

## Case Report

# Renal myopericytoma: a case report and literature review

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**Abstract:** Renal myopericytoma is a rare mesenchymal neoplasm of the kidney. Here, we report a new case that represents the eleventh renal myopericytoma case reported worldwide according to the PubMed database. Myopericytoma is described in the literature as a rare mesenchymal neoplasm that primarily originates from subcutaneous soft tissue and rarely originates from the viscera, and it manifests as spindle-shaped cells that are histologically characterized as concentric perivascular growths. However, renal myopericytoma is not easily recognized. We report a new case of renal myopericytoma which is treated in laparoscopic radical nephrectomy by us. Meanwhile, we reviewed and analyzed cases of renal myopericytoma and summarized the epidemiology, diagnosis, pathology, treatment and prognosis characteristics to improve the recognition of this disease.

**Keywords:** Myopericytoma, kidney, epidemiology, pathology

### Introduction

Renal myopericytoma is a rare mesenchymal neoplasm of the kidney that shares common characteristics with myopericytoma. However, renal myopericytomas can originate from the viscera; thus, these neoplasms also present unique characteristics. However, the lack of clinical research on these neoplasms increases the difficulty of diagnosing renal myopericytoma. Although myopericytoma was defined by Requena in 1996, [2] renal myopericytoma was firstly reported by Sean K in 2010, [1] who defined it as a rare mesenchymal neoplasm that differentiates into perivascular myoid cells. Renal myopericytoma is rarely malignant and commonly originates from subcutaneous muscular and vascular tissue and rarely originates from the viscera. Histologically, myopericytomas contain spindle-shaped myoid cells with bland, round or ovoid nuclei in a concentric perivascular arrangement [1-8]. Morphologically, myopericytomas belong to the class of perivascular myoid cell neoplasms that also includes myofibroma/myofibromatosis, angioleiomyomas and glomus tumors [3]. Myopericytoma is typically misdiagnosed as myofibroma or angioleiomyoma, and it is now

classified separately by the WHO. An analysis of the current data indicates that renal myopericytoma also follows the above classification.

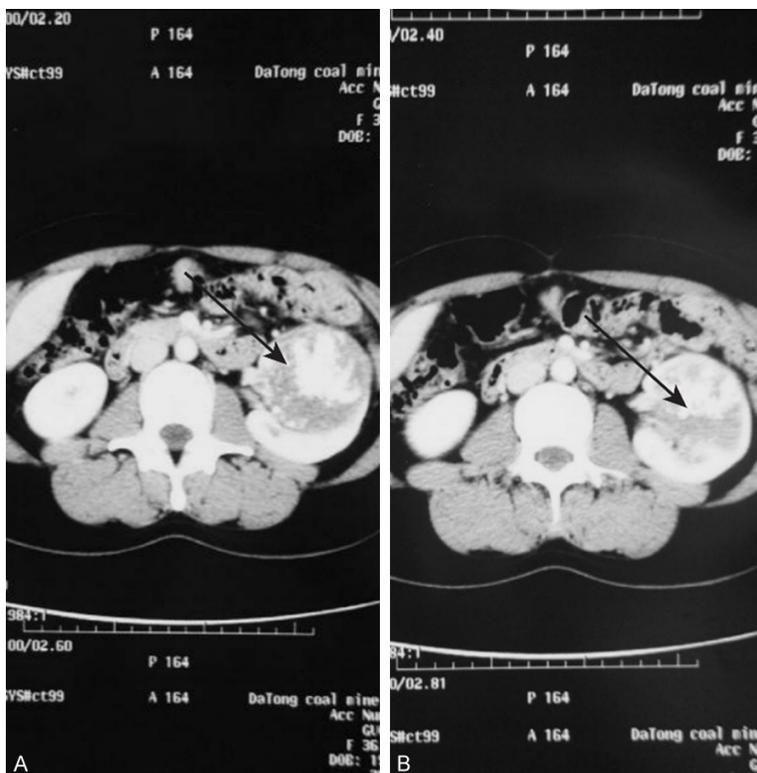
According to the PubMed database, five publications have reported ten cases of renal myopericytoma to date [1, 4-7]. Here, we report and analyze one newly identified renal myopericytoma to broaden the recognition of this neoplasm.

