CASE REPORT

CT and MRI appearance of solitary parapelvic neurofibroma of the kidney

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ABSTRACT. We report an extremely rare case of a renal parapelvic neurofibroma. The mass was an incidental finding on an abdominal CT scan of an asymptomatic 59-year-old male with hepatitis C. Subsequent MRI demonstrated a complex mass occupying the left renal sinus that had mass effect on the pelvicalyceal system. The patient underwent left radical nephrectomy, and histological and immunohistochemical findings confirmed the lesion to be a neurofibroma. Given that similar imaging features have been reported previously in four additional cases, we suggest that it may be possible to include this entity in the pre-operative differential diagnosis.

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Solitary neurofibromas occur in patients without neurofibromatosis-1. Solitary neurofibromas of the kidney are exceedingly rare, constituting a very small percentage of urinary tract tumours [1, 2]. To our knowledge, only four cases of solitary renal parapelvic or parenchymal neurofibromas have been reported in the literature [1–4]. In this report, we present an additional case of a solitary renal parapelvic neurofibroma, and we review the radiological, gross histological and immunohistochemical features.

Case report

Radiological findings

An asymptomatic 59-year-old male with a medical history of hepatitis C, diabetes mellitus and hypertension was found, on screening by abdominal computed tomography (CT), to have a 7.4 × 6.2 cm well-circumscribed mass (Figure 1) occupying the left renal sinus. The mass was associated with deformity and compression of the pelvicalyceal system. Internally, the mass demonstrated heterogeneous density, ranging from about 30 to 50 Hounsfield units, incompatible with a simple cyst. MRI, which was performed for further characterisation, showed an 8.3 × 5.3 cm T_2 hyperintense mass (Figure 2) centred within and expanding the left renal sinus. The mass extended superiorly from the renal sinus along the medial aspect of the left kidney. The mass caused some mild compression of the left renal collecting system, but the left kidney excreted normally. On MRI, the mass showed internal complexity: it contained widespread enhancing septations with areas

of thickening and nodularity (Figure 3). There was no associated invasion of the renal parenchyma, vascular invasion or lymphadenopathy. After imaging, the mass remained non-specific and a malignancy could not be excluded. Diagnostic considerations included tumours known to originate within the renal sinus, such as haemangioma or haemangiopericytoma, and an unusual appearance of renal cell carcinoma or multilocular cystic nephroma, although there was no clear renal parenchymal involvement to suggest these latter diagnoses strongly. No other renal mass was identified.

Pathological findings

The patient underwent left radical nephrectomy. The tumour was located within the renal hilum, separate from the kidney (Figure 4). The kidney itself was uninvolved and grossly unremarkable aside from pelvicalyceal dilatation. Microscopic examination of the tumour revealed interlacing bundles of spindle cells with wavy nuclei (Figure 5). The cells were associated with a fine, wiry collagenous background. Mast cells and lymphocytes were dispersed throughout the stroma. No areas suggestive of Antoni A cells, typical of a schwannoma, were seen. Mitotic figures were rare and no atypia was noted. The neoplastic cells were positive for S-100 and negative for vimentin and HMB-45. On the basis of these histological and immunohistochemical findings, the tumour was diagnosed as a neurofibroma.

Discussion

Solitary neurofibromas are rare and usually found within cutaneous or subcutaneous tissues or along nerves [2]. Their occurrence in the urinary tract is

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Figure 1. Axial contrast-enhanced computed tomography (CT) shows a mass (arrow) expanding the left renal sinus, with associated compression of the pelvicalyceal system.

Figure 2. Coronal half-Fourier acquisition single-shot turbo spin-echo (HASTE) magnetic resonance (MR) image shows a complex T_2 hyperintense mass (arrow) occupying the left renal sinus and extending superiorly along the medial aspect of the left kidney, without invasion of the renal parenchyma.

Figure 3. Axial contrast-enhanced T_1 weighted magnetic resonance (MR) image shows a left renal sinus mass (arrow) with internal enhancing components and compression of the pelvicalyceal system. As seen on CT, the left kidney has maintained excretory function.

Figure 4. Cross-section of a specimen of kidney with the parapelvic tumour measuring $10 \times 7 \times 5$ cm. The cut surface shows a firm, yellow, fatty, well-circumscribed mass (arrow). No areas of haemorrhage or necrosis were noted.

particularly uncommon; they are more likely to occur within the retroperitoneal soft tissues rather than within the kidneys themselves [3]. The rate of malignant transformation of neurofibromas has been estimated at 2–29% in patients with neurofibromatosis-1, but the rate of malignant degeneration of solitary neurofibromas is believed to be extraordinarily low [5]. In addition, solitary neurofibromas are generally associated with a low local recurrence rate if completely excised [6].

