Acute abdominal pain induced by renal leiomyoma in a young patient: a case report

Weina Ma¹, Huali Jiang², Yunzhen Zhang³, Jian Zhang⁴,⁵ and Hualong Jiang³

Abstract
Renal leiomyoma is a rare benign mesenchymal tumor of the kidney that predominantly originates from the renal capsule or pelvis. However, because of its nonspecific clinical and imaging features, renal leiomyoma remains poorly characterized and may even lead to radical or partial nephrectomy on the basis of preoperative suspicion of renal carcinoma. We herein present a case involving a 12-year-old boy with acute abdominal pain who was diagnosed with renal leiomyoma based on both clinical imaging and histopathological examination. One year after radical nephrectomy, the patient recovered to good condition. This case demonstrates that the comprehensive application of imaging and histology are essential for early clinical diagnosis and effective treatment of renal leiomyoma.

Keywords
Kidney, leiomyoma, capsule, case report, clinical imaging, histopathology, nephrectomy

Date received: 25 December 2020; accepted: 25 June 2021

Introduction
Leiomyomas are rare benign mesenchymal tumors originating from smooth muscle cells and were first described by Virchow in 1854.¹ The reported prevalence of leiomyomas based on autopsy findings ranges from 4.0% to 5.5%.² Leiomyomas are most common in the uterus; they are exceptionally rare in the kidney, and fewer than 100 renal leiomyomas have been reported in the

¹Department of Hematology, the Sixth Medical Center, Chinese PLA General Hospital, Beijing, China
²Department of Cardiovascularology, Tungwah Hospital of Sun Yat-sen University, Dongguan, China
³Department of Urology, Tungwah Hospital of Sun Yat-sen University, Dongguan, China
⁴Department of Radiation Oncology, Affiliated Cancer Hospital & Institute of Guangzhou Medical University; State Key Laboratory of Respiratory Diseases, Guangzhou Institute of Respiratory Disease, Guangzhou, China
⁵Guangzhou Medical University, Guangzhou, China

Corresponding author:
Hualong Jiang, Department of Urology, Tungwah Hospital of Sun Yat-sen University, No. 1 Dongcheng East Road, Dongguan 523000, China.
Email: hualongjiang@yahoo.com
The diagnosis of leiomyomas remains challenging. Renal leiomyomas are well-circumscribed tumors without infiltration into surrounding tissue or invasion and metastasis of distant organs. Although they were included in the 2004 World Health Organization (WHO) classification of renal tumors, they are not commonly considered during the clinical evaluation of renal masses because of their rare occurrence in this location. The most common symptoms of renal leiomyomas in clinically evident cases are an abdominal mass (57%), abdominal pain (53%), or both (33%). However, only 20% of patients with renal leiomyomas present with gross hematuria. Thus, it is difficult to clinically distinguish renal leiomyomas from other types of cancer, including renal leiomyosarcoma and malignant renal cell carcinoma (RCC).

In a review of case reports and rare case series of renal leiomyoma, Patil et al. assessed 24 cases initially diagnosed as renal leiomyoma in 10 institutions from different Western areas. All renal leiomyomas were solitary and occurred in women (mean age, 63 years; range, 44–74 years). The tumor size ranged from 0.6 to 7.0 cm (mean, 2.9 cm). To the best of our knowledge, there are no previously published reports of acute abdominal pain induced by a giant renal leiomyoma in a younger patient.

**Case report**

The reporting of this study conforms to the CARE guidelines. A 12-year-old boy was admitted to the hospital because of a half-day history of acute left upper abdominal pain in December 2017. At the time of consultation, he had not experienced abdominal distention, nausea, vomiting, chills, fever, frequent urination, urgent urination, painful urination, gross hematuria, chest tightness, or shortness of breath in the previous several days. B-mode ultrasonography of the abdomen at a local hospital had shown an approximately 81- × 79-mm mass with low and weak echo, clear edges, less uniform internal echo, and an unclear boundary with the left kidney (Figure 1(a)).

The patient presented to our institution with no significant relief of his abdominal pain. Physical examination showed no protuberance in the area of either the right or left kidney; however, palpation revealed a left renal subcostal mass of about 7.0 × 7.9 cm². No percussion pain was noted in either kidney area, and no vascular murmur was heard during auscultation. The rest of the physical examination, including examination of the respiratory system, was unremarkable.

Abdominal computed tomography demonstrated a round tumor in the left kidney, measuring approximately 7.0 × 8.5 × 8.7 cm³ (Figure 1(b)). The tumor showed contrast uptake, and the enhancement range in the venous phase and delayed phase was enlarged while the degree of enhancement was reduced. Areas of irregular and flaky enhancement were present in the tumor, and the boundary between the tumor and renal parenchyma was unclear (Figure 1(c)–(f)).