### Case report

In this case, a 36-year-old Chinese female presented with a mass in the upper pole of the left kidney discovered by routine urinary ultrasound and CTU (CT Urography), and she had no hematuria or abdominal pain. The patient was admitted to Peking Union Medical College Hospital for further diagnosis and treatment. A contrast-enhanced CT scan of the abdomen and pelvic cavity and ultrasound of the urinary system revealed a 6 cm diameter heterogeneously enhancing mass in the upper pole of the left kidney (**Figure 1**).

The patient underwent laparoscopic radical nephrectomy on August 3<sup>rd</sup>, 2016. A mass measuring 7×7×6.5 cm was found in the upper pole

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**Figure 1.** Contrast-enhanced CT scan of the abdomen and pelvic cavity revealed a 6 cm diameter heterogeneously enhancing mass in the upper pole of the left kidney projecting outward from the renal capsule in (A and B).



**Figure 2.** Grossly, a mass measuring 7×7×6.5 cm was visualized in the upper pole of the left kidney. The mass was approximately round, pale brown, solid and tenacious.

of the left kidney. The mass was pale brown, solid and tenacious and presented visible ossification. The pathologic diagnosis was renal myopericytoma of the left kidney adjacent to the renal capsule without invading the pelvis, ureter, the cut end of the vessel or the adrenal gland. The immunohistochemical results were

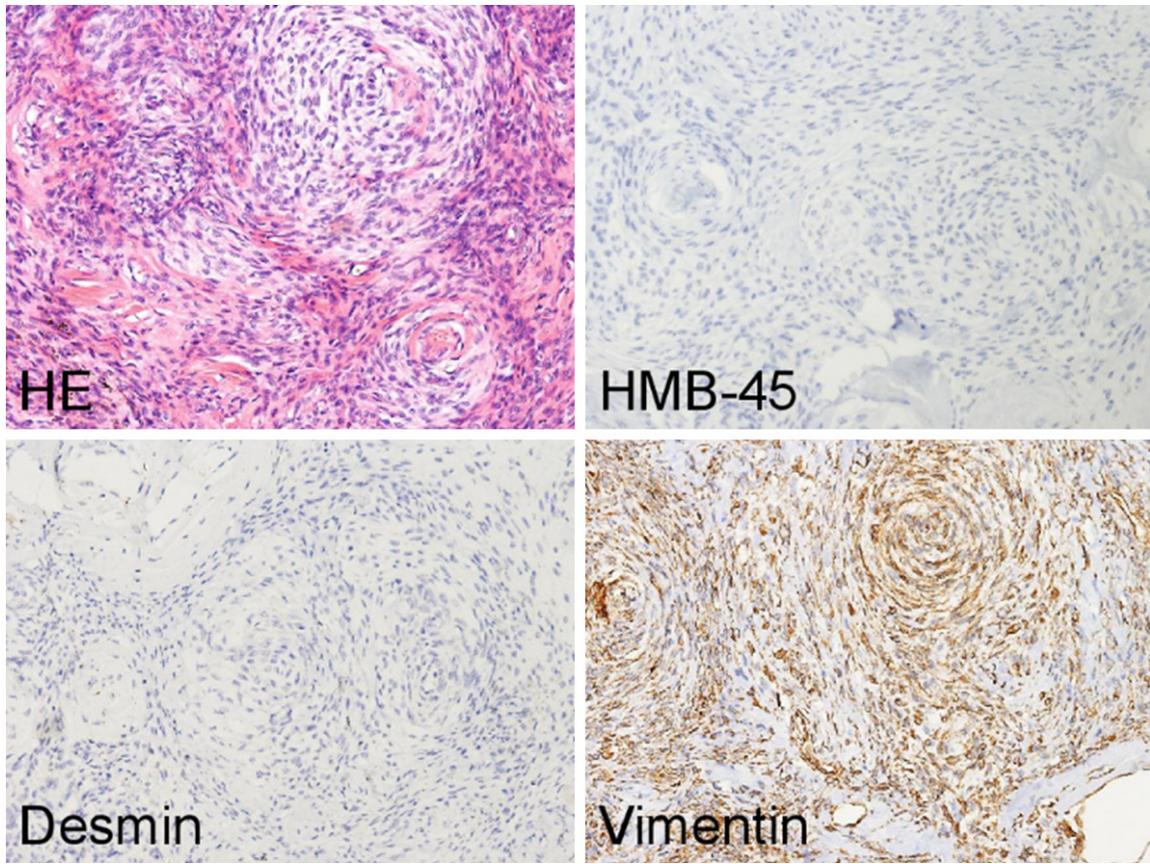
as follows: AE5: AE1/AE3 (-), Bcl-2 (+), CD31 (vessel+), CD34 (vessel+), CD56 (NK-1) (+), CD99 (partial+), desmin (-), HMB-45 (-), Ki-67 (index 1%), Melan-A (-), PAX-8 (-), S-100 (-), SMA (scattered+), vimentin (+), WT-1 (-), CD117 (-), caldesmon (-), calponin (+), ER (-), Myo-D1 (-), PR (-) and SA (-) (Figures 2, 3). The patient recovered well postoperatively and was discharged from the hospital on the 5<sup>th</sup> day after surgery. Patient was followed up for 14 months without recurrence.

