

ORIGINAL ARTICLE

Mucinous tubular and spindle cell carcinoma of the kidney: imaging features

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Abstract

This article describes the features on sonography, computed tomography (CT) and magnetic resonance imaging (MRI) of mucinous tubular and spindle cell carcinoma of the kidney. Six pathologically proven cases of mucinous tubular and spindle cell carcinoma of the kidney were identified (5 females, 1 male); all patients underwent preoperative imaging. The mean age of the patients was 58.5 years. Thirteen imaging studies were available for review: 2 sonograms, 1 unenhanced CT scan, 5 contrast-enhanced CT scans, 1 unenhanced magnetic resonance imaging (MRI) examination, and 4 contrast-enhanced MRI examinations. Two abdominal radiologists evaluated all images retrospectively on a PACS workstation using a standardized data collection sheet until consensus was reached. All mucinous tubular and spindle cell carcinomas presented as well-marginated, small (mean 2.6 cm, range 1.9–3.2 cm) predominantly solid masses. No intratumoral fat or calcification was identified. Unenhanced CT and MRI appearances were variable as was the degree of enhancement following intravenous contrast material administration. There was no evidence of perinephric extension, renal vein involvement or metastatic disease in any of the cases. The radiological appearance of mucinous tubular and spindle cell carcinoma is diverse and therefore indistinguishable from the more common subtypes of renal cell carcinoma.

Keywords: Mucinous tubular and spindle cell carcinoma; kidney; sonography; computed tomography; magnetic resonance imaging.

Introduction

Mucinous tubular and spindle cell carcinoma of the kidney is a rare, low-grade epithelial neoplasm that has a characteristic histologic appearance that differentiates it from other subtypes of renal cell carcinoma^[1,2]. It is typically composed of 3 elements: tightly packed tubules, mucinous stroma, and spindle cells^[1,2]. The tumor has only recently been recognized as a distinct entity in the 2004 World Health Organization classification of adult renal tumors^[1,2]. Prior to this, however, multiple renal tumors with similar or overlapping histologic features were assigned a variety of other names^[3–7]. These included low-grade mucinous tubulocystic renal carcinoma^[3], unusual renal cell carcinoma with prominent spindle cell change^[4], low-grade myxoid renal epithelial

neoplasm^[5], low-grade tubular-mucinous renal neoplasm^[6] and spindle and cuboidal renal cell carcinoma^[7].

Mucinous tubular and spindle cell carcinomas are believed to arise from the distal nephron segments, in particular the collecting duct or the loop of Henle^[5,8]. There is a female preponderance (4:1) with a wide age range reported^[9]. This subtype of renal carcinoma is believed to portend a better prognosis than conventional renal cell carcinoma due to the presence of less cellular atypia, making the correct classification of clinical relevance^[5,10]. Mucinous tubular and spindle cell carcinoma is typically confined to the kidney, however, rare cases of local recurrence [6] and metastases to lymph nodes, bone and lung^[4,7,8,11,12] have been reported.

Although multiple small case series of this entity exist in the pathology literature^[13–17], there are only a few

