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Cystic Renal Leiomyosarcoma Treated With Partial Nephrectomy

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ABSTRACT

Renal sarcoma is rare among all renal malignancies. The present report is a case of renal leiomyosarcoma in a 59-year-old woman. She had an asymptomatic cystic renal tumor that was incidentally found by abdominal computed tomography. The characteristics of the tumor mimicked cystic renal cell carcinoma. Partial nephrectomy was completed. The pathological diagnosis was low-grade leiomyosarcoma. The patient has remained well without recurrence for 44 months following the surgery.

INTRODUCTION

Leiomyosarcoma constitutes between 0.5% and 1.5% of renal tumors in adults [1,2]. The tumors usually arise from either the renal capsule or smooth muscle tissue in the renal pelvic wall [2]. Prognosis is usually good at the early stage of the disease, following radical excision. However, despite intervention, 40% of patients die from the disease due to progression [3].

There are no specific indicators for renal leiomyosarcoma in imaging studies. Abdominal computed tomography (CT) scan typically demonstrates a large soft tissue mass involving the kidney. Cystic change in leiomyosarcoma is occasionally observed. Differentiation of renal leiomyosarcoma from renal cell carcinoma is often difficult on the basis of clinical manifestation and radiographic findings. The authors present a case of cystic leiomyosarcoma in which the mass was composed of a thick cystic wall with a large cystic portion, mimicking cystic renal cell carcinoma.

CASE REPORT

A 59-year-old woman with an asymptomatic renal tumor that was incidentally found by CT scan was admitted to the hospital. There was no significant family history of neoplastic disease. Physical examination revealed no abnormality.

Complete blood count, blood biochemical values, and urinalysis were all within the normal limits. Abdominal CT indicated a cystic renal mass, 4 cm in diameter, in the lower pole of the right kidney (Bosniak classification, category IV). The wall of the cystic renal mass was thick, and it was enhanced in the early phase of contrasted CT (Figure 1). The authors made a presumptive diagnosis of cystic renal cell carcinoma.

Partial nephrectomy was carried out by means of lumbotomy. The surgical specimen revealed a clearly defined cystic mass measuring 5.0 cm in diameter. The cystic lesion was clearly demarcated and did not extend outside Gerota's fascia. Microscopically, there were myofibrils that were arranged in cross formation. There was pleomorphism, mitotic activity, hyalinization, and vascular proliferation in the specimen. Surgical margins were negative histologically (Figure 2a; Figure 2b). Sections were stained with a broad panel of immunohistochemical markers using monoclonal antibodies. The tumor tissue was positive for vimentin and

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Abbreviations and Acronyms
CT = computed tomography
NSS = nephron-sparing surgery

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case report

Figure 1. Abdominal CT Scan Showing an Enhanced 4 cm Cystic Tumor, Located Within the Right Kidney.

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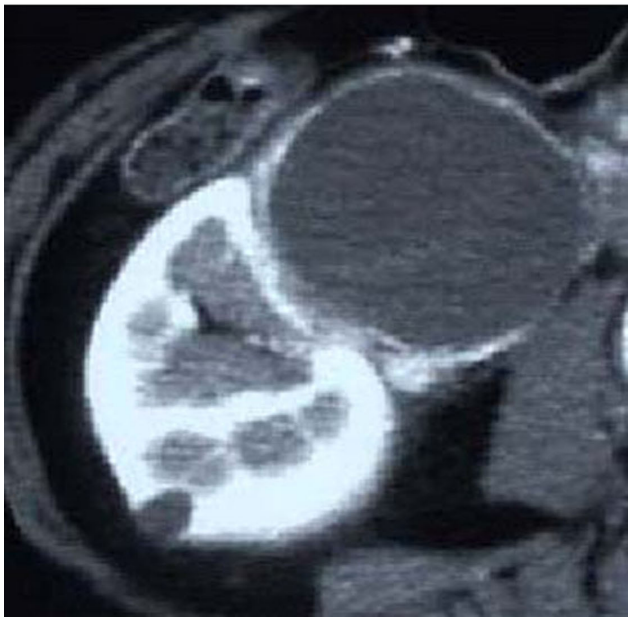

α-smooth muscle actin (ASMA) and negative for desmin, S-100, and CD34. The final pathological diagnosis of the specimen was low-grade leiomyosarcoma.

Neither adjuvant chemotherapy nor radiotherapy was applied to the patient. No evidence of local recurrence or distant metastasis was noted during 44 months of follow-up with CT scanning.

DISCUSSION

In this case, the initial clinical diagnosis was cystic renal cell carcinoma. Therefore, the authors applied partial nephrectomy to this localized renal mass in order to spare the nephron. The first nephron-sparing surgery (NSS) for a renal tumor was done by Czerny in 1887 [4]. According to the current literature, NSS for renal cell carcinoma is an acceptable therapeutic approach in patients who have a single, small localized renal mass with a normal contralateral kidney; the 5-year survival rates after radical nephrectomy and NSS are almost the same for this kind of patient [5]. There are several reports supporting partial nephrectomy rather than radical nephrectomy. Long-term follow-up revealed that renal function was well preserved in patients treated with partial nephrectomy [6,7,8]. Recently, Huang et al [9] reported that mortality rate and cardiovascular events were reduced in patients with partial nephrectomy when compared with patients following radical nephrectomy. During partial nephrectomy, the surgeon must attempt to avoid complications such as postoperative hemorrhage, urinary fistula, ureteral obstruction, and infection. Moreover, close follow-up is important to detect local recurrence.