### Discussion

Myopericytoma is a rare mesenchymal neoplasm that commonly originates from subcutaneous soft tissues and rarely arises from the viscera, and it is infrequently malignant. Reports in the literature indicate that myopericytoma could originate from the nervous system, lung, liver, kidney and urinary bladder [6, 9-11]. Renal myopericytomas present low morbidity, although large-scale data on its epidemiologic characteristics are not available. As previously mentioned, information on 11 cases of renal myopericytoma, including the case reported here, were collected [1, 4-7] for a review to determine the epidemiologic characteristics of this neoplasm (Table 1).

Among the published cases, six males (54.5%) and five females (45.5%) are represented. The reported neoplasms originated from the right kidney in two cases (18.2%) and the left side in nine cases (81.8%). Thus, these neoplasms tend to originate from the left kidney. Three of the reported cases occurred in the upper pole of the kidney (30%), five cases occurred in the middle part (50%) and 2 cases involved the lower pole (20%). Thus, these neoplasms tend to occur in the middle part of the kidney. In these 11 cases, the largest neoplasm was 20 cm in diameter and the smallest was 1.8 cm in diameter, and the average was approximately 5.8 cm (4.2 cm median). As previously reported, the diameter of myopericytomas is always

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**Figure 3.** Pathology of the neoplasm showing spindle-shaped cells characterized by concentric perivascular growth. The immunohistochemical results were as follows: AE5: AE1/AE3 (-), Bcl-2 (+), CD31 (vessel+), CD34 (vessel+), CD56 (NK-1) (+), CD99 (partial+), Desmin (-), HMB-45 (-), Ki-67 (index 1%), Melan-A (-), PAX-8 (-), S-100 (-), SMA (scattered+), Vimentin (+), WT-1 (-), CD117 (-), Caldesmon (-), Calponin (+), ER (-), Myo-D1 (-), PR (-) and SA (-). These four figures showed above are  $\times 200$  in magnification.

less than 4 cm [4]. When myopericytomas originate from soft tissue, they tend to be smaller at less than 2 cm diameter, and when neoplasms originate from the viscera, they tend to be larger, [8] which is consistent with our data. In all cases, only one (9.1%) had an infiltrative border, whereas the other ten cases (90.9%) had well-circumscribed borders, which is typical of the benign characteristic of myopericytomas. Renal myopericytomas are infrequently accompanied by other types of neoplasms. In the 11 cases described, only one case (9.1%) was accompanied by a papillary renal cell carcinoma. Limited research has been performed to determine whether renal myopericytomas are always accompanied by other diseases. However, reports have indicated that AIDS patients may suffer from myopericytoma associated with the Epstein-Barr virus. Currently, myopericytoma in patients with AIDS is identi-

fied as a new class of Epstein-Barr virus-associated tumor [12]. As reported, myopericytomas in patients with AIDS always arise from soft tissue but not from the kidney. Moreover, the case reported here did not present with AIDS. Accordingly, this case of myopericytoma is unlikely associated with the Epstein-Barr virus.

