Primary Renal Sarcoma with Inferior Vena Cava Thrombus Presenting with Tumor Rupture

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hemorrhagic shock; inferior vena cava thrombus; primary renal sarcoma

Case Report

A 22-year-old Thai man suffered from right flank pain off and on for 2 weeks. He was sent to a provincial hospital because of exacerbation of symptoms one day before admission to our hospital. Abdominal sonography revealed a mixed hyperechoic and hypoechoic mass arising from the right kidney. He was then transferred to our hospital for further treatment.

Abdominal CT disclosed a 16 cm tumor occupying most of the right kidney with IVC thrombus and tumor rupture, as well as retroperitoneal hematoma (Fig. 1). Intraoperative pyelography and venocavogram were unavailable due to hemorrhagic shock. Emergent radical nephrectomy and vena caval thrombectomy were performed under the impression of renal cell carcinoma with IVC thrombus and tumor rupture. Vena caval thrombectomy was done by the following steps: (1) Place Satinsky clamp in the subhepatic vena cava, proximal renal vein, and left renal vein; (2) In case of an elipse of Chinese Medical Journal (Taipei) 2001;64:183-186

Primary renal sarcoma is a rare neoplasm.\(^1,\) Only a few cases of leiomyosarcoma,\(^2-4\) rhabdomyosarcoma,\(^2\) malignant hemangiopericytoma,\(^2\) liposarcoma,\(^2\) fibrosarcoma,\(^2,3\) osteogenic sarcoma\(^4\) and malignant schwannoma\(^2\) have been reported. Complete surgical excision is the gold standard of therapy for soft tissue sarcomas at any location.\(^7\) Adjuvant radiation therapy or chemotherapy has been suggested, but has not been proven to increase overall survival of renal sarcoma. Here, we report a case of primary clear cell sarcoma of kidney with inferior vena cava (IVC) thrombus.

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vena cava around the right renal vein and carefully dissect the thrombus free from the cava; (3) Close the vena cava with a running 5-0 Prolene suture.

A 14 × 8.0 × 4.0 cm yellowish gray-white, soft-and hemorrhagic cystic tumor originating from the upper pole of the right kidney was seen with weight of 600 gm. The tumor involved the renal parenchyma, pelvis and ureter, and protruded through the capsule near the hilum with hematoma formation.

Microscopically, the tumor was composed of small-sized spindle and focal clear cells. The tumor cells were arranged in fascicular and focal storiform pattern with multifocal necrosis and vascular invasion (Figs. 2 and 3). On immunohistochemical study, tumor cells stained negative for periodic acid-Schiff (PAS) and periodic acid-Schiff diastase (PASD). Using peroxidase-antiperoxidase (PAP) stains, tumor cells were positive for vimentin and alpha-1-antichymotrypsin, but negative for cytokeratin, smooth muscle actin M851, skeletal muscle actin M874, S-100 protein and desmin. From the combination of clinical, microscopic and immunohistochemical findings, the tumor was a primitive high-grade sarcoma, without smooth muscle, skeletal muscle or neurogenic differentiation. Clear cell sarcoma was diagnosed.

The patient made an uneventful recovery. Adjuvant chemotherapy and radiotherapy were suggested, but he returned to Thailand 3 months after surgery.

**Discussion**

Sarcomas of various types can arise from the kidney, although the frequency is extremely low. Farrow et al reported 26 cases of primary renal sarcoma from a population of 2386 renal neoplasms, with 15 leiomyosarcomas, 5 malignant hemangiopericytomas, 5 liposarcomas and one rhabdomyosarcoma.  

Flank pain is the most common presenting complaint of renal sarcoma. Gross hematuria and a palpable mass are seldom found. Clinically man i fes-
tion of tumor rupture with hemor rhagic shock is very rare.

Imaging studies are inconclusive for renal sarcoma. Con trast en hanced CT and MRI help to de fine the ex tent of the tu mor and its re la tion ship to vital vas cu lar struc tures for pre-operative plan ning.

Clear cell sar coma of the kid ney is a highly ma lignant tu mor that tends to oc cur in chil dren with a male pre pon der ance, and has a ten dency to meta sas is to the bone.10-12 Re ported prin ci pal sites of dis tant meta stases are bones, lungs, and liver.12 Con tra lateral kid ney me ta tas is is sel dom re ported. IVC tu mor throm bus pre sent ing with tu mor rupture has never been re ported. In the lit er a ture, the old est pa tient with clear cell sar coma of the kidney was 14 years and 10 months.13 This case is the old est at pres ent. For young pa tients with re nal tu mor, clear cell sar coma should be con sid ered.

References