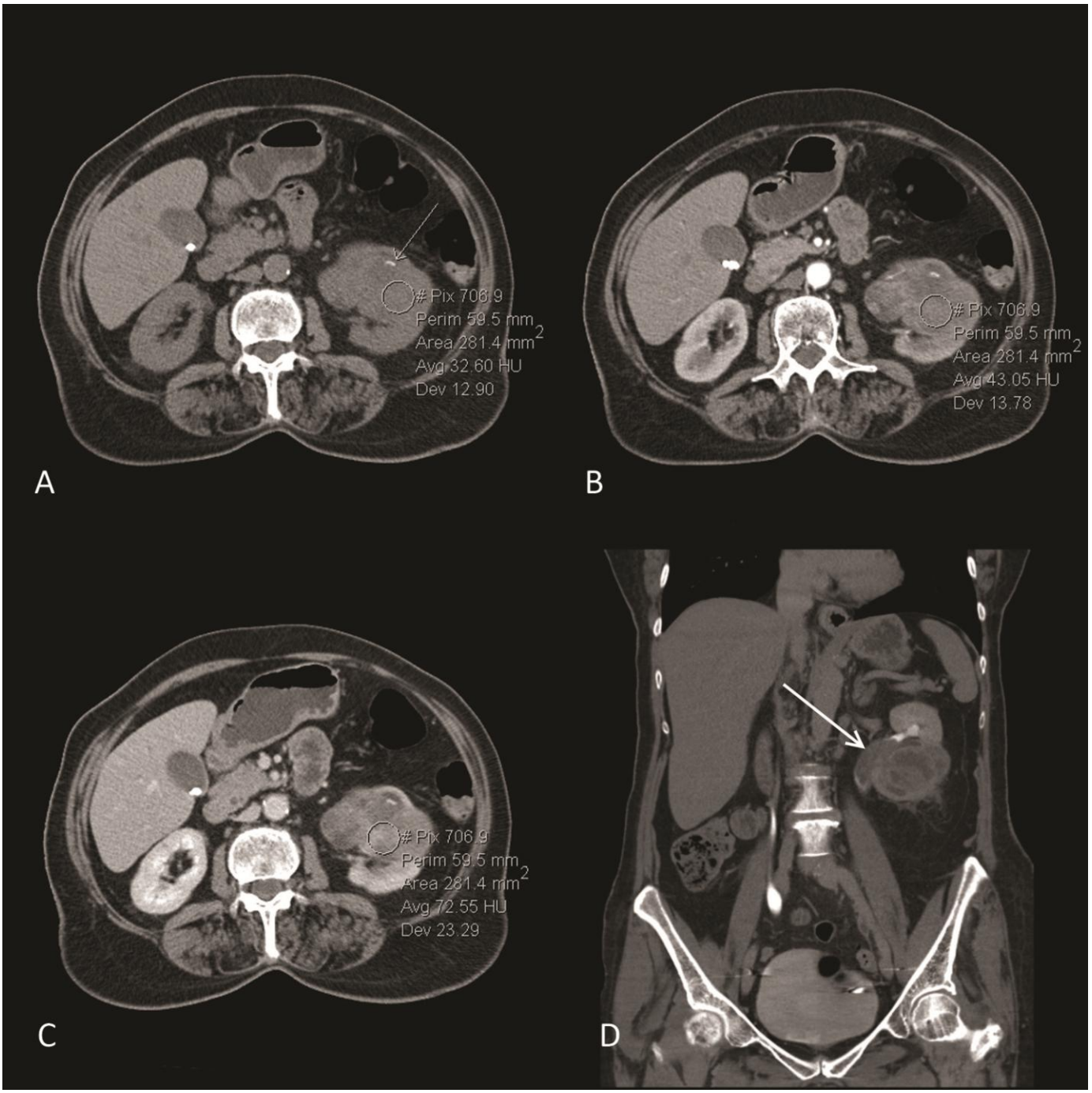

Figure 1: 87 year old woman diagnosed with sarcomatoid renal cell carcinoma. Axial and coronal computed tomographic images of the abdomen demonstrating a 6.0 x 5.2 x 5.5 cm mass centered within the inferior pole of the left kidney. On the axial noncontrast image (A), the mass is heterogeneous with the peripheral portions being isodense to the normal renal parenchyma and with central hypoattenuating areas suggestive of necrosis. The thin arrow denotes the calcification. During the corticomedullary phase (B), the peripheral solid component of the mass enhances but not as avidly as normal renal cortex. The central low density necrotic areas become more conspicuous during the nephrographic phase (C). Intraluminal soft tissue