CASE REPORT

Parapelvic solitary neurofibroma of the kidney

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SUMMARY

A middle-aged man presented with intermittent gross haematuria and dull aching left flank pain for 1 month. He was a chronic smoker for 15 years. Contrast-enhanced CT scan of the abdomen detected a 4.5×3.0×2.5 cm, heterogeneous and poorly contrast-enhancing mass in the left renal sinus and upper ureteric region causing narrowing of the pelviureteric junction and upper ureter. Considering it to be a transitional cell carcinoma of upper urinary tract, the patient underwent laparoscopic radical nephroureterectomy. Histopathology revealed the mass to be a neurofibroma in the renal sinus extending around the upper ureter.

BACKGROUND

Neurofibroma is a benign tumour arising from nerve sheaths. A renal or parapelvic solitary neurofibroma that occurs de novo without any genetic mutation/syndrome is quite rare and only seven cases have been reported in the literature so far. Preoperative diagnosis is usually difficult as the tumour may closely mimic transitional cell carcinoma of the pelvis or renal cell carcinoma. We report a case of a neurofibroma in the region of the renal pelvis and upper ureter, masquerading as transitional cell carcinoma of the upper urinary tract. The purpose of this case report is to highlight the fact that a neurofibroma can occur in highly unusual locations such as the renal sinus where it can be misdiagnosed clinically and radiologically as transitional cell carcinoma of the upper tract. Awareness of this rare differential diagnosis of tumours involving the renal sinus or pelvis is important.

CASE PRESENTATION

A 47-year-old man presented with intermittent gross haematuria and dull aching left flank pain for 1 month. There was no history of other urinary symptoms, anorexia or significant weight loss. The patient was a chronic smoker for 15 years. His medical history, abdominal examination and systemic examinations were unremarkable.

INVESTIGATIONS

The patient's haemoglobin and serum creatinine were 12g/dl and 0.8 mg/dl, respectively. Urine microscopy revealed 6–8 red blood cells per high-power field. Complete blood counts, coagulation profile, serum biochemistries including liver function tests, chest X-ray, urine cytology for malignant cells and urine culture were unremarkable. Ultrasonography scan detected a mass with hydronephrosis in the left renal pelvis. Contrast-enhanced CT scan of the abdomen revealed a mass of about 4.5×3.0×2.5 cm in the left renal pelvis region extending to upper ureter and causing irregular narrowing of the pelviureteric junction and upper ureter (figure 1A,B). The tumour was heterogeneous, poorly enhancing on contrast administration and suggestive of transitional cell carcinoma of the upper urinary tract. There was also a small 5 mm stone at the pelviureteral junction. The right kidney was normal in size, shape and contrast excretion.

DIFFERENTIAL DIAGNOSIS

- ▶ Transitional cell carcinoma of renal pelvis.
- ▶ Renal cell carcinoma.
- ▶ Mesenchymal tumours: leiomyosarcoma, fibrosarcoma, liposarcoma, haemangiopericytoma, malignant fibrous histiocytoma, haemangioma,

Figure 1 (A and B) contrast-enhanced CT scan showing a mass in right renal pelvic and upper ureteric region causing narrowing of pelviureteric junction and upper ureter.

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Figure 2 (A) specimen showing tumour in parapelvic region; (B) cut-section of specimen showing myxoid tumour in renal sinus; (C) histopathology showing spindle cells loosely arranged in fascicles with neural elements (thin arrows) in the background of loose fibromyxoid stroma (thick arrow).

fibroma, leiomyoma, angiomyolipoma, neurogenic tumour, teratoma, etc.

TREATMENT

After discussing management options and counselling, the patient underwent laparoscopic left radical nephroureterectomy and bladder cuff excision considering it to be an invasive transitional cell carcinoma. Intraoperatively, renal vein, artery, psoas muscle and surrounding structures were free from tumour invasion. The specimen revealed a myxoid tumour located in the renal sinus and upper periureteric region, compressing the renal pelvis and upper ureter (figure 2A, B). Histopathological examination revealed hypocellular proliferation of spindle cells loosely arranged in fascicles and neural elements in the background of loose fibromyxoid stroma (figure 2C). Tumour cells

were positive for S-100 and CD-34 immunostaining. The tumour involved adventitia of the renal pelvis and ureter, sparing the lamina propria and urothelial lining.

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful. The patient is doing well at 3 months follow-up.

DISCUSSION

Neurofibroma of the parapelvic region or renal sinus is a very rare tumour. A literature search revealed only seven reported cases of this specific entity so far. The details of these cases are summarised in table 1.