The case we describe involved a solitary parapelvic neurofibroma located at the hilum of the kidney, without attachment to the pelvicalyceal system or renal parenchyma. The occurrence of a solitary neurofibroma within the urinary tract is incredibly rare [3], with only four such tumours previously reported in the literature to our knowledge (summarised in Table 1). All of these tumours were located at the renal hilum and produced mass effect on the pelvicalyceal system. Despite their similar anatomical location, the clinical presentations of the tumours differed. Two cases presented with lumbar

Figure 5. Section from the tumour demonstrating spindle cells with wavy nuclei in a collagenous stroma (haematoxylin-eosin, original magnification $\times 100$).

Table 1. Summary of previously reported cases and current case of solitary parapelvic neurofibroma

Case	Age	Sex	Location	Size	Available imaging	Treatment
Freund et al, 1967 [3]	45	F	Inferomedial left kidney, involving lower pole calyces and a portion of renal pelvis	10.2×8.5×4.5 cm	Excretory urogram, conventional angiography	Local tumour resection
Borrego et al, 1995 [4]	41	NR ^a	Left renal sinus, with mass effect on upper pole calyces	7.5×6.0 cm	CT, US, excretory urogram	Complete nephrectomy
Nishiyama et al, 2000 [1]	33	F	Right renal sinus, with mass effect on upper renal pelvis	4.0×2.0×1.0 cm	CT	Retroperitoneoscopic resection of tumour
Kostakopoulos, et al 2003 [2]	37	F	Right renal sinus, with mass effect on renal pelvis	7.0 cm	CT	Radical nephrectomy
Present case	59	M	Left renal sinus, with mass effect on renal pelvis	10.0×7.0×5.0 cm	CT, MRI	Radical nephrectomy

^aNR, not reported.

pain [1, 2], one case was detected during the investigation of long-standing renal disease [3], and both of the remaining previously reported cases [4] as well as our case were found incidentally in asymptomatic patients. The renal parenchyma was completely uninvolved in all five cases, excluding the diagnosis of a tumour of renal cell origin.

Our report is the first to show the MRI appearance of a renal parapelvic neurofibroma. Specifically, MRI demonstrated a circumscribed, internally complex mass expanding and growing out of the renal sinus. It caused compression of the collecting system without invasion. Review of the CT images from three earlier reports [1, 2, 4] of this lesion show a similar imaging appearance, although in one case [1] the mass was smaller in size without extension out of the renal sinus. The described imaging appearance suggested a renal sinus tumour of primary mesenchymal origin, but the findings were non-specific; it remains a challenge to distinguish this benign tumour from a malignancy pre-operatively [7].

Given the non-specific imaging findings, resection and histopathological analysis remains necessary to establish the diagnosis. Although radical nephrectomy [2] and a nephron-sparing approach by way of retroperitoneoscopy [1] have been reported in the literature, none of the reported cases had any renal parenchymal involvement. Intervention is necessary, but, given the similar imaging

features reported in this case and previous cases, it may be possible to include benign mesenchymal neoplasms, such as a neurofibroma, in the differential diagnosis. When such a diagnosis is suspected, a frozen section analysis may be considered at the time of surgery to establish both the benign nature of the tumour and the absence of renal parenchymal involvement, thereby assisting the avoidance of unnecessary radical nephrectomy.

References

1. Nishiyama T, Ikarashi T, Terunuma H. Parapelvic neurofibroma of the kidney. *Int J Urol* 2000;7:470–1.
2. Kostakopoulos A, Chorti M, Protogerou V, Kokkinou S. Solitary neurofibroma of kidney: clinical, histological and chromosomal appearance. *Int Urol Nephrol* 2003;35:11–13.
3. Freund ME, Crocker DW, Harrison JH. Neurofibroma arising in a solitary kidney. *J Urol* 1967;98:318–21.
4. Borrego J, Cuesta C, Allona A, Navio S, Escudero A. Myxoid neurofibroma of the renal sinus. *Actas Urol Esp* 1995;19:415–18.
5. Enzinger FM, Weiss SW. *Soft tissue tumors*. Philadelphia: Mosby-Elsevier, 2008.
6. Jeyaretna DS, Oriolowo A, Smith MEF, Watkins RM. Solitary neurofibroma of the male breast. *World J Surg Oncol* 2007;5:23.
7. Rha SE, Byun JY, Jung SE, Oh SN, Choi YJ, Lee A, et al. The renal sinus: pathologic spectrum and multimodality imaging approach. *Radiographics* 2004;24:S117–31.