Currently, clinical diagnoses and screening mainly depend on ultrasound, computerized tomographic (CT) scans and magnetic resonance imaging (MRI). The review of 11 cases of renal myopericytoma indicated that three patients (27%) complained of abdominal pain symptoms, whereas eight patients (27%) had no symptoms. Renal myopericytomas always develop without symptoms and are detected by routine health examinations, which highlights the importance of imaging technology for the diagnosis and screening for renal myopericyto-

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**Table 1.** Summary of 11 cases, including the case reported herein, of renal myopericytoma diagnosed after 2010 based on reports in the PubMed database

Author	Date	Gender	Age	Symptom	Kidney (L/R)	Location	Size	Border	Immunohistochemical result	Coexistence	Treatment	Follow-up (month)
Sean K, et al	2010	F	59	Asymptomatic	L	U	3	Well-C	MSA (+), CD34 (+), bcl-2 (+), collagen type IV (+), keratin (-), EMA (-), desmin (-), S-100 protein (-), HMB-45 (-), Melan-A (-), CD31 (-), chromogranin (-), synaptophysin (-)	No	PN	ANED (8)
Sadhna Dhingra, et al	2011	F	40	Pain on the left side of the abdomen and frequent urination	L	-	3.8×3.0×3.0	Well-C	SMA (+), MSA (+), CD34 (+), Ki-67 (+) <5%, Desmin (-), S100 protein (-), HMB-45 (-), Mart 1 (-), EBV (-)	No	PN	ANED (24)
Zhiqian G Zhan, et al	2013	M	39	Pain on the left side of the abdomen	L	U	20×13×10	Well-C	Smooth muscle actin (+), CD10 (+), CD34 (-), desmin (-), S-100 protein (-), cytokeratin (-), HMB-45 (-), Bcl-2 (-), CD99 (-), Ki-67 index <1%	No	RN	ANED (20)
Ming Zhao, et al	2013	F	59	Asymptomatic	L	M	3.6×2.8×2.7	Well-C	SMA (+), desmin (patchy+), caldesmon (+), vimentin (+), calponin (+), cathepsin K (-), CD34 (-), CD31 (-), AE1/3 (-), EMA (-), C-kit (-), NSE (+), S100 protein (-), HMB45 (-), melan-A (-), chromogranin A (-), synaptophysin (-), CD10 (-), ER (-), PR (-), EBER-ISH (-)	No	RN	ANED (14)
Jun Li, et al	2000-2014	F	56	Pain on the right side of the abdomen	R	L	1.8×1.6×1.2	Well-C	VM (+), SMA (+), caldesmon (+), MSA (+), desmin (-), CD34 (-), Ki-67 (+) <2%	No	PN	ANED (66)
Jun Li, et al	2000-2014	M	33	Asymptomatic	L	M	4.5×4.5×4.0	Well-C	VM (+), SMA (+), caldesmon (+), MSA (+), desmin (-), CD34 (-), Ki-67 (+) <1%	No	RN	ANED (64)
Jun Li, et al	2000-2014	M	46	Asymptomatic	L	L	7.3×6.3×6.0	Well-C	VM (+), SMA (+), caldesmon (+), MSA (partial+), desmin (-), CD34 (-), Ki-67 (+) <2%	No	RN	ANED (46)
Jun Li, et al	2000-2014	M	70	Asymptomatic	L	M	4.8×4.3×4.0	Inf	VM (+), SMA (+), caldesmon (+), MSA (partial+), desmin (-), CD34 (partial+), Ki-67 (+) 5%	Papillary adenoma	RN	ANED (16)
Jun Li, et al	2000-2014	M	69	Asymptomatic	R	M	4.2×3.0×2.8	Well-C	VM (+), SMA (+), caldesmon (+), MSA (+), desmin (-), CD34 (-), Ki-67 (+) <1%	No	RN	ANED (14)
Jun Li, et al	2000-2014	M	59	Asymptomatic	L	M	3.6×2.8×2.7	Well-C	VM (+), SMA (+), caldesmon (+), MSA (+), desmin (patchy+), CD34 (-), Ki-67 (+) <1%	No	RN	ANED (26)
Weigang Yan, et al	2016	F	36	Asymptomatic	L	U	7×7×6.6	Well-C	AE5: AE1/AE3 (-), Bcl-2 (+), CD31 (Vessel+), CD34 (Vessel+), CD56 (NK-1)(+), CD99 (partial+), desmin (-), HMB45 (-), Ki-67 (index 1%), melan-A (-), PAX-8 (-), S-100 (-), SMA (+), vimentin (+), WT-1 (-), CD117 (-), caldesmon (-), calponin (+), ER (-), Myo-D1 (-), PR (-), SA (-)	No	RN	ANED (2)