The clinical presentation in the reported cases varied from asymptomatic incidental detection to lumbar pain and

Table 1 Summary of cases of parapelvic neurofibroma reported in the literature

Case report	Age	Gender	Presentation	Preoperative imaging	Size of tumour (cm)	Site of tumour	Surgery procedure
Freund <i>et al</i> ¹	45	F	Dull aching lumbar pain	Angiography, excretory urogram	10.2×8.5×4.5	Left lower pole calyces and renal pelvis	Local tumour excision
Borrego <i>et al</i> ²	41	NA	Asymptomatic	CT, US, excretory urogram	7.5×6.0	Left renal sinus	Nephrectomy
Nishiyama <i>et al</i> ³	33	F	Dull aching lumbar pain	CT	4.0×2.0×1.0	Right renal sinus	Retroperitoneoscopic tumour resection
Kostakopoulos <i>et al</i> ⁴	37	F	Right lumbar pain	CT	7.0	Right renal sinus	Nephrectomy
Eljack <i>et al</i> ⁵	59	M	Asymptomatic	CT, MRI	10.0×7.0×5.0	Left renal sinus	Nephrectomy
Mondal <i>et al</i> ⁶	54	F	Right lower back pain and haematuria	CT, MRI	4×3.6	Right upper pole calyces	Nephrectomy
Corbellini <i>et al</i> ⁷	47	M	Right lumbar pain and fever	US, CT	5.3×4.4×6.0	Right retroperitoneal para-aortic-pararenal	Local tumour excision
Our case	47	M	Left flank Pain, intermittent haematuria	US, CT	4.5×3.0×2.5	Left renal sinus, upper ureter	Laparoscopic nephroureterectomy +bladder cuff excision

NA, not available; US, ultrasonography.

haematuria. Four of the reported tumours involved the renal sinus while two involved calyces and renal parenchyma. In the case report by Corbellini *et al.*,⁷ the tumour was a retroperitoneal neurofibroma in the para-aortic region situated away from the renal sinus. The diagnosis of neurofibroma was not suspected in the majority of these lesions (4 of 7) due to close proximity with or involvement of the renal pelvis or kidney, and they were treated with radical nephrectomy. Only when the tumours are distinguished from the renal pelvis, could they be locally excised.^{1–3}

Other mesenchymal tumours, renal lymphoma and metastasis to renal sinus are some rare differential diagnoses for such renal sinus tumours. Image findings of the mesenchymal tumours are usually non-specific. They are mostly misdiagnosed preoperatively as the more common renal cell carcinoma or transitional cell carcinoma.

In the published literature, ours is the first case of parapelvic solitary neurofibroma where the tumour also encased the upper ureter to cause ureteral narrowing on imaging. Inability of the CT scan to differentiate it from upper urinary tract transitional cell carcinoma and presence of haematuria biased the diagnosis towards transitional cell carcinoma. Retrospectively, the stone seems to be responsible for the haematuria in our case as the tumour was not invading the pelvis/ureteral muscle or urothelium. Because of infiltration of the adventitia of the renal pelvis and upper ureter by the tumour, renal salvage would have been difficult in our case.

Learning points

- An isolated solitary neurofibroma can occur at highly unusual locations such as the renal sinus, or parapelvic, para-aortic or paraureteral region.
- A parapelvic neurofibroma may mimic transitional cell carcinoma of renal pelvis on imaging and clinically.
- For a parapelvic tumour distinguishable from that of the renal pelvis, one should have a high index of suspicion. Needle biopsy or surgical exploration with frozen section may be needed to determine the nature of the tumour to avoid unnecessary radical nephrectomy.

As renal neurofibroma is a benign tumour, conservative surgical treatment in the form of local tumour excision or partial nephrectomy is considered adequate treatment. However, the treatment of solitary neurofibroma may still be controversial, varying from radical nephrectomy⁴ to conservative management,⁸ depending on various factors such as size of the lesion, involvement of the pelvicalyceal system and kidney, symptomatic status and suspicion of malignancy.

In neurofibroma of the kidney or renal sinus, there are no preoperative pathognomonic imaging features that can differentiate it from other renal neoplasms—either benign or malignant, though perfusion imaging to differentiate it from malignant lesions of the kidney has been advocated.⁹ A high index of suspicion may be required for performing a preoperative needle biopsy or surgical exploration with frozen section, especially in cases where imaging identifies the mass to be in the parapelvic region, distinguishable from the renal pelvis.

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REFERENCES

- 1 Freund ME, Crocker DW, Harrison JH. Neurofibroma arising in a solitary kidney. *J Urol* 1967;98:318–21.
- 2 Borrego J, Cuesta C, Allona A, *et al.* Myxoid neurofibroma of the renal sinus. *Actas Urol Esp* 1995;19:415–18.
- 3 Nishiyama T, Ikarashi T, Terunuma H. Parapelvic neurofibroma of the kidney. *Int J Urol* 2000;7:470–1.
- 4 Kostakopoulos A, Chorti M, Protogerou V, *et al.* Solitary neurofibroma of kidney: clinical, histological and chromosomal appearance. *Int Urol Nephrol* 2003;35:11–13.
- 5 Eljack S, Rosenkrantz AB, Das K. CT and MRI appearance of solitary parapelvic neurofibroma of the kidney. *Br J Radiol* 2010;83:e108–10.
- 6 Mondal SK, Mallick MG, Bandyopadhyay R, *et al.* Neurofibroma of kidney: an uncommon neoplasm and diagnostic dilemma with solitary fibrous tumor. *J Can Res Ther* 2010;6:388–90.
- 7 Corbellini C, Vingiani A, Maffini F, *et al.* Retroperitoneal pararenal isolated neurofibroma: report of a case and review of literature. *Ecancermedicalscience* 2012;6:253.
- 8 Kubiena H, Entner T, Schmidt M, *et al.* Peripheral neural sheath tumors (PNST)—what a radiologist should know. *Eur J Radiol* 2013;82:51–5.
- 9 Liu WG, Liang WJ. Letter to the editor: proper imaging method for evaluation of solitary renal parapelvic neurofibroma. *Br J Radiol* 2011;84:771.

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