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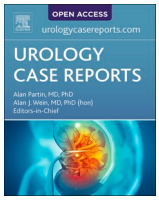

Oncology

Renal myopericytoma: A case report with a literature review

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ABSTRACT

Myopericytoma is a rare soft tissue tumor characterized by differentiation into perivascular muscle-like cells or perimuscular cells. This tumor primarily affects adults and is uncommon in children. It is predominantly found in the subcutaneous soft tissues of the distal limbs, and cases originating in the kidney are exceedingly rare. In this report, we present a case of a patient with renal myopericytoma admitted to our hospital. We also summarize the diagnostic and therapeutic features by reviewing relevant domestic and international literature.

1. Introduction

Myopericytoma was first systematically described and named by Granter et al., in 1998.[1] It is classified within the tumor spectrum of pericyte/perivascular cell tumors, alongside angiomyolipomas, myofibromas, and vascular leiomyomas. The underlying mechanisms of its development remain poorly understood. However, there are reports suggesting that trauma and the Epstein-Barr virus (EBV) may be significant risk factors.[2,3] Since the initial report of renal myopericytoma in 2010, only 12 additional cases have been documented,[4–10] bringing the total to 13 cases, including the one discussed in this article. This article provides a summary of the clinical presentation, histopathological characteristics, treatment, and prognosis of these 13 patients. (Table 1).

2. Case presentation

The patient, a 51-year-old female, was admitted for "intermittent pain in the left lumbar and back region, persisting for six days." Her physical and laboratory examinations showed no abnormalities. She has a medical history that includes chronic hepatitis B, renal lithotripsy using extracorporeal shock wave, and a subtotal hysterectomy. Contrast-enhanced computed tomography (CT) revealed a soft tissue density nodule, approximately 2.1 cm × 2.3 cm, at the upper pole of the left kidney. This nodule protrudes beyond the kidney contour and demonstrates moderate enhancement following contrast administration (Fig. 1). Contrast-enhanced magnetic resonance imaging (MRI) indicated that the upper pole of the left kidney has an abnormal, quasi-circular lesion measuring 2.5 cm × 2.2 cm. It shows isointense signals on both T1 and T2-weighted images. DWI and ADC images also display isointense signals. The lesion exhibits delayed, uniform, and marked enhancement during both the nephrographic and excretory phases, suggesting a benign neoplastic lesion, possibly an angiomyolipoma lacking fat. After confirming there were no contraindications for surgery, the patient underwent a robot-assisted laparoscopic partial nephrectomy under general anesthesia. During the procedure, a 2 cm solid tumor protruding from the surface of the upper pole of the left kidney was observed. The boundaries between the tumor, the adrenal gland, and the diaphragm were indistinct. Following careful dissection, the tumor and part of the kidney were successfully removed. Postoperative pathology results are as follows: (1) Gross examination: An irregularly shaped piece of tissue, gray-yellow and gray-red, measuring approximately 3 cm × 2.5 cm × 2.5 cm, was observed with abundant fat on the surface. A gray-white nodule, approximately 2 cm × 2 cm × 1.8 cm, was visible on the cut surface, exhibiting clear boundaries with the surrounding tissues. (2) Light microscopy examination Fig. 2: The specimen contains numerous round, oval, and short spindle-shaped muscle-like cells arranged in concentric circles around thin-walled blood vessels. The tumor cells are relatively uniform in size, feature eosinophilic cytoplasm, and the nuclei are round to short spindle-shaped and deeply stained. There were no significant mitotic figures or necrosis observed. In some areas, tumor cells are grouped and clustered with inconspicuous small central blood vessels. Some local blood vessels are dilated, forming a branching or sinusoidal pattern, and are surrounded by uniformly distributed spindle-shaped tumor cells, resembling an "extravascular endothelioma-like" configuration. (3) Immunohistochemical analysis: Tumor cells diffusely express smooth muscle actin (SMA), Vimentin, and

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Table 1
Clinical data for 13 patients diagnosed with renal myopericytoma.