Figure 2a. Microphotograph Showing Normal Kidney Tissue with Glomeruli and Tumor Tissue (Hematoxylin and Eosin Stain, 100x).

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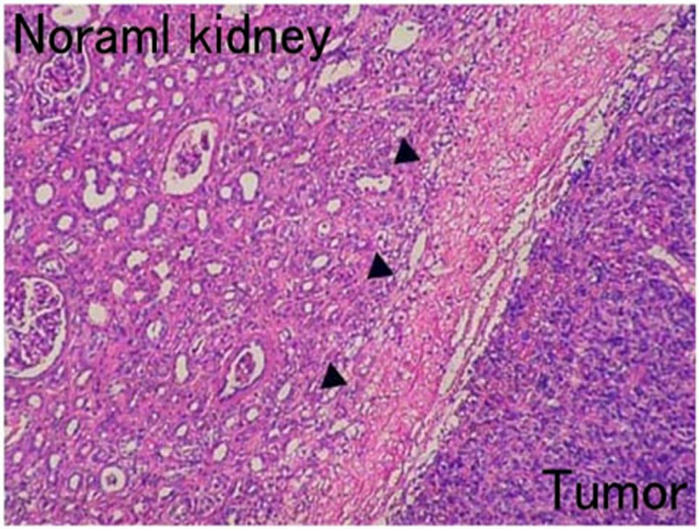

Arrowheads show the renal capsule

Figure 2b. High Power Microphotograph of Leiomyosarcoma Exhibiting Irregular Bundles of Spindle and Round Cells (Hematoxylin and Eosin Stain, 400x).

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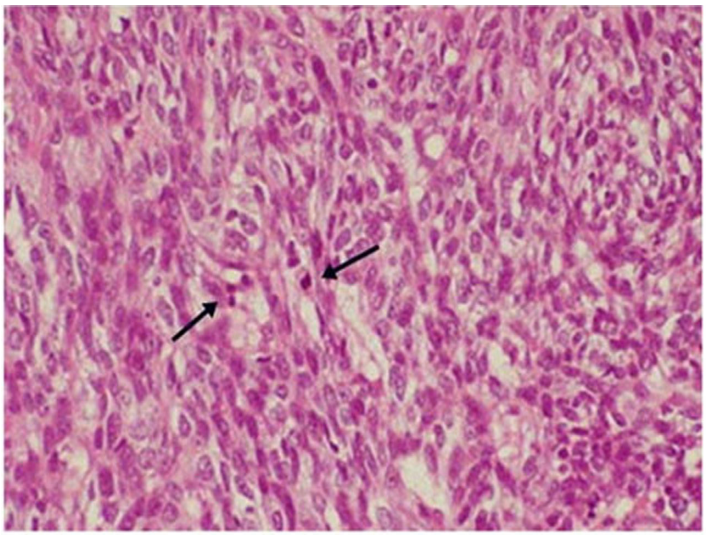

Arrows indicate mitotic features

Leiomyosarcoma of the kidney has a predominance in women and is more frequent within the fourth decade of life. However, it can be found at almost any age, with a gradually increasing incidence in the later period of life [10]. The most common presenting sign of leiomyosarcoma is an abdominal mass with or without abdominal pain. These clinical manifestations are the same as those for renal cell carcinoma. There is no specific radiographic finding for leiomyosarcoma. In general, myogenic tumors such as leiomyoma and leiomyosarcoma contain densely packed spindle-shaped muscle cells and variable amounts of fibrous connective tissue. The fibrous connective tissue is of low intensity on both T1- and T2-weighted magnetic resonance images. However, these findings are observed not only in myogenic tumors but also in neoplasms of various organs and radiation fibrosis [11].

A case of cystic leiomyosarcoma mimicking cystic renal cell carcinoma, such as the present case, is very rare. However, 2 published case reports are available [1,12]. Leiomyoblastomas are often accompanied by cystic lesions due to their rapid growth, as demonstrated by Choi et al [13]. It is speculated that the cystic lesion in the present case was secondarily formed by central necrosis or hemorrhage, because necrotic tissue with hemorrhage was histologically identified within the cyst. The case presented by Choi et al was similarly enhanced in the early phase of contrast CT, but their patient's lesion was much larger.

High-grade sarcomas often metastasize and the lungs are the primary site of spread. Prognosis is poor. The majority of patients die within a matter of months, due to the progression of the disease. Low-grade sarcomas tend to pursue a more indolent course, although local recurrence is common [14]. For renal sarcomas, the best treatment modality is radical nephrectomy. Evidence in the literature indicates that, even when confined to the kidney, leiomyosarcomas have a generally poor prognosis. The 5-year survival rates are between 29% and 36% [15]. There are not enough data regarding the indication of partial nephrectomy for leiomyosarcoma. Although radical nephrectomy has been suggested as the treatment, some of the reports [1,12,16] demonstrated good results from partial nephrectomy for leiomyosarcoma, as shown in the present case. Long-term follow up is necessary to evaluate the indication of partial nephrectomy for leiomyosarcoma.

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