Preoperative examinations (blood tests, electrocardiography, and chest radiography) showed no abnormalities. The patient’s laboratory results revealed a leukocyte count of 6.82 × 10⁹/L, erythrocyte count of 4.30 × 10⁹/L, hemoglobin concentration of 127 g/L, and platelet count of 378 × 10⁹/L. Biochemical examinations showed a prealbumin concentration of 18.9 mg/dL (reference range, 20–40 mg/dL) and creatinine concentration of 80.9 μmol/L (reference range, 27–65 μmol/L). Urinalysis showed occult blood (+) (reference, negative).

On intraoperative examination, we found a well-circumscribed, grayish, encapsulated round tumor with soft consistency.
The tumor seemed to originate from the renal capsule. Given the size of the lesion and because malignant entities such as cystic RCC or leiomyosarcoma could not be excluded, a surgical approach was deemed necessary. Therefore, radical left nephrectomy was carried out under laparoscopy.

On postoperative examination, the kidney specimen measured approximately $14.0 \times 9.5 \times 8.5$ cm$^3$. The specimen was dissected, and the tumor was gray-white and $8.5 \times 8.5 \times 8.0$ cm$^3$ in volume; it had a clear boundary and soft quality. On microscopic examination, the tumor consisted of well-oriented fascicles of long spindle cells immersed in stromal tissue. No mitosis, necrosis, or atypical cells were found (Figure 2). Immunohistochemical evaluation was positive for vimentin, desmin, and CD34 and negative for pan-cytokeratin, S100, and Ki67 ($<1\%$). The final pathologic diagnosis was renal leiomyoma.

The perioperative course was uneventful, and the patient was discharged on the fifth postoperative day. One year after surgery, the patient returned for CT re-examination. The results of plain and enhanced CT showed that the patient was disease-free (Figure 3(a)–(d)).

**Discussion**

Leiomyomas are rare benign mesenchymal tumors that mainly originate from smooth muscle cells. Renal leiomyoma is an uncommon benign mesenchymal neoplasm of the urinary system and is included in the 2016 WHO classification of renal tumors. Renal leiomyomas can be classified into three groups based on the detection scenario: (1) discovered at autopsy, (2) rare clinically significant symptomatic lesions, and (3) discovered incidentally on imaging examination. This tumor shows a female predilection (2:1), and patients’ mean age is 47 years. The most common symptoms are a palpable flank mass, abdominal pain, and hematuria. To the best of our knowledge, the present case report is the first to describe acute abdominal pain
induced by a renal leiomyoma in a pediatric patient (12 years of age).

Because of degenerative phenomena of the tumor, most renal leiomyomas have lost their typical homogeneous properties, making it difficult to differentiate them from benign lesions such as angiomyolipoma (AML) and oncocytoma as well as malignant cancers such as leiomyosarcoma and RCC. CT and magnetic resonance imaging are widely used to achieve a clinical diagnosis; these imaging techniques show that the lesion has regular margins and no evidence of local invasion. However, regular and well-defined margins and the absence of radiological signs of local invasion are uncommon features of leiomyosarcomas. RCCs typically appear hyperintense in T2-weighted images, and large RCCs usually show areas of necrosis. Meanwhile, with the absence of a macroscopic fat component and the presence of calcifications and hemorrhagic areas on imaging examination, AML and its less common fat-poor variant can be ruled out with a reasonable degree of certainty. Mixed epithelial and stromal tumor of the kidney often presents as a well-circumscribed, multicystic, and solid mass with delayed enhancement and often occurs in perimenopausal women. However, rare kidney sarcomas such as dedifferentiated liposarcoma or undifferentiated pleomorphic sarcoma also have fat

Figure 2. Histopathologic examination and immunohistochemical staining results of renal leiomyoma (×200). The immunohistochemical findings were as follows: vimentin (+), desmin (+), CD34 (+), CKpan (−), S100 (−), and Ki67 (−).
HE, hematoxylin–eosin staining; CKpan, pan-cytokeratin.
components, and they must also be included as differential diagnoses. Thus, these radiologic features are not completely sufficient to exclude papillary RCC and other rare kidney sarcomas as differential diagnoses.

Microscopically, malignant spindle cells usually display variable degrees of nuclear pleomorphism, nuclear hyperchromasia, and mitotic activity. Immunohistochemical staining of HMB45 in renal leiomyosarcomas is reportedly negative and can be used to differentiate these tumors from leiomyomas. In one study, cathepsin K reportedly showed positive expression in 8 (67%) of 12 leiomyosarcomas. Desmin is also a useful marker in the distinction between lipid-poor AML and leiomyoma. Cathepsin K is 100% sensitive in both common and leiomyoma-like AMLs and shows reactivity in >80% of cells. However, the expression of cathepsin K has not been investigated in leiomyomas. Despite the potential role of hormones in the pathogenesis of renal leiomyomas, estrogen receptor and progesterone receptor expression cannot be used in distinguishing renal leiomyoma from AMLs.