Abbreviations are as follows: in the *Kidney (L/R)* column, L: left, R: right; in the *Location* column, U: upper pole, L: lower pole, M: middle part; in the *Border* column, Well-C: well-circumscribed, Inf: infiltrative border; In the *Follow-up* column, ANED: no evidence of disease.

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ma. Currently, limited research has been performed to determine the imaging features of renal myopericytoma. By analyzing these features, we expect to contribute insights to the imaging diagnosis of renal myopericytoma. By combining the imaging features of myopericytoma of the neck and other subcutaneous or soft tissue, Lee et al summarized the manifestations of myopericytoma. Ultrasound scan shows that myopericytoma presents as a heterogeneous, hypoechoic, solid mass with well-circumscribed borders and marked internal vascularity on color Doppler sonography. CT scan shows that the neoplasm is a homogeneous mass with a lower density that presents homogeneous enhancements with contrast [13]. Chu ZG et al also reported the imaging features of myopericytoma of the parotid gland, and the CT scan showed a heterogeneous neoplasm with poorly circumscribed borders and heterogeneous peripheral enhancement with contrast [14]. In conclusion, the imaging features of myopericytoma are inconsistent with its abundance of blood supply, which suggests marked internal vascularity. However, the imaging density and the degree of enhancement of the neoplasm vary in different tissues or locations. Furthermore, myopericytoma does not clearly present well-circumscribed borders upon imaging, and the border is occasionally unclear. The above 11 cases are consistent with these features. In the three cases with radiologic information, the renal myopericytomas present the same density as the renal soft tissue. However, well-circumscribed borders are not reported in all cases (a well-circumscribed border is reported in one case and not reported in one case, and there is no border information in the other one). The neoplasm in one case is heterogeneously enhanced and presents enhancement in the periphery but is absent in the central region (there is no information from other cases). For renal cell carcinoma, an imaging feature of the CT scan is an irregular mass in the parenchyma of the kidney and well-circumscribed borders. The CT presentation of renal cell carcinoma is similar to the parenchyma of the kidney, although renal cell carcinoma can be enhanced with contrast at a relatively lower CT value compared with that of the parenchyma of the kidney. Because renal myopericytoma may not present a well-circumscribed border upon imaging, the neoplasm may be distinguished from renal cell carcinoma.

However, these two neoplasms share a considerable number of common radiological features, which increases the difficulty of clinically differentiating these cases. Selin et al reported one case in which the patient had a subcutaneous mass with an SUVmax value of 5.8 on the right side of the back as identified by FDG PET/CT for colon cancer. Pathological examination confirmed that the mass was a myopericytoma [15]. Previous reports have suggested that the median SUVmax value is 2.6 (1.1-5.6) for FDG PET/CTs of the primary renal cell carcinoma without metastasis, whereas it is 5.0 (2.9-7.6) in primary renal cell carcinoma with metastasis [16]. Although only one such case has been reported, myopericytoma appears to present a relatively higher SUVmax value upon FDG PET/CT than primary renal cell carcinoma, especially in carcinoma without metastasis. However, this conclusion is restricted by the sample size of the research. Furthermore, the features of FDG PET/CT in renal myopericytoma have not been directly investigated. Therefore, whether FDG PET/CT can be used to clinically distinguish renal myopericytoma from renal cell carcinoma is an issue that requires further study. Currently, distinguishing these two neoplasms using imaging is difficult; therefore, the diagnosis depends on the pathology.

