

Primary Synovial Sarcoma of the Kidney

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Key Words

Synovial sarcoma · Renal tumor · Renal mass · PET-CT · SYT-SSX

Abstract

The case was a 40-year-old female. She visited a local doctor with a chief complaint of right side abdominal pain. A right kidney tumor measuring 10 cm in diameter was observed in an abdominal Computed Tomography (CT) scan. Based on the CT image, the possibility of angiomyolipoma (AML) could not be ruled out, but a high maximum standardized uptake value (SUVmax) of 7.8 was observed in a Positron Emission Tomography CT (PET-CT) scan and there was a possibility of malignancy. We therefore performed a transperitoneal right radical nephrectomy. Although adhesion of the tumor to the duodenum and the inferior vena cava was observed, it was possible to perform an excision. The tumor accounted for a large proportion of the excised kidney; the surrounding areas had taken on a cyst-like structure, and the interior comprised grayish brittle tissue exhibiting solid growth. Histologically, gland-like and cyst-like structures composed of cylindrical cuboidal cells and mainly characterized by the solid growth of short fusiform-shaped and oval-shaped basophilic cells were observed, and we believed it was a synovial sarcoma. There were no malignant findings in the adrenal gland. There have been approximately 30 reported cases around the world of synovial sarcoma that developed in the kidney, and we herein report this case with bibliographic considerations.

Case Report

The patient was a 40-year-old female with a chief complaint of right side abdominal pain. Her previous history included appendicitis at the age of 10 and an ovarian cyst at the age of 29. Her family history and history of substance use were not remarkable.

History of Events

In February 2009, the patient became aware of discomfort in the right abdominal area. In March, she visited a local doctor and a right kidney tumor was observed in a Computed Tomography (CT) image. Based on the CT image, a post-hemorrhagic tumor accompanying renal AML was suspected, but in an additional experimental Positron Emission Tomography CT (PET-CT) scan, fluorodeoxyglucose (FDG) accumulation was observed in the region corresponding to the tumor and the possibility of malignancy could not be ruled out. The patient visited our hospital for the purpose of surgery ([fig. 1a, b](#)).

Physical Findings

Other than palpation of a tumor the size of an infant's head in the right hypochondrium, there were no other physical findings in particular.

Examination Findings

Hematological values: RBC: 395/ μ l; WBC: 7,900/ μ l; Hb: 11.8 g/dl; Plt: 229,000/ μ l.

Biochemistry: Alb: 3.6 g/dl; TP: 7.0 g/dl; T-bil: 0.4 mg/dl; AST: 15 IU/l; ALT: 14 IU/l; ALP: 176 IU/l; LDH: 170 IU/l; BUN: 11 mg/dl; Cre: 0.7 mg/dl; Na: 136 mEq/l; K: 3.9 mEq/l; Cl: 102 mEq/l; CRP: 0.07 g/dl.

Urinalysis: pH: 5.5; RBC: 0–1/hpf; WBC: 0–1/hpf; Protein (–); Glucose (–).

Image Findings

Abdominal enhanced CT: A tumor 9×9 cm in size with a clearly defined border and an uneven interior was observed ([fig. 1a](#)).

PET-CT: High FDG accumulation was observed in the region corresponding to the tumor region (maximum standardized uptake value; SUVmax: 7.8) ([fig. 1b](#)).

Surgical findings: As the tumor was large in size, a reverse L-shaped incision was made and a transperitoneal radical nephrectomy was performed. As fibrous adhesion was observed between the tumor and the duodenum, a sharp detachment was made in some parts to perform an excision.

Macroscopic Findings

The tumor was 8.5×8.5×7.5 cm in size and accounted for a large proportion of the right kidney. The surrounding areas had taken on a cyst-like structure, and the interior comprised grayish brittle tissue exhibiting solid growth ([fig. 2a](#)).

Pathological Findings

In hematoxylin-eosin (HE) staining, gland-like and cyst-like structures composed of cylindrical cuboidal cells and histologically mainly characterized by the growth of short fusiform-shaped and oval-shaped basophilic cells were observed ([fig. 2b](#)). In an additional immunohistological stain, Bcl-2, MIC2, VIMENTIN, and AE1/AE3 were partially positive, while S100, WT1, and SMA were negative. Moreover, SYT-SSX1 fusion gene was also positive in a polymerase chain reaction (PCR) test. Based on these results, the patient was diagnosed to have synovial sarcoma of the kidney.

