Primary Renal Leiomyosarcoma
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ABSTRACT
To optimise the diagnosis and treatment of primary renal leiomyosarcoma, we present our experience with a similar case and review the literature.

A 49-year woman was incidentally found to have a palpable mass in the right kidney. CT scan revealed an enhancing, heterogeneous, 6×7×9 cm right renal mass. On suspicion of renal carcinoma, a right radical nephrectomy was performed.

The pathological diagnosis was primary renal leiomyosarcoma. After 40 months of regular follow-up, local recurrence was found, and she had liver metastases. She died of cachexia due to tumor metastasis, and the survival period was just 44 months.

With unobvious clinical manifestations, preoperative imaging and postoperative pathological examination might be helpful for an accurate diagnosis. Radical nephrectomy to completely remove the tumor is recommended, and the combination of neoadjuvant or postoperative therapy should also be considered.

Key Words: Renal neoplasms, Leiomyosarcoma, Diagnosis, Therapy.

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uneventful. She presented with recurrence of tumor at 40 months after the radical operation and had liver metastases. She died of cachexia due to tumor metastasis, and the survival period was 44 months.

**DISCUSSION**

Adult primary renal sarcomas of rare tumor types, accounts for approximately 1 - 2% of all primary renal tumors. Of these, leiomyosarcoma is the most common histological subtype of renal sarcoma, accounts for 50% - 60% of later. These tumors can occur at any age and are more common in women aged 40 - 60 years. The incidence observed in females may be related to the deletion of one or more genes on the X chromosome, which evades X chromosome inactivation. By analysing 57 cases of renal leiomyoma and leiomyosarcoma characteristics, Gupta et al. observed that leiomyosarcomas exhibited a higher mitotic rate, Ki67-labelling index, and lack of expression of the estrogen receptor (ER) and progesterone receptor (PR). Primary renal leiomyosarcoma is genetically complex and usually shows a "chaotic" karyotype, including aneuploidy or polyploidy with no detectable recurrent tumor-specific translocations. In addition, cases have also been reported with the EBV virus infection, AIDS and after organ transplantation. In the present case, a 49-year woman, without obvious and specific clinical manifestations, was incidentally found to have a palpable mass in the right kidney by physical examination.
Renal leiomyosarcoma is most likely derived from the renal capsule, the renal pelvic smooth muscle cells, or blood vessels. Renal leiomyosarcoma has a high degree of invasiveness, high metastatic potential and often infiltrates renal parenchyma and renal sinus and perirenal adipose tissue. Microscopic examination reveals spindle cells, which are mainly arranged as intersecting bundles. Immunohistochemical staining was positive for α-SMA, desmin and vimentin; and tumor cells were invading the perirenal adipose tissue.

There were no significant clinical manifestations in this tumor. B-mode ultrasound showed a hyperechoic tumor, lacking blood supply. CT scan revealed an enhancing, heterogeneous, right renal mass. A provisional diagnosis of renal cell carcinoma was made. Due to rapid and aggressive growth and poor prognosis, primary renal leiomyosarcoma is more prone to metastasis in the early stage, and blood and lymph nodal metastases are common. Common metastatic sites are the liver and perirenal lymph nodes; however, other sites have been reported, such as lung, bone, stomach and soft tissue. In this case, the tumor stage of T2 was prone to metastasis.

Compared with other urinary tract sarcomas, renal sarcoma exhibits a poor prognosis. According to statistics, the 5-year survival rate of patients with retroperitoneal sarcoma was 82%, while the 5-year survival rates of patients with bladder sarcoma, prostate sarcoma, and renal sarcoma were 73%, 44%, and 39%, respectively. However, our patient did not survive for 5 years. Currently, radical nephrectomy is the primary treatment of primary renal leiomyosarcoma. Multivariate survival analysis was performed on 143 patients with primary retroperitoneal leiomyosarcomas by van Dalen et al.

They concluded that complete tumor resection by radical surgery and low histology grade were the only factors affecting prognosis. However, there are also reports of partial nephrectomy with postoperative survival of 44 months. After 40 months of regular follow-up in our patient, local recurrence was found, and she had liver metastases. She died of cachexia due to tumor metastasis, and the survival period was 44 months. Adjuvant radiotherapy and chemotherapy may be used in the case of patients with renal leiomyosarcoma. Reports have indicated that chemotherapy (specifically gemcitabine and docetaxel) and radiotherapy may benefit patients’ survival after radical nephrectomy or partial nephrectomy. In addition, phase II studies report the possibility of using a tyrosine kinase inhibitor, such as sunitinib for the treatment of primary renal leiomyosarcoma. Reports have indicated that combined treatment with chemotherapy or radiotherapy after radical surgery can not only prolong the tumor recurrence time but also slightly prolong the survival time. There is a need for more studies to strengthen the evidence base.

In summary, primary renal leiomyosarcoma is a rare, aggressive tumor of kidney with a poor prognosis. A high level of suspicion should be kept in mind when preoperative B-ultrasound shows a hyperechoic solid tumor with a lack of blood supply; whereas, the CT scan shows uneven density of tumor with delayed enhance-

**REFERENCES**

1. Narula V, Siraj F, Bansal A. Renal leiomyosarcoma with soft tissue metastasis: An unusual presentation. Can Urol Assoc J 2015; 9(3-4):E139-41. doi: 10.5489/cuaj.2396.
2. Venkatess K, Lamba Saini M, Niveditha SR, Krishnagiri C, Babu S. Primary leiomyosarcoma of the kidney. Patholog Res Int 2010; 2010:652398. doi: 10.4061/2010/652398.
3. Tajima S, Waki M, Fukuyama M. Pleomorphic leiomyosarcoma with a dedifferentiation-like appearance in the kidney: Case report and literature review. Med Mol Morphol 2016; 49(4):238-42. doi: 10.1007/s00795-015-0103-6.
4. Ozturk H. High-grade primary renal leiomyosarcoma. Int Braz J Urol 2015; 41(2):304-11. doi: 10.1590/S1677-5538.IBJU.2015.02.17.
5. Tanaka T, Koie T, Iwabuchi I, Ogasawara M, Kawaguchi T, Ohyama C. Primary leiomyosarcoma of a horseshoe kidney in a woman with Turner syndrome: A case report. BMC Research Notes 2014; 7:491. doi: 10.1186/1756-0500-7-491.
6. Evans D, Fowlkes N. Renal leiomyosarcoma in a cat. J Vet Diagn Invest 2016; 28(3):315-8. doi: 10.1177/1040637816638126.
7. Gupta S, Jimenez RE, Folpe AL, Cheville JC. Renal leiomyosarcoma and leiomyosarcoma: A study of 57 cases. Am J Surg Pathol 2016; 40(11):1557-63. doi: 10.1097/PAS.0000000000000681.
8. Miettinen M. Smooth muscle tumors of soft tissue and non-uterine viscer: Biology and prognosis. Mod Pathol 2014; 27(Suppl1):S17-29. doi: 10.1038/modpathol.2013.178.
9. Nicolos MM, Tamboli P, Gomez JA, Czerniak BA. Pleomorphic and dedifferentiated leiomyosarcoma: Clinicopathologic and immunohistochemical study of 41 cases. Hum Pathol 2010; 41(5):663-71.
10. van Dalen T, Plooij JM, van Coevorden F, van Geel AN, Heskstra HJ, Albus-Lutter C, et al. Long-term prognosis of primary retroperitoneal soft tissue sarcoma. Eur J Surg Oncol 2007; 33(2):234-8. doi: 10.1016/j.ejso.2006.09.020.