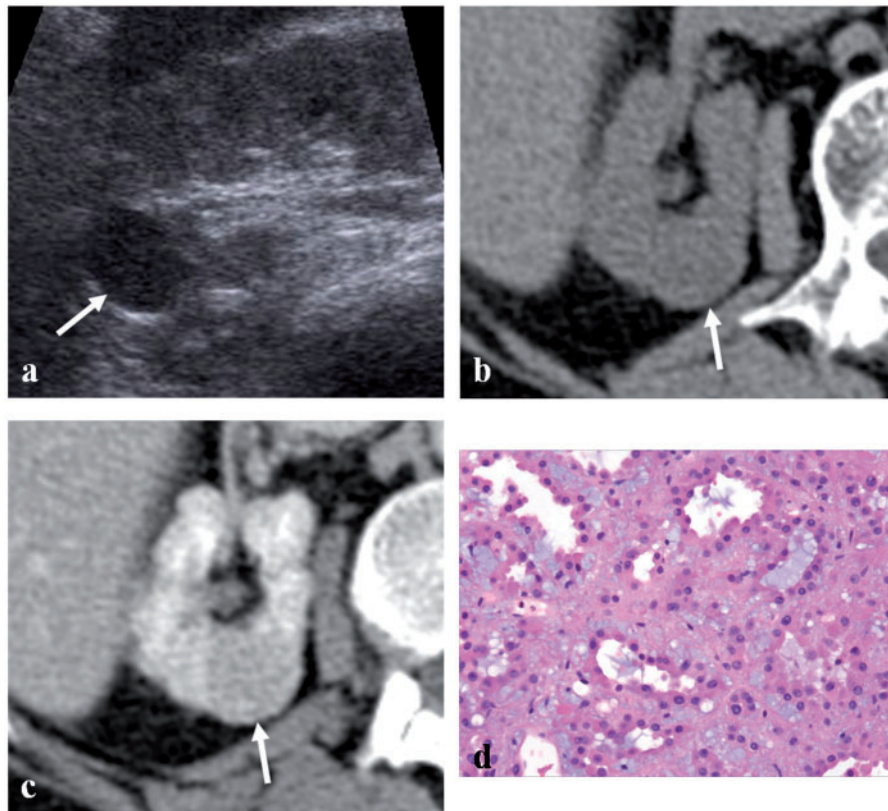


Figure 1 A 65-year-old woman with mucinous tubular and spindle cell carcinoma of right kidney. (a) Sagittal sonogram shows a well-margined hypoechoic mass (arrow). (b,c) Axial CT scans during unenhanced (b) and nephrographic (c) phases demonstrate a 1.9-cm intraparenchymal enhancing mass (arrows). (d) An 18-gauge biopsy specimen demonstrates oncocytic epithelioid cells with a tubular architecture, associated with a prominent myxoid matrix.

isolated case reports describing the imaging appearance^[18–20]; 1 case series demonstrated the computed tomography (CT) appearance in 2 patients^[21]. Therefore, the objective of this study was to present the radiological findings from a series of mucinous tubular and spindle cell carcinomas of the kidney using sonography, CT and magnetic resonance imaging (MRI).

Methods

After obtaining Institutional Review Board approval, a retrospective review of the electronic surgical pathology database at our institution was performed from 2004 to 2010. Informed consent was waived. Six pathologically proven cases of mucinous tubular and spindle cell carcinoma of the kidney were identified; all patients underwent preoperative imaging. The mean age of the patients (5 females, 1 male) with mucinous tubular and spindle cell carcinoma was 58.5 years (range 38–69 years). Thirteen imaging studies were available for review: 2 sonograms, 1 unenhanced CT scan, 5 contrast-enhanced CT scans, 1 unenhanced MRI examination, and 4 contrast-enhanced MRI examinations.

Two abdominal radiologists (SGS and VAS) evaluated all images retrospectively on a picture archiving and

communication system workstation using a standardized data collection sheet until consensus was reached. The maximum diameter of the mass was determined in 3 orthogonal planes. Location was evaluated for side (left or right), renal pole (upper, interpolar or lower) and whether the tumor was primarily exophytic, intraparenchymal or central, and whether it extended into the renal sinus. The imaging characteristics of the tumor reviewed included margin (poorly or well-margined), extension into the perinephric space, involvement of the ipsilateral renal vein and the presence of calcification, macroscopic fat or cystic change within the tumor. The remainder of the kidneys were reviewed for other focal lesions. The presence of metastases was also evaluated.

