

Oncology

Renal myopericytoma: A case report and literature review

Thomas Riley^{a,*}, Patrick Shenjere^b, Arun Jain^a, Shyam Sunder^a^a North Manchester General Hospital, Pennine Acute NHS Hospitals Trust, Delaunay's Road, Manchester, M8 5RB, UK^b The Christie Hospital, Christie NHS Foundation Trust, Wilmslow Road, Manchester, M20 4BX, UK

ARTICLE INFO

Keywords:

Renal myopericytoma
Pericytic tumour
Rare
Case report
Diagnosis

ABSTRACT

Renal myopericytoma is an extremely rare entity with just 11 cases reported in the literature. We report the case of a 57 year old Caucasian man who was found to have a renal myopericytoma following nephrectomy for suspected renal cell carcinoma. Renal myopericytoma has a distinct morphological overlap with other pericytic tumours and significant histological variation has been noted between cases reported to date. Further characterising this novel tumour is vital to identify subtypes within this spectrum, understand its behaviour and to identify imaging trends which may lead to pre-operative diagnosis in order to potentially avoid radical treatment.

Introduction

Myopericytoma is a rare mesenchymal tumour originating from myopericytes.¹ They typically occur in the skin and soft tissues and are extremely rare in visceral organs, with only 11 cases previously reported in the kidney [Table 1]. Renal tumours tend to be larger than peripheral tumours (5.6cm vs < 2cm).^{2,3} All primary cases of renal myopericytoma have been benign.

The World Health Organisation recognised myopericytoma as a distinct entity in its classification of tumours of soft tissue in 2013. They are classified as pericytic tumours displaying differentiation towards perivascular myoid cells or myopericytes.⁴ Histologically, myopericytomas contain spindle-shaped myoid cells with bland, round or ovoid nuclei in a concentric perivascular arrangement.³ However, certain reports have shown variant morphology in renal involvement. This case report further characterises this rare entity.

Case report

We report the case of a 57 year old British man who presented to the emergency department with acute onset left flank pain, vomiting and an elevated white cell count. Prior to this the patient had been asymptomatic with no visible haematuria, other urinary symptoms or weight loss. He had no significant past medical history. Physical examination revealed mild tenderness in the left flank but was otherwise unremarkable. The ureteric stone passed spontaneously. Radical

nephrectomy was performed due to the suspicion of renal cell carcinoma (RCC). Histological analysis revealed a vascular tumour with intervening stromal tissue composed of spindle shaped cells leading to a diagnosis of renal myopericytoma. The patient remains alive and disease free at eight months following diagnosis.

Imaging findings

A computed tomography (CT) scan of the abdomen and pelvis revealed a 5mm obstructing left mid-ureteric stone with mild calyceal dilatation and an incidental 5.5 cm lesion in the inter-polar region of the left kidney [Image 1]. The lesion showed marked heterogeneity with internal high density followed by a hyperdense rim. There was no evidence of local invasion or metastasis. The ureter and remaining urinary tract were unremarkable. The radiological differential diagnosis was thought to be an angiomyolipoma or RCC with internal haemorrhage. Follow-up CT renal triple phase did not identify any features suggestive of the presence of adipose tissue or acute internal haemorrhage. There was progressive contrast enhancement starting centrally and moving peripherally between arterial and portal venous phases leaving a rim of reduced enhancement.

Pathological findings

Macroscopic analysis revealed a solid 33 × 33 × 35mm inter-polar solitary renal tumour with a well-defined pseudocapsule and greyish-

* Corresponding author.

