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Primary Fibrosarcoma of Kidney

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Pathological features of renal masses can be classified as malignat, such as renal cell carcinoma (RCC), urothelial-based, sarcoma, and embryonic or pediatric tumor; benign, such as simple cyst, angiomyolipoma, oncocytoms, and renal adenoma; and inflamatorty, such as abscess, focal pyelonephritis, infected cyst, and tuberculosis.[1] Among malignant urologic tumors, RCC was the 3rd most prevalent after prostate carcinoma and transitional cell carcinoma (TCC) of the bladder.[2] Malignant tumors of the kidney in adults may arise from the renal parenchyma and renal palvis. Often, renal pelvic cancers are TCC type, comprising less than 10% of the microscopically confirmed kidney carcinomas. Renal cell carcinoma arise from renal parenchyma accounting for over 90% of kidney carcinomas.[3]

Sarcomas of the kidney are uncommon, 1% to 3%, as compared to other renal malignant tumors.[4,5] Histopathologically, renal sarcomas are malignant mesenchymal tumors of the kidney with a variety of histological types, among which fibosarcoma is the most frequent type. Other rarer sarcomas may occur and include leiomyosarcoma, rhabdomyosarcoma, osteogenic sarcoma, and liposarcoma. Fibrosarcoma of the kidney usually develops from the capsule of the kidney, which contains much fibrous and connective tissue with smooth muscle, and thus, it is thought to be the origin of mesenchymal tumors of the kidney.[5]

Microscopically, fibrosarcoma is composed of elongated spindel cells with delicate cytoplasm and long tapered nuclei. Cells were in broad fascicles, which focally produced herring bone pattern. Nuclear pleomorphism is not a feature, althogh mitoses are frequent, and necrosis is present focally. Immunohistochemical staining studies usually show positive staining for vimentin in tumor cells and negative for actin and S-100 stains.[5]

Fibrosarcoma of the kidney clinically occurs mainly in adults, and both genders are equally affected. The tumors are usually encapsulated and grow rapidly. About 40% of tumors are found to have renal vein invasion upon operation.[5]

Retroperitoneal sarcoma arises primarily from soft tissue of fibrous and adipose origin as well as muscle, nerve, and lymphatic tissue. These tissues are derived from primitive mesenchyme from the mesoderm with the same contribution from neuroectoderm. In classic reviews of these lesions, the tissue distribution in descending order is liposarcoma, leiomyosarcoma, and fibrosarcoma,

followed by other histologies. Malignant fibrous histocytoma occurs more predominantly; however, owing to intensive pathologic interest in defining this disorder, many tumors previousely were described as variant of fibrosarcoma or liposarcoma, which heve been reclassified as malignant fibrous histocytoma. Therefore, fibrosarcoma has been replaced in frequency order by this condition.[6]

Because of a lack of significant symptoms, the majority of renal fibrosarcomas are difficult to diagnose early. The usual presenting symptoms, including abdominal mass, hematuria, and flank pain, may be the late symptoms, indicating that the patient might have huge tumors. Some patients with renal fibrosarcomas are asymptomatic initially, and these are diagnosed incidentally when gastrointestinal symptoms or metastatic lesions have developed.[7] Excretory urography or computed tomography may show a mass lesion and renal arteriography usually reveals the lesion to be hypovascular.[5]

In the current issue of the *Iranian Journal of Kidney Diseases*, Chaudhari and coworkers presented a 70-year-old man with increasing pain and lumps in the right flank and constitiotional symptoms such as fever, anorexia, and weight loss.[8] Contrast enhanced computed tomography of the abdomen showed a heterogenous enhanced lobulated mass in the right kidney without any extention to renal vein. Microscopic examination of right radical nephrectomy tissue revealed partially encapsulated tumor, comprising of spindle cells in fasicular arrangement and immunohistochemistry staining showed the tumor cells were positive for vimentin and negative for cytokeratin, that indicates the presence of fibrosarcoma of the kidney and excludes sarcomatoid RCC and leiomyosarcomas.

For difinitive diagnosis of primary sarcoma of the kidney, the major criteria include: (1) the abscence of sarcoma in other sites for disproving metastase, (2) kidney, as the origin of tumor, should be confirmed grossly rather than invasion by a retroperitoneum origin, and (3) RCC sarcomatoid type should be excluded.[5] In this case report, the mass is totally limited to the kidney and no invasion to the adjasent tissue or from other retroperitoneal tissue to the kidney was reported, but microscopic and immunohistochemical staining results, all compatible with primary fibrosarcoma of kidney.

Radical nephrectomy seems to be the only modality of treatment for renal fibrosarcoma, since the tumor is resistant to radiotherapy and chemotherapy. Immunotherapy for fibrosarcoma is still bing studied in animal models. The 5-year survival after nephrectomy is less than 10%. Overall, renal fibrosarcoma is a highly malignant neoplasm and in general prognosis is bad.[5]

CONFLICT OF INTEREST

None declared.

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