Predominant sonographic echogenicity compared with renal cortex (hyper-, iso or hypoechoic) and MRI signal characteristics of the mass on T1-weighted and T2-weighted sequences compared with renal cortex were evaluated. CT attenuation (HU) or MRI signal intensity values (SI units) were obtained using regions of interest (ROI) of at least 3 pixels, on unenhanced, nephrographic and excretory phases. Masses were considered enhancing if after contrast medium, there was an increase in attenuation of at least 20 HU^[22], or a 20% increase in signal intensity^[23]. Enhancement was deemed equivocal if the

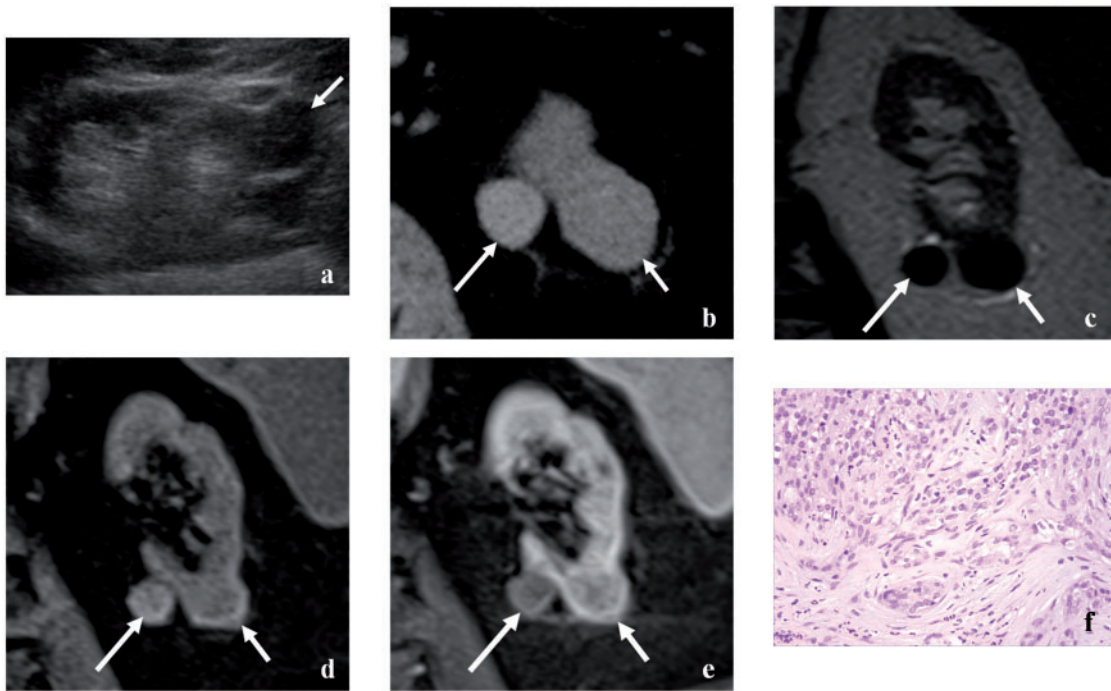


Figure 2 A 67-year-old man with mucinous tubular and spindle cell carcinoma of left kidney. (a) Sagittal sonogram shows a well-margined hypoechoic mass (arrow) arising from the lower pole. (b) Axial CT scans during the unenhanced phase demonstrates a 3.2-cm exophytic hyperdense mass (short arrow), and a nearby hyperdense cyst (long arrow). (c) Coronal T2-weighted MR image demonstrates T2-hypointensity of the mass (short arrow). The adjacent cyst (long arrow) is also T2-hypointense in keeping with hemorrhage. (d,e) Coronal T1-weighted fat-suppressed sequence in unenhanced (d) and nephrographic (e) phases shows enhancement of the mass (short arrow) and no enhancement of the cyst (long arrow). (f) An 18-gauge biopsy specimen demonstrates a mixture of epithelioid and bland spindle cells associated with a desmoplastic matrix; confirmatory cytogenetics helped make the correct diagnosis in this case.

increase in CT attenuation was between 10 and 20 HU, and if the increase in signal intensity was between 15% and 20%.

