CASE REPORT

Solitary fibrous tumor of the kidney: a case report

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A solitary fibrous tumor of the kidney is a rare neoplasm that was often misdiagnosed as hemangiopericytoma, until recently. We report a case of a 35-year-old male patient with a solid, 7 cm tumor located centrally in a solitary right kidney. The patient underwent successful bench surgery and autotransplantation.

Key Words: kidney neoplasm, solitary fibrous tumor

Introduction

A solitary fibrous tumor (SFT) is a rare entity that can develop at any site. Most frequently it is found arising from the pleura. SFTs of the kidney are extremely rare. Until recently, however, some SFTs were probably misdiagnosed as hemangiopericytomas.[1] Microscopy and immunohistochemistry can successfully differentiate between renal sarcomas, gastrointestinal stromal tumors, and benign peripheral nerve sheath tumors. We report the case of a SFT in a patient with an unusual past history.

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Case report

A 35-year-old patient was referred to us for surgical removal of a solid tumor in his right kidney. The tumor was detected during diagnostic work up of idiopathic thrombocytopenia.

The patient's history included radical surgery at age 1 year for a hormone producing cancer of the left suprarenal gland. Surgery was followed by radiotherapy. At age 29 years, the patient experienced painless gross hematuria. At that time, only a central renal cyst of approximately 4 cm was diagnosed by ultrasound. The ultrasound was no longer available.

The patient was currently being seen because of idiopathic thrombocytopenia. On ultrasound of the abdomen, a solid tumor of the right kidney was seen. A magnetic resonance image (MRI) scan revealed a centrally located, 7 cm solid tumor compressing mainly

the upper pole of the right kidney and leading to mild hydronephrosis, Figure 1.

After the patient's idiopathic thrombocytopenia was successfully treated with high doses of dexamethasone, the decision was made to perform bench surgery because of a well demarcated, centrally situated solid tumor with no prior histology results.

Only the upper pole had to be partially resected. However, the vessels could not be spared due to the tumor being trapped in the pseudocapsule.

Autotransplantation to the right iliac fossa was performed. The patient's serum creatinine was 1.7 mg/dl 10 days after surgery. His metastatic work up was unremarkable, and his platelets remained stable at a high level.

The patient's follow up MRI did not reveal a local recurrence of cancer, Figure 2.

The resected tumor was solid, well circumscribed, and encapsulated, and had a maximum diameter of 8.0 cm. The cut surface appeared brownish yellow to white. Histologically the tumor was composed of fibroblastic

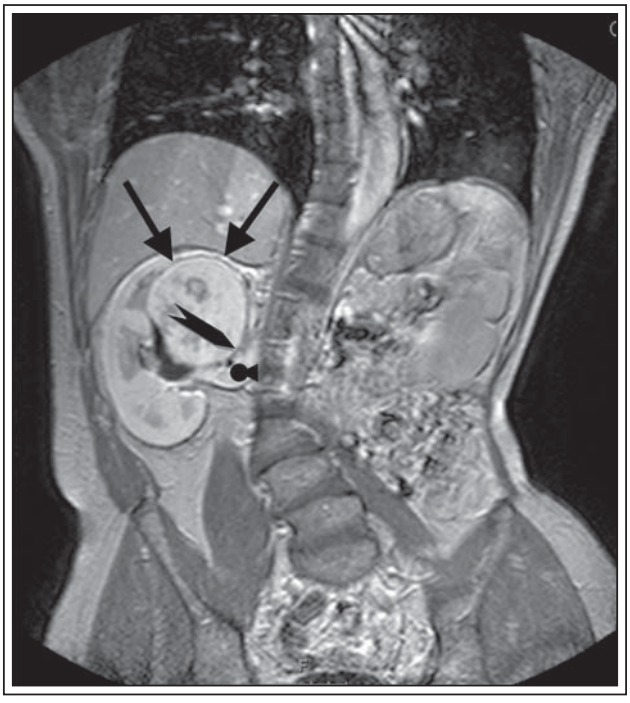

Figure 1. A predominantly hyperintense mass (arrows) can be delineated in the right kidney after intravenous contrast material delivery (Gd-DTPA) in this coronal MR-scan (T1w-FFE). The tumor margin is well defined, the renal vein is caudally displaced ( ) and the renal pelvis is slightly obstructed. Also some central necrotic areas are visible (with hypointense signal within the tumor). Furthermore, a right-convex thoraco-lumbar scoliosis is visible.