density is suggested within the proximal left ureter (arrow) on the coronal excretory phase image (D), which was confirmed to not represent tumor extension at the time of pathologic analysis. (Protocol: 64 slice, Siemens, 100 mAs, 120 kV, 3 mm slice thickness. W:400, L:40. 100 mL intravenous Omnipaque).

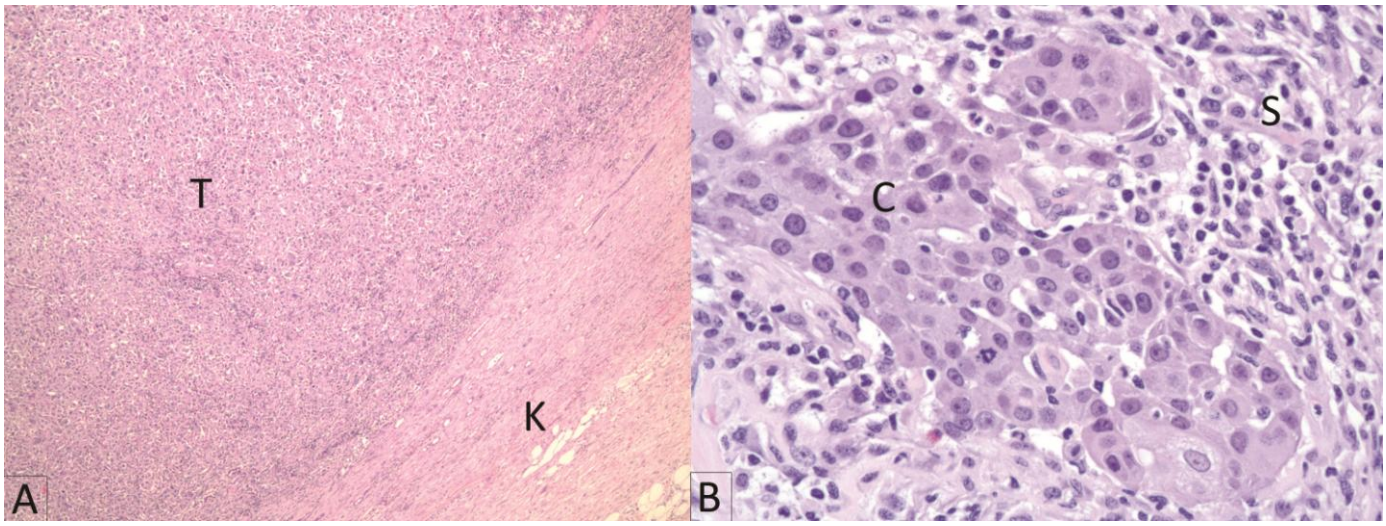

Figure 2: Hematoxylin and eosin-stained sections. Low power (4X) view (A) shows the interface between tumor (T) and uninvolved kidney (K). High power (40X) view (B) reveals cells with pale eosinophilia centrally (C) and a few perinuclear halos indicative of the chromophobe subtype of renal cell carcinoma (RCC) as well as pleomorphic spindled cells with large, bizarre nuclei (S) typical of sarcomatoid differentiation.