Results

All mucinous tubular and spindle cell carcinomas appeared as well-margined, small (mean 2.6 cm, range 1.9–3.2 cm) predominantly solid masses without calcification or fat (Figs. 1 and 2). Five masses demonstrated enhancement after intravenous contrast material administration (one mass demonstrated equivocal enhancement on CT but enhanced unequivocally on MRI). No contrast-enhanced imaging was available in 1 case. A small non-enhancing cystic component was found in association with 1 mass. This may have represented a simple cystic component of the lesion or an adjacent cyst. All masses were confined to the kidney and were found to be predominantly intraparenchymal in location in 5 cases and exophytic in 1 case. No mass involved the renal pelvis. No synchronous renal neoplasms were seen in any case. There was no evidence of perinephric extension, renal vein involvement or metastatic disease in any of the cases. An enlarged left paraaortic lymph node

measuring 2.4×1.5 cm was present in 1 case. This was shown subsequently to be stable for 28 months and likely reactive to the patient's known ulcerative colitis. In the 2 patients who underwent ultrasonography, both masses appeared homogeneously hypoechoic and well margined. In 5 masses where unenhanced CT was available, 3 tumors were isodense to renal parenchyma and 2 were hyperdense. The T1 signal was isointense to renal parenchyma in all 5 cases where MRI scans were available for review. The T2 signal, however, demonstrated hypo-, iso- and hyperintensity in 2, 1 and 2 cases, respectively (Table 1).

Discussion

Mucinous tubular and spindle cell carcinoma is a rare tumor of the kidney that has only recently been described. Although reported in the pathology literature^[13–17], limited information exists regarding its imaging appearance. To the best of our knowledge, no prior studies have evaluated the multimodality imaging appearance of this neoplasm.

In summary, our review of the imaging of 6 patients found all tumors to be well-margined and confined to

Table 1 Imaging features of mucinous tubular and spindle cell carcinoma

Variable	Patient					
	1	2	3	4	5	6
Age (years)	65	47	38	67	65	69
Gender	Female	Female	Female	Male	Female	Female
Maximum size (cm)	1.9	2.8	2.4	3.2	2.1	3.1
Side	Right	Left	Right	Left	Left	Left
Predominant location	Parenchyma	Parenchyma	Parenchyma	Exophytic	Parenchyma	Parenchyma
Cystic component	No	No	Yes	No	No	No
Sonographic echogenicity relative to renal parenchyma	Hypoechoic	N/A	N/A	Hypoechoic	N/A	N/A
Unenhanced CT density relative to renal parenchyma	Isodense	N/A	Isodense	Hyperdense	Hyperdense	Isodense
CT enhancement in nephrographic phase	76 HU	N/A	86 HU	13 HU	34 HU	N/A
T1 signal relative to renal parenchyma	Isointense	N/A	Isointense	Isointense	Isointense	Isointense
T2 signal relative to renal parenchyma	Isointense	N/A	Hyperintense	Hypointense	Hyperintense	Hypointense
MRI enhancement in nephrographic phase (%)	N/A	N/A	107	63	151	116

CT enhancement, attenuation of mass after intravenous contrast material administration minus unenhanced attenuation; MR enhancement, signal intensity of mass after intravenous contrast administration minus unenhanced signal intensity divided by unenhanced signal intensity; N/A, not available.

the kidney without metastatic spread. All tumors were predominantly solid and the 5 cases in which intravenous contrast was administered demonstrated enhancement.

Comparison of our imaging findings with prior reports is difficult due to limited information available in published articles. Prior to 2004, the entity was known by a variety of names. These included low-grade mucinous tubulocystic renal carcinoma^[3], unusual renal cell carcinoma with prominent spindle cell change^[4], low-grade myxoid renal epithelial neoplasm^[5], low-grade tubular-mucinous renal neoplasm^[6] and spindle and cuboidal renal cell carcinoma^[7]. MacLennan et al.^[3] described the sonographic and/or CT appearance of 11 of the 12 tumors in this series. Eight were considered solid, 2 purely cystic and 1 was a complex solid/cystic mass. No further feature analysis was performed. Moreover, subsequent review of the pathology of these tumors revealed that only 5 cases in the series were mucinous tubular and spindle cell carcinoma and retrospective identification of these cases with regard to imaging characteristics was not possible^[24].