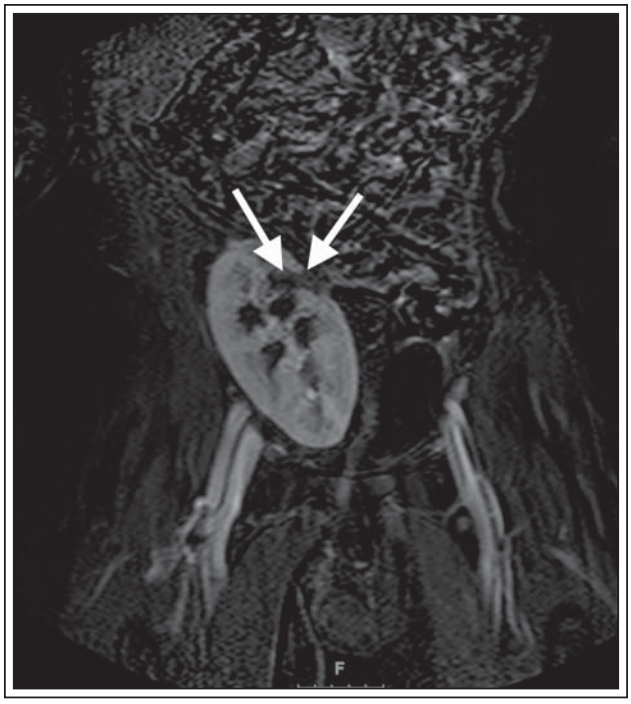

Figure 2. Subtracted T1w-image in the coronal plane (pre/postintravenous Gd-DTPA application). After autotransplantation, the kidney is now seen in the right iliac fossa. A homogeneous signal is seen throughout the renal parenchyma. After tumor enucleation, a small scar is visible in the upper medial portion of the kidney (arrows). Note the homogeneous opacification of the iliac arteries and veins.

appearing cells and thin bands of collagen. The capillaries showed a hemangiopericytoma like growth

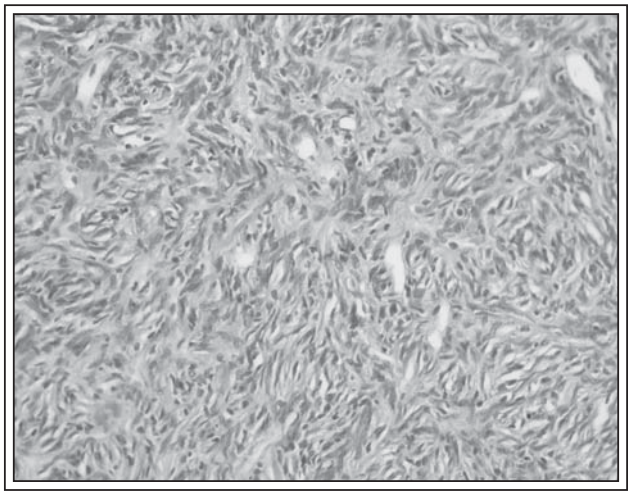

Figure 3. Histological appearance of fibroplastic appearing cells and thin bands of collagen. The capillaries show a hemangiopericytoma like growth pattern.

pattern consisting of elongated vessels with thickened, often hyalinized walls. The tumor cells stained positive with CD34, CD99, and bcl2. The proliferation rate (Ki67) was 5%, and histomorphologically, less than 4 mitoses per 10 high power fields were seen. The presence of a tumor section with fewer cells and prominent fibrosis led to the diagnosis of an SFT, Figure 3.

Discussion

SFTs are mesenchymal tumors arising at any site.[2] In the kidney, they arise from the capsule, renal pelvis, or hilar fatty tissue.[3-5] They rarely invade the kidney, even in cases of larger masses.[3] Symptoms do not differ from those reported by patients with renal cell cancer. In our patient, gross hematuria had been present several years before his current presentation. The age range of patients with reported SFTs is from 28 to 85 years.[4]

On gross appearance, the tumor is solid and grayish with a pseudocapsule. This pseudocapsule allows the development of a surgical plane and resection of the tumor with negative surgical margins. The extreme proximity of the renal vessels makes it difficult to perform nephron sparing surgery when the lesion is centrally located.

The microscopic appearance shows a cellular spindle cell proliferation. Immunohistochemical staining typically shows positive staining for CD34, vimentin, and CD99. The tumors usually do not stain positive for S-100 protein, desmin, cytokeratin, and alpha smooth muscle actin. Controversy concerning the diagnosis of SFT and hemangiopericytoma still exists due to overlapping features of the two tumors.

Poor prognosis of SFT occurs when there is incomplete resection and presence of cellularity with nuclear crowding, nuclear pleomorphism, necrosis, and more than 4 mitoses per 10 high power fields. Although pleural SFTs with these features metastasize in 80% of patients, to our knowledge, until now, no case of a renal SFT with metastases has been described in the literature. However, to our knowledge, all cases reported until now were patients who underwent radical nephrectomy. Long term follow up is necessary, since metastases can still occur after more than 15 years. □

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