E-mail addresses: Tom.Riley@doctors.org.uk (T. Riley), p.shenjere@nhs.net (P. Shenjere), Arun.Jain@pat.nhs.uk (A. Jain), Shyam.Sunder@pat.nhs.uk (S. Sunder).<https://doi.org/10.1016/j.eucr.2020.101537>

Received 23 November 2020; Accepted 14 December 2020

Available online 15 December 2020

2214-4420/© 2020 Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Table 1
Summary of all cases to date; adapted from Qiao et al.³

Author	Year	Gender	Age (years)	Size (largest dimension, cm)	Treatment
Lau et al.	2010	F	59	3	Partial nephrectomy
Dhingra et al.	2011	F	40	3.8	Partial nephrectomy
Zhang et al.	2013	M	39	20	Radical nephrectomy
Zhao et al.	2013	F	59	3.6	Radical nephrectomy
Li et al.	2000–2014	F	56	1.8	Partial nephrectomy
Li et al.	2000–2014	M	33	4.5	Radical nephrectomy
Li et al.	2000–2014	M	46	7.3	Radical nephrectomy
Li et al.	2000–2014	M	70	4.8	Radical nephrectomy
Li et al.	2000–2014	M	69	4.2	Radical nephrectomy
Yang et al.	2016	F	19	7	Radical nephrectomy
Qiao et al.	2018	F	36	6	Radical nephrectomy
Riley et al.	2020	M	57	3.5	Radical nephrectomy

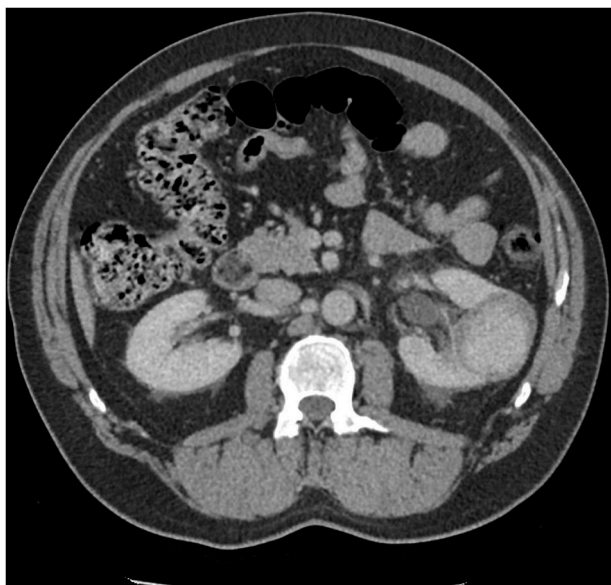


Image 1. CT renal triple phase.

brown solid cut surface. The tumour abutted the renal capsule but showed no invasion into perinephric or renal sinus fat. The remaining renal parenchyma was unremarkable. Microscopically, the tumour was well circumscribed and variably cellular. It consisted of prominent vascular spaces, many with staghorn or cavernous appearance, surrounded by a moderately wide stroma composed spindle cells [Image 2]. The spindle cells had eosinophilic cytoplasm and plump, round to oval nuclei lacking any nuclear atypia or hyperchromasia. The cells were arranged in concentric layers around the vessels and occasionally formed fascicles. Mitotic activity was not identified and there was no evidence of necrosis. There was focal nodular protrusion of lesional stroma covered by regular endothelium into the lumen of a vessel, but no lymphovascular invasion was noted.

The lesional cells were diffusely positive for alpha smooth muscle

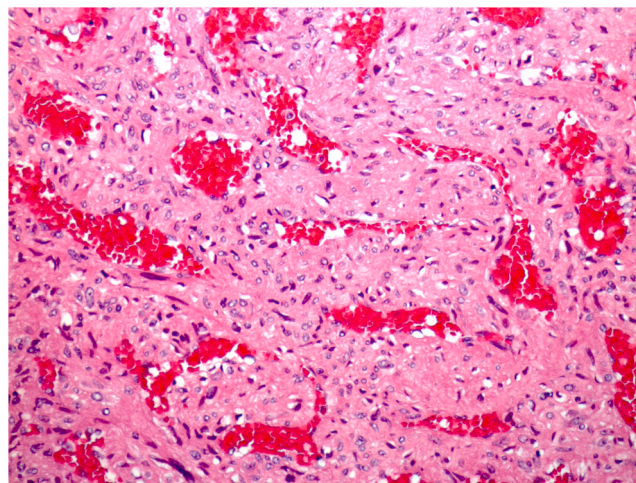


Image 2. H&E stain.