Discussion

Synovial sarcoma is a tumor that accounts for 5–10% of soft-tissue sarcomas; approximately 80% of synovial sarcomas develop in the limbs, and they also develop in the trunk of the body, the head region, and other parts of the body [1]. One of its characteristics is that in 90% of cases, SYT-SSX1 fusion genes formed by the reciprocal translocation of the chromosome X short arm and the chromosome 18 long arm are observed [2]. Regarding treatment methods, surgical excision is the first choice for primary tumors, local recurrent tumors, and pulmonary metastases. Moreover, it has been reported that for primary tumors exceeding 5 cm, combined modality therapy including radiation therapy and chemotherapy mainly consisting of ifomide is useful [3].

Synovial sarcoma of the kidney is extremely rare; it was first reported by Faria et al. in 1999, and to the extent of our research, our case will be the 28th reported case [4–6]. The age distribution of the cases ranged from 15–59 years with a median age of 35 years, and in the male:female ratio, 15 cases (59%) were males and 13 cases (41%) were females. Symptoms included hematuria, abdominal pain, and palpation of an abdominal tumor in 9 cases each. The tumor diameters ranged from 5–19 cm and the median diameter was 10 cm. Recurrence was observed in 15 cases (56%); the most frequently affected area was the lungs, which were affected in 5 cases, followed by local recurrence in 3 cases and bone and the liver in 2 cases each.

As the number of cases of synovial sarcoma of the kidney is low, its long-term prognosis is unknown. However, because the outcome in many cases involves recurrence, it was believed that additional treatment was necessary at an early stage after diagnosis. Moreover, 4 out of 9 cases of postoperative recurrence expired within 1 year after surgery and, therefore, strict follow-up is necessary. As cases involving the kidney are extremely rare, no clear medical guidelines have yet been established, and it is therefore believed that further accumulation of therapeutic experience is necessary.

Fig. 1. **a** Enhanced CT of the abdomen. **b** FDG-PET CT. SUVmax: 7.8 (arrow).

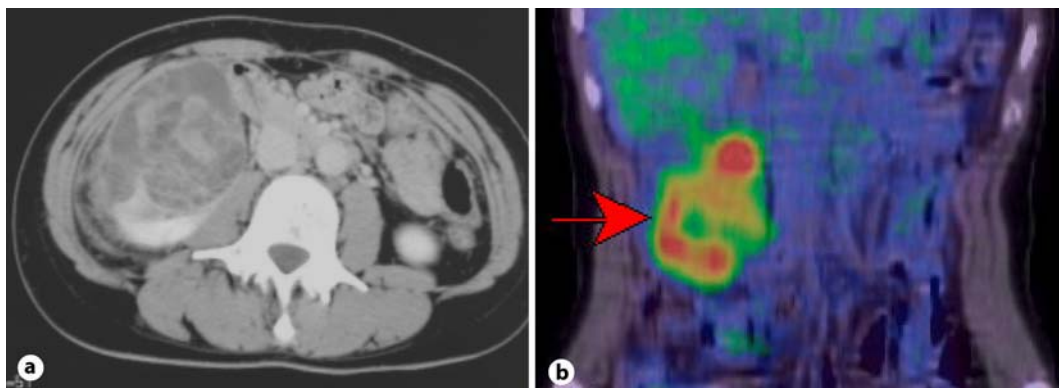
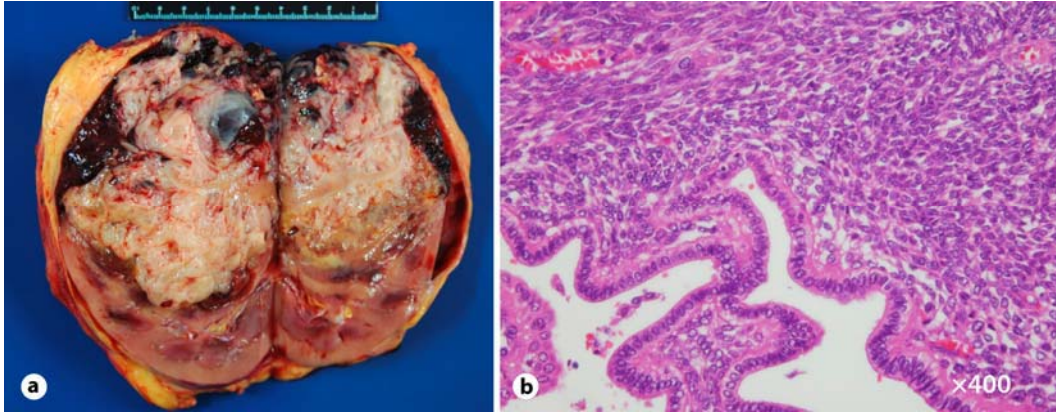


Fig. 2. **a** Gross findings. A cyst-like structure; the interior comprised grayish brittle tissue exhibiting solid growth. **b** HE staining, $\times 400$. Gland-like and cyst-like structures composed of cylindrical cuboidal cells.



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