Primary Renal Malignant Fibrous Histiocytoma
Four-Case Report and Review of the Literature

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OBJECTIVE To evaluate the diagnosis and treatment for primary renal malignant fibrous histiocytoma, a rare tumor arising from the kidney.

METHODS The clinical and pathological data from 4 cases of malignant fibrous histiocytoma of the kidney detected in our hospital are described. One case of special interest involved a giant cell subtype, the first to be reported in the oncology literature. The clinicopathologic features and prognostic factors of this tumor were analyzed and summarized after reviewing 55 documented cases in the English and Chinese literature.

RESULTS A palpable mass (71.2%), emaciation (54.2%), and pain (54.2%) were common manifestations in renal MFH. Of all the cases, 51 were identified as a storiform–pleomorphic subtype by pathologists. In consideration of all the prognosis related factors, the residual tumor and high TNM stage predicted a shortened survival duration, but the symptom of a fever served as a better prognostic factor.

CONCLUSION Malignant fibrous histiocytoma which arises from the kidney is a rare pathologic type, and possesses a high tendency towards local recurrence and distant metastasis. Despite the poor prognosis, early detection and radical surgery can prolong survival in selected cases.

KEYWORDS: kidney, malignant, fibrous histiocytoma.

Malignant fibrous histiocytoma (MFH) is considered to be the most common soft tissue sarcoma in adult life. Typical sites are the extremities (67–75%), and retroperitoneum (6–16%). Even though involvement of the genitourinary tract is rare, isolated cases of involving the urinary bladder, prostate, penis, and spermatic cord have been described. A primary malignant fibrous histiocytoma of the kidney is very rare. Through the end of 2003, we found only 38 patients with renal MFH reported in the English literature, and only 17 cases have been mentioned in the Chinese. Here we discuss four additional cases of primary renal malignant fibrous histiocytoma including one giant cell subtype, which is the first published report. In addition, we reviewed the literature to discuss the diagnosis, morphology, prognosis, and therapeutic approaches of this infrequent tumor.
CASE REPORTS

Case 1
A 53-year-old woman presented with a 10-month history of fatigue, a slight fever of 38°C, and weight loss of about 15 kg. Physical examination revealed a palpable mass in the left flank with a mild left flank tenderness. Hematological tests showed an anemia (hemoglobin 7.9 g/dl), an elevated white blood count (9.8 x 10^9/L), and a raised erythrocyte sedimentation rate (140 mm/hr). Serum biochemistry was normal except for an elevated alkaline phosphatase at 376 U/L. Urinalysis was normal. The excretory urogram showed deformity of a bifid ureter on the right side, and a large mass arising from the upper pole of the left kidney. Ultrasound examination confirmed the presence of an upper pole mass on the left side which had a mixed echo pattern consistent with a renal cell carcinoma. A radical nephrectomy was performed through a transperitoneal approach. A solid, whitish mass, 8 cm in greatest dimension projected from the upper pole of the kidney and extended through the Gerota's fascia, but with no invasion of the renal parenchyma. The cut surface exhibited local hemorrhage and necrosis. Histologically, the tumor was composed of spindle-shaped cells frequently in a storiform pattern. The cell nuclei were hyperchromatic and pleomorphic. The incomplete fibrous capsule was infiltrated with histiocytes and lymphocytes. A 2 cm swollen lymph node resected from the left renal artery was negative. Immunohistochemically, there was strong reactivity for vimentin, lysozyme, CD68, and negative for the epithelial membrane antigen (EMA), the cytokeratin and S-100 protein. The pathological diagnosis was storiform-pleomorphic MFH (Fig.1).

After nephrectomy, the levels of the erythrocyte sedimentation rate, alkaline phosphatase and white blood count were all normalized as well as the patient's temperature. Afterwards the patient received a course of 5,000 cGy over five weeks to the left upper abdominal quadrant. The patient was rehospitalized seven months later for an afternoon low-grade fever that persisted for a week. Computerized tomography (CT) revealed multi-focal liver and lumbar vertebral metastases, and the patient expired 4 months later.

Case 2
A 48-year-old woman was admitted to the department of chest surgery because of a cough, fatigue, 10 kg weight loss over a 6-month period, and an afternoon fever (37.5°C ~38.3°C) which she had for one week. There was no history of flank pain or hematuria. Physical examination showed a large, fixed, non-tender mass in the left flank. The hemogram showed an anemia (hemoglobin 11.2 g/dl). Both erythrocyte sedimentation rate (72 mm/hr) and alkaline phosphatase (112 U/L) were over the normal range. Ultrasonography and computerized tomography of the abdomen revealed a 5 x 4 cm size, well-demarcated mass in the lower pole of the left kidney. There was no involvement of the renal vein, inferior vena cava or lymphadenopathy. Chest X-ray and CT showed multi-focal metastatic nodes in the right lung. Suspecting a stage IV renal carcinoma, the patient underwent a left nephrectomy. Grossly, the tumor was solitary, sharply defined, and the cut section was solid and gray-yellowish. The pyelocaliceal system, renal vessels, Gerota's fascia were free of tumor involvement. Microscopically, the tumor was composed of pleomorphic spindle cells arranged in fascicles or in a storiform pattern, intermixed with more rounded histiocytic appearing cells. The nuclei were enlarged and hyperchromatic with considerable variation in size and shape. More than 20 mitoses were observed in 10 high power fields. Lympho-plasmacytic...