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Diagnosis and Treatment of Primary Adult Renal Sarcoma

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Key words: renal sarcoma; adult; pathology

Objective To investigate the clinical characteristics of primary adult renal sarcoma.

Methods A total of 1654 cases with adult renal tumors were treated during 1985 to 2009 in Peking Union Medical College Hospital. Of all, 17 cases were diagnosed as primary renal sarcoma and underwent radical nephrectomy. The clinical features of 17 such patients were retrospectively analyzed.

Results The first symptom of 10 (59%) cases in all renal sarcomas was abdominal mass. The pathological diagnosis was leiomyosarcoma (7 cases), rhabdomyosarcoma (2 cases), malignant fibrous histiocytoma (2 cases), low-differentiated sarcoma (2 cases), chromophobe renal cell carcinoma coexisting with liposarcoma (1 case), fibrosarcoma (1 case), embryonic sarcoma (1 case) and leiomyosarcoma (1 case). One patient died of tumor thrombus of the inferior vena cava during surgery. Finally, 15 cases were regularly followed up for 4 to 60 months. Till now, 1 had tumor-free survival for 9 months, and the other 14 cases died 2-38 months after the operation with a median survival time of 18 (range, 5-60) months. The median survival time of leiomyosarcoma group was 28 (range, 11-60) months, and 2 cases of malignant fibrous histiocytoma died 4 and 8 months after the operation respectively.

Conclusions The primary renal sarcoma has the clinical symptom similar with advanced renal cell carcinoma and has poor prognosis. Leiomyosarcoma might have relative good prognosis.

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P RIMARY sarcomas of the kidney are rare tumors in adults, accounting for approximately 1% of all primary renal malignancies. Most cases continue to be the subjects of single case reports or reports grouping two or three cases. In general, these tumors have a poor prognosis, although occasionally long-

term survivors are reported. This report presented the clinical, pathologic, therapeutic, and prognostic features of 17 cases with primary renal sarcoma.

PATIENTS AND METHODS

A total of 1654 cases of renal tumors were treated from 1985 to 2009 in Peking Union Medical College Hospital, wherein, 17 cases were diagnosed as primary renal sarcoma, accounting for 1.0%. The clinical characteristics,

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diagnosis, and outcome of the 17 patients undergoing radical nephrectomy were retrospectively analyzed.

RESULTS

Clinical characteristics

In the 17 cases, 9 were male and 8 were female with age from 29 to 67 years, and the median age at diagnosis was 54 years. The tumor was 5-20 cm in diameter. The presenting symptoms were abdominal mass (10 of 17, 59%), abdominal and/or back pain (8 of 17, 47%), hematuria (4 of 17, 24%), and low grade fever (2 of 17, 12%). Clinical features of the 17 cases are summarized in Table 1.

Imaging examination findings

All patients underwent ultrasonography and CT scan. Twelve cases received MRI scan and eight received kidney-ureter-bladder (KUB) radiography and intravenous pyelography (IVP).

The ultrasonograms of primary renal sarcomas in 12 cases showed a non-homogeneous echo mass with unclear boundary and that in 1 case showed strong homogeneous echo. The images of CT and MRI showed that there were inhomogeneous necrosis and liquefied areas in tumor. Enhanced CT and MRI scans revealed the boundary of the renal parenchyma and mass was clear; and the perirenal space of 1 case was homogeneous with fat density tissues. KUB and IVP images showed displacement and distortion of

the kidney or the renal collecting system. The tumor thrombus of the inferior vena cava occurred in 2 cases and renal vein tumor thrombus did in 2 cases. Primary renal sarcoma with colon (3 cases), pancreas and spleen invasion (2 cases) was observed.

Diagnosis

Stage of the tumors at diagnosis were stage II in 3 of 17 (18%) patients; stage III in 9 (53%) patients; and stage IV in 5 (29%) patients (Table 1).

Pathology results

Routine staining and immunohistochemical detection were performed for pathological diagnosis. The pathological diagnosis was leiomyosarcoma (7 cases), rhabdomyosarcoma (2 cases), malignant fibrous histiocytoma (2 cases), low-differentiated sarcoma (2 cases), chromophobe renal cell carcinoma coexisting with liposarcoma (1 case), fibrosarcoma (1 case), embryonic sarcoma (1 case) and leiomyosarcoma (1 case) (Table 1). Immunohistochemically, all cases were completely negative for Wilm's tumor gene (WT-1) protein and cytokeratin in spindle cells, and focally positive for epithelial membrane antigen (EMA) and bcl-2 in spindle cells. Seven cases with leiomyosarcomas were positive for vimentin, desmin and smooth muscle actin (SMA). Two rhabdomyosarcomas cases expressed vimentin, and two cases with malignant fibrous histiocytomas expressed lysozyme, α 1-antitrypsin (α 1-AT) and α 1-antichymotrypsin (α 1-ACT).