Yusukuku et al.^[18] reported the imaging appearance of mucinous tubular and spindle cell carcinoma in a 71-year-old woman. A CT scan showed a well-defined 5.6 × 5.2 cm mass arising from the left kidney. It showed no enhancement on early phase scans after intravenous contrast material administration and slightly enhanced on a later phase. The timing of imaging relative to the contrast material administration and the degree of enhancement were not quantified. The lesion was isointense on both T1- and T2-weighted MR imaging.

Geamizadeh et al.^[19] and Gaafar et al.^[21] also presented select CT images of mucinous tubular and spindle cell carcinoma. Again no feature analysis of the imaging appearances was provided. All 3 cases showed a well-defined hypodense mass located centrally within the renal parenchyma.

Noon et al.^[20] recently reported the MRI findings of an incidentally detected mucinous tubular and spindle cell carcinoma in a 35-year-old woman presenting with an ectopic pregnancy. The mass was isointense to renal parenchyma on T1-weighted imaging and intermediate signal intensity on T2-weighted imaging. It demonstrated mild early phase enhancement. The authors highlighted that the relatively low T2 signal and mild contrast enhancement are similar to papillary renal cell carcinoma. Similarly, 2 cases in our series demonstrated low T2 signal and 1 of these enhanced only mildly. The low T2 signal of papillary carcinoma is well documented^[25–27] and shown to be due to papillary architecture with some contribution from the presence of hemosiderin in some cases^[25]. Papillary renal cell carcinoma has also been shown to enhance less than clear cell renal cell carcinoma^[28].

Based on our imaging finding and those in the literature, it appears that mucinous tubular and spindle cell carcinoma may have a variable imaging appearance. Although some features such as low signal on T2-weighted MRI or lack of metastatic spread may suggest the diagnosis, making a prospective diagnosis by imaging alone would be difficult.

Mucinous tubular and spindle cell carcinoma is considered a low-grade carcinoma with a favorable prognosis and a low propensity for local recurrence or metastasis^[29]. Given this natural history, it would be an ideal candidate for nephron-sparing treatment such as partial nephrectomy or percutaneous ablation. This would require prospective knowledge of the pathology via percutaneous biopsy. Percutaneous biopsy should also be considered when an enhancing mass is hyperattenuating on unenhanced CT and hypointense on T2-weighted MRI sequences^[30–32] as seen in case 4 of our series. This is because benign neoplasms, in particular angiomyolipoma with minimal fat, may have an identical

imaging appearance^[33]. Caution should be applied, however, when interpreting biopsy results as sarcomatoid differentiation can be confused with the banal-appearing spindle cells of mucinous tubular and spindle cell carcinoma. In addition, sarcomatoid differentiation has been described in rare cases of patients with mucinous tubular and spindle cell carcinoma^[11,12,34] leading to a more aggressive behavior with metastases to bone and lung. Sarcomatoid differentiation may only affect part of the lesion and therefore could be missed by percutaneous biopsy. These reported cases, however, were much larger than the tumors in our series (size range was 7 to 15 cm) and tumors with sarcomatoid differentiation are frequently associated with necrosis.

A limitation of our study was its retrospective nature. As a result, data from all 3 imaging modalities were not available for each patient. In addition, because this entity is rare, only a small number of cases were available for review. However, to the best of our knowledge, our study is the largest published radiological review of mucinous tubular and spindle cell carcinoma of the kidney.

In summary, mucinous tubular and spindle cell carcinoma of the kidney appears as a well-marginated, solid enhancing renal mass. The sonographic, CT and MRI features appear to be too diverse to allow a specific diagnosis to be rendered on the basis of imaging features alone. Mucinous tubular and spindle cell carcinoma is therefore indistinguishable from the more common subtypes of renal cell carcinoma.

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