In conclusion, because of the nonspecific clinical and imaging features of renal leiomyoma, comprehensive application of imaging and histology is an effective way to achieve a diagnosis. Thus, better identification of this neoplasm is still needed to avoid unnecessary aggressive treatments, particularly in younger patients.

**Acknowledgement**

We thank the patient’s father for allowing us to use the patient’s data.
Ethics statement
This study was reviewed and approved by the Ethics Committee of Tungwah Hospital of Sun Yat-sen University. Written informed consent was obtained from the patient’s legal guardian for the publication of any potentially identifiable images or data included in this article.

Declaration of conflicting interest
The authors declare that there is no conflict of interest.

Funding
This work was supported by a grant from the Social Science and Technology Development Key Project of Dongguan (No. 201750715046462).

Author contributions
WNM, HLiJ, and YZZ wrote the case report. JZ and HLoJ reviewed and edited the manuscript. All authors contributed to manuscript revision and approved the final version.

ORCID iD
Jian Zhang https://orcid.org/0000-0002-0557-886X

References
1. Virchow R. On macroglossia and pathologic neogenesis of striated muscle [German]. Arch Pathol Anat 1854; 7: 126–138.
2. Xipell JM. The incidence of benign renal nodules (a clinicopathologic study). J Urol 1971; 106: 503–506.
3. Steiner M, Quinlan D, Goldman SM, et al. Leiomyoma of the kidney: presentation of 4 new cases and the role of computerized tomography. J Urol 1990; 143: 994–998.
4. MacLennan GT and Cheng L. Neoplasms of the kidney. In: Bostwick DG and Cheng L (eds) Urologic Surgical Pathology. 3rd ed. China: MOSBY Elsevier, 2014, pp.74–156.
5. Kho GT and Duggan MM. Bizarre leiomyoma of the renal pelvis with ultrastructural and immunohistochemical findings. J Urol 1989; 141: 928–929.
6. Patil PA, McKenney JK, Trpkov K, et al. Renal leiomyoma: a contemporary multi-institution study of an infrequent and frequently misclassified neoplasm. Am J Surg Pathol 2015; 39: 349–356.
7. Gagnier JJ, Kienle G, Altman DG, et al. The CARE guidelines: consensus-based clinical case reporting guideline development. Headache 2013; 53: 1541–1547.
8. Comperat EM, Burger M, Gontero P, et al. Grading of Urothelial Carcinoma and The New “World Health Organisation Classification of Tumours of the Urinary System and Male Genital Organs 2016”. Eur Urol Focus 2019; 5: 457–466.
9. Wagner BJ, Wong-You-Cheong JJ and Davis CJ Jr. Adult renal hamartomas. Radiographics 1997; 17: 155–169.
10. Petersen RO, Sesterhenn IA and Davis CJ. Kidney. In: Urology Pathology. Philadelphia: Lippincott William & Wilkins, Wolters Kluwer Health, 2009, pp.74–75.
11. Nagar AM, Raut AA, Narlawar RS, et al. Giant renal capsular leiomyoma: study of two cases. Br J Radiol 2004; 77: 957–958.
12. Andreoiu M, Drachenberg D and Macmahon R. Giant renal leiomyoma: a case report and brief review of the literature. Can Urol Assoc J. 2009; 3: E58–E60.
13. Murray CA, Quon M, McInnes MD, et al. Evaluation of t1-weighted mri to detect intratumoral hemorrhage within papillary renal cell carcinoma as a feature differentiating from angiomyolipoma without visible fat. AJR Am J Roentgenol 2016; 207: 585–591.
14. Chu LC, Hruban RH, Horton KM, et al. Mixed epithelial and stromal tumor of the kidney: Radiologic-pathologic correlation. Radiographics 2010; 30: 1541–1551.
15. Li CC, Li CZ, Liu CY, et al. Dedifferentiated liposarcoma of the left kidney: a rare case report. Urol Case Rep 2018; 21: 24–26.
16. Mellas S, Bouchikhi AA, Tazi MF, et al. Primary pleomorphic undifferentiated sarcoma—A rare renal localization: a case report. Case Rep Urol 2012; 2012: 862493.
17. Bonsib SM. HMB-45 reactivity in renal leiomyomas and leiomyosarcomas. *Mod Pathol* 1996; 9: 664–669.

18. Hoon V, Thung SN, Kaneko M, et al. HMB-45 reactivity in renal angiomyolipoma and lymphangioleiomyomatosis. *Arch Pathol Lab Med* 1994; 118: 732–734.

19. Zheng G, Martignoni G, Antonescu C, et al. A broad survey of cathepsin K immunoreactivity in human neoplasms. *Am J Clin Pathol* 2013; 139: 151–159.

20. Martignoni G, Bonetti F, Chilosi M, et al. Cathepsin K expression in the spectrum of perivascular epithelioid cell (PEC) lesions of the kidney. *Mod Pathol* 2012; 25: 100–111.