actin and h-caldesmon. Desmin was largely negative, with only limited staining of occasional spindle cells noted. CD34 showed patchy positivity. Focal non-specific weak staining for Cathepsin-K was seen. CD31 and ERG stained only the vascular endothelial lining of the prominent blood vessels within the lesion. BRAF V600E showed scanty focal staining in a small minority of lesional cells. The lesional cells were negative for PAX8, c-kit, oestrogen receptor, progesterone receptor, MNF116, AE1/E, melan-A, HMB-45, S100, SOX-10, STAT-6, p63, CD31 and ERG.

Discussion

Due to its rarity, the morphologic features, immunohistochemical profiles and biological behaviour of this tumour have not been entirely understood.⁴ The lack of available clinical research serves to increase the difficulty of diagnosing renal myopericytoma.³

CT findings of renal myopericytoma are similar to RCC, often leading to misdiagnosis.^{1,2} CT typically reveals a well-circumscribed solid lesion with heterogenous contrast enhancement, occasionally peripheral enhancement with lack of central enhancement.³ Just four of the twelve case reports have included images for comparison underlining the importance of documenting the characteristics of this lesion. This case appears to be consistent with previously described radiological findings.

Myopericytoma is morphologically heterogenous and forms part of a histological spectrum of tumours including myofibroma/myofibromatosis, glomus tumours and angioleiomyoma.⁴ Given this significant overlap, myopericytomas are often misdiagnosed.³ Typically, the cells in renal myopericytoma widely express SMA and h-caldesmon and, more rarely, CD34 and desmin on immunohistochemical staining. This case was partially positive for both CD34 and desmin which is an unusual finding with only two of the twelve documented tumours expressing patchy desmin reactivity.

All cases to date have undergone surgical excision. The prognosis of these lesions is excellent with no cases of a renal primary leading to metastasis with the longest follow-up being 66 months. This further strengthens the case for early diagnosis in order to avoid invasive radical treatment and its complications. Surveillance may be substituted in lieu of radical treatment if early diagnosis was more readily achievable. This would rely primarily on identifying imaging characteristics with sufficient confidence to prompt biopsy over surgical excision. Further studies should continue to examine imaging features which may help to identify this rare lesion radiologically.

Conclusion

Given the significant overlap between established clinical entities in this relatively novel tumour with unclear radiological characteristics, thorough reporting of variation between cases is paramount to distinguish subtypes and recognise patterns of behaviour which may influence treatment options and follow-up. Particular emphasis should be placed on histological distinction of morphological subtypes within the established spectrum of disease. If pre-operative diagnosis becomes possible, this may obviate the need for invasive surgical treatment and the comorbidity that entails.

Declaration of interests statement

None.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Author statement

First Author: Mr Thomas Riley; conceptualisation, methodology, writing.

Second Author: Mr P Shenjere: review and editing.

Third Author: Mr A Jain: supervisor.

Fourth Author: Mr S Sunder: supervisor.

Consent

Signed consent for publication, including above images, was obtained from the patient prior to publication.

References

1. Dhingra S, Ayala A, Chai H, et al. Renal myopericytoma: case report and review of literature. *Arch Pathol Lab Med.* 2012;136(5):563–566.
2. Zhao M, Williamson SR, Sun K, et al. Benign perivascular myoid cell tumor (myopericytoma) of the urinary tract: a report of 2 cases with an emphasis on differential diagnosis. *Hum Pathol.* 2014;45(5):1115–1121.
3. Qiao Y, Zhou Z, Yan W, et al. Renal myopericytoma: a case report and literature review. *Int J Clin Exp Med.* 2018;11(5):5193–5199.
4. Lau SK, Klein R, Jiang Z, et al. Myopericytoma of the kidney. *Hum Pathol.* 2010;41(10):1500–1504.