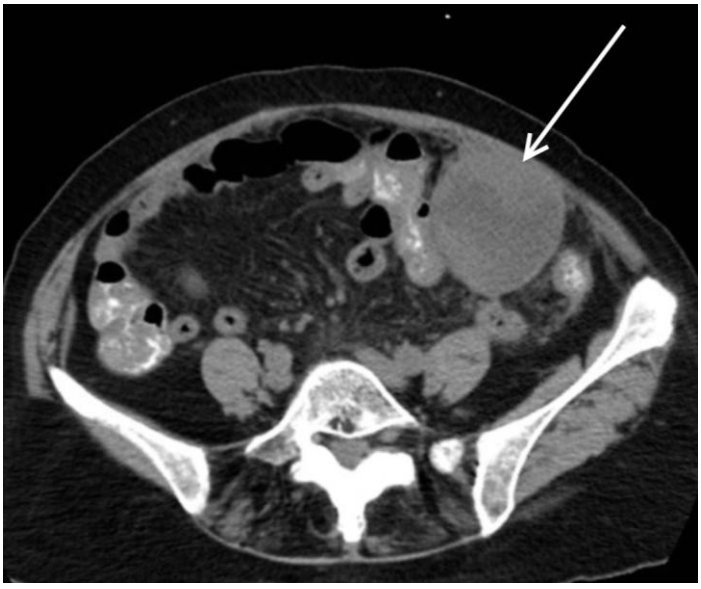

Figure 3 (left): 87 year old woman diagnosed with sarcomatoid renal cell carcinoma. Axial CT image of the abdomen obtained 5 months after left nephrectomy demonstrates a new round heterogeneous mass within the left hemiabdomen, abutting the transverse colon (arrow). (Protocol: 64 slice, Siemens, 100 mAs, 120 kV, 5 mm slice thickness. W:400, L:40. 100 mL intravenous Omnipaque).

Etiology	Renal cell carcinoma occurs in both sporadic (non-hereditary) and hereditary forms
Incidence	Sarcomatoid component reported to occur in 1-8% of renal cell carcinomas
Gender ratio	Renal cell carcinoma is slightly more common in men, 1.6:1 male:female ratio
Age predilection	No age predilection, renal cell carcinoma is most common in patients older than 50
Risk factors	Obesity and cigarette smoking are strongest associated risk factors for renal cell carcinoma
Treatment	Nephrectomy is primary therapy for renal cell carcinoma. With sarcomatoid component, use of chemotherapy agents has shown success in prolonging patient survival
Prognosis	Very poor prognosis for sarcomatoid renal cell carcinoma with median survival less than 1 year
Imaging features	Sarcomatoid renal cell carcinoma typically demonstrates aggressive features, such as necrosis, renal vein invasion and renal sinus invasion

Table 1: Summary table for sarcomatoid renal cell carcinoma

	CT	MRI	Pattern of contrast enhancement
Sarcomatoid renal cell carcinoma	- Low density areas represent necrosis - Renal sinus or perirenal fat invasion is common	Necrosis is evident as hypointense signal on T1 with corresponding T2 hyperintensity	Heterogeneous appearance due to non-enhancing necrotic areas
Clear cell renal cell carcinoma	Heterogeneous density due to presence of hemorrhage, necrosis or cysts	- Hypo to isointense on T1 - Iso to hyperintense on T2 - May show drop in signal on in phase/out of phase sequence due to microscopic fat	Hyperenhancement of mass compared to normal renal cortex due to tumor hypervascularity
Papillary renal cell carcinoma	Homogeneous in appearance compared to clear cell renal cell carcinoma; cystic change is common	- Hypo to isointense on T1 - Commonly hypointense on T2	Hypovascular and enhances to a lesser extent than clear cell variety
Chromophobe renal cell carcinoma	Mass with homogeneous soft tissue density	- Hypo to isointense on T1 - May have hypointense signal on T2	- Relative homogeneous enhancement - Enhances to greater extent than papillary renal cell carcinoma but not as robustly as clear cell

Table 2: Differential imaging characteristics for different renal cell carcinoma histological subtypes

ABBREVIATIONS

CT = Computed tomography
MRI = Magnetic resonance imaging
RCC = Renal cell carcinoma
WHO = World health organization

KEYWORDS

renal cell carcinoma; sarcomatoid dedifferentiation