Table 1. Clinical features of 17 cases with primary renal sarcomas

Case no.	Age	Sex	Presenting symptom	Stage	Histological type	Primary treatment	Follow-up (mon)	Outcome
1	29	Female	Abdominal mass	II	Leiomyosarcoma	RN	12	DOD
2	54	Female	Fever	IV	Low-differentiated sarcoma	RN	NA	NA
3	61	Male	Hematuria, abdominal mass	III	Leiomyosarcoma	RN	33	DOD
4	43	Female	Abdominal mass	IV	Leiomyosarcoma/chromophobe RCC	RN	16	DOD
5	62	Male	Abdominal pain	III	Leiomyosarcoma	RN	18	DOD
6	51	Male	Abdominal mass	II	Leiomyosarcoma	RN	60	DOD
7	48	Male	Abdominal pain, hematuria	III	Rhabdomyosarcoma	RN	14	DOD
8	54	Female	Abdominal mass	III	Leiomyosarcoma	RN	36	DOD
9	67	Male	Back pain, hematuria	III	Liposarcoma/chromophobe RCC	RN	32	DOD
10	65	Male	Abdominal mass	IV	MFH	RN	8	DOD
11	46	Male	Abdominal mass, abdominal pain	III	Leiomyosarcoma	RN	28	DOD
12	51	Female	Abdominal pain	III	Embryonic sarcoma/chromophobe RCC	RN	22	DOD
13	62	Female	Fever, back pain	III	Rhabdomyosarcoma	RN	12	DOD
14	47	Male	Abdominal pain	IV	Low-differentiated sarcoma	RN	0	Die on operation
15	43	Male	Abdominal mass, hematuria	IV	Fibrosarcoma/chromophobe RCC	RN	11	DOD
16	64	Female	Abdominal mass, abdominal pain	III	MFH	RN	4	DOD
17	58	Female	Abdominal mass	II	Leiomyosarcoma	RN	9	Alive without metastatic disease

RCC: renal cell carcinoma; MFH: malignant fibrous histiocytoma; RN: radical nephrectomy; NA: not available; DOD: dead of disease.

Outcome

One patient died of tumor thrombus of the inferior vena cava during surgery and one case was lost during the follow-up period. Finally, 15 cases were regularly followed up for 4-60 months. Of them, 1 was alive and free of tumor after the operation for 9 months, the other 14 cases were found to have local recurrence or metastasis 2-38 months after the operation with a median survival time of 18 (range, 5-60) months. The median survival time of leiomyosarcoma group was 28 (range, 11-60) months, and 2 cases of malignant fibrous histiocytoma died 4 and 8 months after the operation respectively.

DISCUSSION

Primary renal sarcoma is rarely reported. It is reported that primary renal sarcoma accounts for 1.1% of renal tumors in western countries.¹ The renal tumors incidence statistics from our country show that primary renal sarcoma accounts for 0.6%-2.7% of renal tumors.²⁻⁵ Pathologic types of renal sarcoma mainly include leiomyosarcoma, liposarcoma, fibrosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma and so on, wherein, leiomyosarcoma is the most common, accounting for 50%.⁶

The symptoms of most patients with primary renal sarcoma are similar to those of patients with late stage of renal cell carcinoma. In this cohort, the first symptom was orderly abdominal mass, lumbar and abdominal pain, hematuria, and low grade fever. In 2 early stage cases, routine examination showed no special clinical manifestations. Regular physical examination might be beneficial to diagnosis of primary renal sarcoma at the early stages.

Primary renal sarcoma has no specific imaging features. Ultrasonic examination reveals the renal or retroperitoneal masses are often inhomogeneous. Typical CT image of primary renal sarcoma is a huge inhomogeneous soft-tissue mass with necrotic areas in an exophytic growth pattern.⁷ The imaging examinations are used to detect the tumor size, blood supply, its relationship to adjacent tissues and organs, and organ invasion, which help doctors find out how far the cancer has spread, determine the stage and set out treatment planning.⁸

However, the confirmed diagnosis of primary renal sarcoma is dependent on pathology. Renal leiomyosarcoma is composed of spindle cells in a fascicular pattern with cytoplasmic eosinophilia. The nucleus is spindle-shaped with atypical shape. Nuclear fusion and/or necrosis can be seen.⁹ Liposarcoma contained a lot of lipid

contents, which should be distinguished from lipoma.¹⁰ The pathological differentiated diagnosis of malignant fibrous histiocytoma with soft tissue sarcomas, such as fibrosarcoma, synovial sarcoma and so on, is difficult. Positive reaction for lysozyme and α 1-AT is helpful.¹¹ Rhabdomyosarcoma tumor shows round-shaped nuclei, prominent nuclei and eosinophilic cytoplasm, with positive reactivity for vimentin and negative reactivity for cytokeratin, leukocyte common antigen (LCA) and CD99.¹²

The primary renal sarcoma has poor prognosis. Stage of tumor, pathological type and classification are the major prognostic factors. Leiomyosarcoma and liposarcoma have relative good prognosis.

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