No	Age(years)	Gender	Symptoms	Size(cm)	Location	Treatment	Follow-up(month)	Metastases
1[4]	59	F	No	3.5	Left	PN	8	No
2[5]	40	F	Abdominal pain	3.8	Left	PN	24	No
3[6]	59	F	No	3.6	Left	RN	14	No
4[7]	39	M	Renal mass	9.0	Left	RN	20	No
5[8]	56	F	No	1.8	Right	PN	66	No
6[8]	33	M	No	4.5	Left	RN	64	No
7[8]	46	M	No	7.3	Left	RN	46	No
8[8]	70	M	No	4.8	Left	RN	16	No
9[8]	69	M	No	4.2	Right	RN	14	No
10[8]	59	M	No	3.6	Left	RN	26	No
11[9]	46	M	No	3.2	Left	RN	25	No
12[10]	57	M	No	5.5	Left	RN	8	No
13	51	F	Low back pain	2.5	Left	PN	13	No

F = female; M = male; PN = partial nephrectomy; RN = radical nephrectomy.

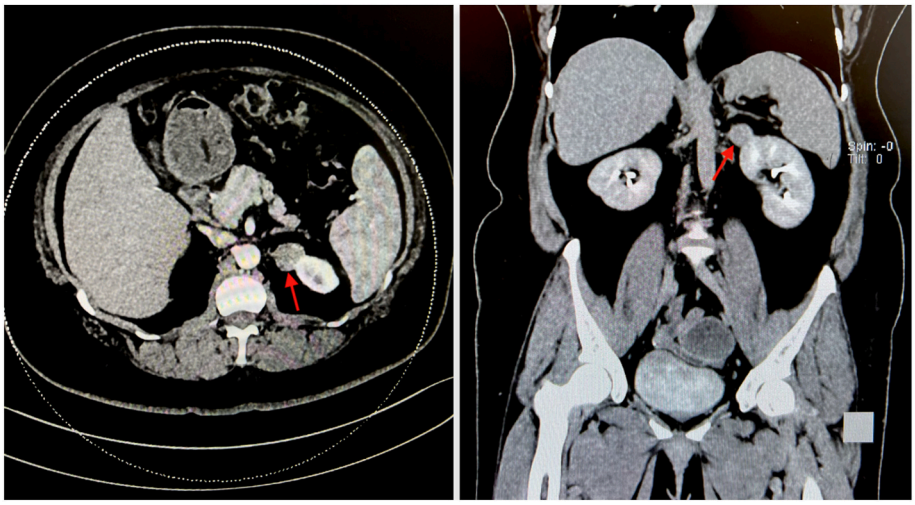

Fig. 1. Radiologic findings.

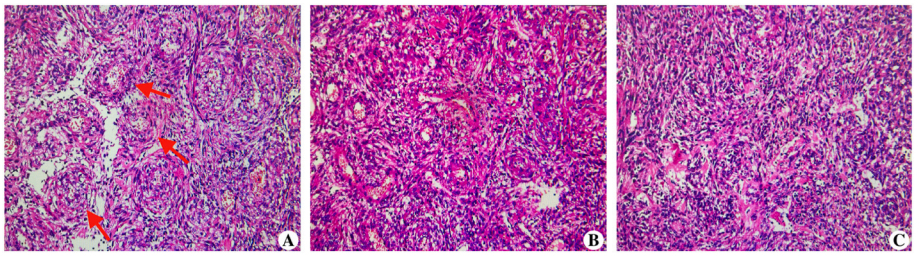

Fig. 2. Histopathological findings (HE × 200).
A: The tumor cells, which are round or oval-shaped and have eosinophilic cytoplasm with deeply stained nuclei, are arranged in concentric circles around thin-walled blood vessels of varying sizes (indicated by arrows). B: The blood vessels exhibit dilation in a branching or sinusoidal pattern and are surrounded by spindle-shaped tumor cells, presenting a configuration reminiscent of hemangiopericytoma. C: Excessive proliferation of tumor cells leads to compression of the blood vessels, resulting in occlusion of the lumen and the formation of solid cell masses.

Caldesmon, with focal expression of Desmin. The Ki-67 proliferation index is approximately 1 %. The cells do not express epithelial membrane antigen (EMA), Melan-A, S-100, HMB45, CD34, cytokeratin panel (CKP), carbonic anhydrase IX (CA9), CD31, CD117, or DOG-1. Pathological diagnosis: Renal myopericytoma. The patient recovered well after surgery. During a follow-up of 13 months, no recurrence or metastasis was observed.