The primary pathologic feature of myopericytoma involves spindle-shaped cells characterized by concentric perivascular growth. Immunohistochemistry studies have revealed that these neoplasms are positive for MSA (muscle-specific actin), SMA (smooth muscle actin) and h-caldesmon and negative for markers of the epithelial, endothelial and neuroendocrine systems, such as keratins, EMA, factor VIII-related antigen and CD31. Occasionally, the tumor tissue is immunoreactive to desmin [17]. The ten previously reported cases of renal myopericytoma also present these features. The pathology of the case report herein also presents spindle-shaped cells arranged concentrically around a vessel. However, the immunohistochemistry findings are slightly different from the previous cases, which show CD31-positive and caldesmon-negative results. Myopericytomas belong to the class of perivascular myoid cell neoplasms that includes myofibroma/myofibromatosis, angioleiomyoma and glomus tumors, [3] and myopericytomas have been previ-

ously misdiagnosed as these three neoplasms. Therefore, methods of distinguishing among these neoplasms are also required. Angioleiomyoma can also present myoid cells arranged concentrically around vessels. Hence, this pathologic feature cannot be used to distinguish myopericytoma from angioleiomyoma. However, the immunoreactivity to desmin varies between these two neoplasms, with a positive reaction presented by angioleiomyoma and a negative reaction presented by myopericytoma. Furthermore, in approximately 90.6% cases of myopericytoma, the h-caldesmon reaction is positive; [18] therefore, this feature might be useful for differentiation. The pathologic features of myofibroma are different than that of myopericytoma. Specifically, a biphasic pattern characterizes the central primitive spindle cells and peripheral myoid cells [19]. Moreover, differentiating between glomus tumors and myopericytoma is relatively easier because glomus tumors contain relatively smaller, rounder cells with well-circumscribed borders compared with myopericytomas but do not present a concentric perivascular arrangement.

For the treatment of the 11 reported cases of renal myopericytoma, three patients (27.3%) underwent partial nephrectomy and eight patients (72.7%) underwent radical nephrectomy. Researchers have suggested that treatment of renal myopericytoma should include partial nephrectomy for neoplasms smaller than 4.0 cm in diameter and radical nephrectomy for neoplasms larger than 4.0 cm in diameter [5]. A review of these 11 cases indicates that all of the patients underwent partial nephrectomy for neoplasms smaller than 4.0 cm in diameter. However, among the eight patients who underwent radical nephrectomy, the diameters of the neoplasms in 2 cases were smaller than 4.0, although the diameters in 6 cases were larger than 4.0 cm, which is in consistent with the above rule. Currently, few studies have focused on the prognosis of renal myopericytoma. However, research on myopericytoma indicates that malignant myopericytomas usually share biological features that include the presence of poor circumscription, high cellularity, nuclear pleomorphism, tumor necrosis, increased mitotic activity and metastasis [8]. Overall, after long-term follow up, myopericytoma shows an excellent prognosis. Phyu P. Aung

et al reviewed 45 reported cases of myopericytoma obtained from skin and vessels, and they included only 2 cases of recurrence after treatment [20]. In the 11 cases of renal myopericytoma analyzed here, the longest follow-up time was 66 months and the shortest was two months (27.3 months on average), and none of the cases of renal myopericytoma presented recurrence or metastasis. In conclusion, renal myopericytoma presents an excellent prognosis and outcomes after nephrectomy. Although recurrence or metastasis did not occur among these 11 cases, the small sample size and relatively short follow-up time were limiting factors. Whether the above treatment method can achieve a radical effect requires further research using a larger sample size and a longer period of observation.

### Conclusions

Renal myopericytomas are rare mesenchymal neoplasms of the kidney, and most are benign. The case reported herein is the eleventh case reported worldwide. Distinguishing renal myopericytoma from renal cell carcinoma is difficult, although FDG PET/CT has the potential to perform such diagnoses. Pathologically, renal myopericytomas exhibit spindle-shaped cells arranged concentrically around vessels. Combined with immunohistochemical features, renal myopericytomas can be distinguished from other neoplasms. The treatment of renal myopericytoma mainly involves nephrectomy, which presents excellent prognoses. To date, recurrence or metastasis have not been reported.

### Disclosure of conflict of interest

None.

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