3. Discussion

Renal myopericytoma is a rare benign tumor that originates from the renal mesenchyme. Most patients are asymptomatic, and the tumor is often discovered incidentally during imaging studies conducted for physical examinations or other unrelated medical conditions. A CT scan typically reveals the lesion as an isodense or hypodense mass with a regular shape and distinct boundaries. On contrast-enhanced scans, the mass demonstrates irregular enhancement. Due to the nonspecific nature of ultrasound and CT findings, determining whether the lesion is benign or malignant can be challenging. MRI is recommended for further evaluation, and a preoperative biopsy is generally not required. In this case, the MRI suggested a benign neoplastic lesion, and subsequent postoperative pathology and immunohistochemical analysis

confirmed the diagnosis of renal perimuscular leiomyoma.

Gross examination reveals that the tumor typically has clear boundaries with surrounding tissues and appears isolated. In only one case did the tumor exhibit invasive growth. No cases showed signs of tumor necrosis, bleeding, or vascular invasion. Microscopically, all cases featured numerous eosinophilic round and oval cells arranged in concentric circles around thin-walled blood vessels. The cells generally showed mild morphology with uniform size, even chromatin, and prominent nucleoli. However, in one instance, a portion of the tumor displayed slight cellular pleomorphism with deeply stained nuclei of varying shapes and binucleation, suggesting intermediate malignancy. The tumor stroma may exhibit degenerative changes such as edema, hyalinization, or myxoid degeneration. Renal myopericytoma demonstrates a broad range of histomorphological variations: five cases showed local angioendotheliomatous changes; four cases exhibited features of angiomyolipomas; five cases had abundant smooth muscle cells arranged in bundles or spiral patterns, resembling leiomyomas; three cases presented with myofibromatosis features; and four cases had areas resembling cavernous hemangiomas. Due to overlapping histological features among myopericytoma, vascular leiomyomas, myofibromas, and angiomyolipomas, including angioendotheliomatous structures, and the characteristic smooth muscle cell features in all tumor cells, histomorphological examination alone is insufficient for diagnosis. A definitive diagnosis requires immunohistochemical staining results.

Characteristic immunohistochemical markers for renal myopericytoma, such as SMA, Vimentin, and H-caldesmon, show extensive strong positive expression. Occasionally, focal expression of CD34 and Desmin is observed, with no expression of HMB-45, EMA, Melan-A, or S-100. The immunohistochemical staining results of this case align with those reported previously, confirming the diagnosis of renal myopericytoma. A high Ki-67 proliferation index (>10 %) is considered indicative of malignancy in myopericytomas. In the reported renal myopericytoma cases, only one exhibited about 5 % Ki-67, along with invasive growth characteristics, while the remaining 12 cases were classified as benign lesions, all displaying Ki-67 levels below 5 %.

The vast majority of renal myopericytomas are benign, and surgical removal remains the treatment of choice. For leiomyomas smaller than 4 cm in diameter as indicated by imaging, partial nephrectomy is usually performed. Conversely, for tumors larger than 4 cm or those displaying preoperative invasive characteristics and suspected malignancy, radical nephrectomy is advisable, with no postoperative chemotherapy or radiotherapy required. Eight patients underwent radical nephrectomy, while five opted for partial nephrectomy. None of the patients received radiation or chemotherapy. The average follow-up period was approximately 26 months post-surgery, during which no recurrences or metastases were observed. Recent studies have confirmed that PDGFRB mutations are present in myopericytoma, indicating that tyrosine kinase inhibition could be a viable therapeutic target.[11] This finding suggests the potential for both preoperative diagnosis and the use of medication as an alternative to surgical intervention in treating perimuscular leiomyomas. Nevertheless, the current body of research and clinical reports remains too limited to fully evaluate the diagnostic and therapeutic efficacy for renal myopericytoma; extensive long-term studies and additional case data are required.

4. Conclusion

In summary, renal myopericytomas are exceptionally rare benign mesenchymal tumors with a low propensity for recurrence or metastasis. Their clinical presentations and imaging findings are generally nonspecific, making definitive diagnosis dependent on a combination of cytology and immunohistochemistry. Currently, surgical removal is the only effective treatment for these tumors, necessitating long-term follow-up postoperatively.

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Consent

Informed consent was obtained from the patient for the publication of this case report.

CRediT authorship contribution statement

Yunhan Huang: Conceptualization, Investigation, Methodology, Writing – original draft. **Qian Yang:** Resources, Supervision, Validation. **Haidi Lv:** Investigation, Supervision. **Baihong Guo:** Funding acquisition, Resources, Validation, Writing – review & editing.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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