Primary Synovial Sarcoma of the Kidney: a case report

YU-KUN TSUI, CHUNG-JUNG LIN, JIA-HWIA WANG, SHU-HUEI SHEN, CHIN-CHEN PAN, YEN-HWA CHANG, CHENG-YEN CHANG

Department of Radiology, Pathology, Division of Urology, Taipei Veterans General Hospital
Medical School of National Yang-Ming University

Primary synovial sarcoma of the kidney is a rare renal malignancy. A chromosomal translocation, t(X;18), is characteristic of this disease. We report a case of primary synovial sarcoma of the kidney showing the following imaging patterns: The tumor occupied the entire left kidney, invaded into the left renal vein and inferior vena cava, and the tumor thrombus extends upward to the level of right atrium, downward to infrarenal IVC. Enlarged lymph nodes were noted in the retroperitoneum. Nodular lesions were also noted in the liver and the possibility of liver metastasis was considered. The SYT-SSX fusion transcripts associated with the t(X;18) translocation was demonstrated by RT-PCR and the diagnosis of primary synovial sarcoma was confirmed by pathology. Primary synovial sarcoma of the kidney can only be confirmed by molecular analysis. It is difficult for the radiologist to differentiate renal synovial sarcoma from other primary renal tumors by image studies. We report a case of primary synovial sarcoma of the kidney with invasive growth pattern, which was demonstrated by MRI.

CASE REPORT

A 19-year-old male presented with right flank pain and soreness for three months. Thrombosis of inferior vena cava (IVC) was first impressed by imaging studies in another hospital at that time. Anticoagulant drug (Coumadin) was given but no improvement. He suffered from hematuria and severe flank pain several days ago. Due to IVC thrombosis and mass lesions in bilateral kidneys and liver by imaging study in another hospital, he was transferred to the division of urology of our hospital for help.

In our hospital, sonogram was performed to evaluate the condition of inferior vena cava (curved linear array 4-2, HDI3000, Advanced Technology Laboratories, Bothell, WA, USA). Presence of echogenic material in the intrahepatic part of the IVC was noted and Doppler’s examination showed no blood flow signals inside. IVC thrombosis was compatible (Fig.1). MRI was performed for preoperative further evaluation using a 1.5T scanner (GE medical system, Genesis_Signa, Milwaukee, U.S.A.). The MRI images showed a tumor occupied the entire left kidney, and invaded into the left renal vein and inferior vena cava...
The tumor thrombus extended upward to the level of right atrium of heart and downward to the level of infrarenal IVC (Fig. 3). Enlarged lymph nodes were noted in the retroperitoneum. Three nodular lesions were also found in the liver and the possibility of liver metastasis was considered (Fig. 4). Renal cell carcinoma of left kidney was impressed preoperatively by MR imaging study. The patient underwent the procedure of left radical nephrectomy, IVC and right atrial tumor thrombectomy. Histopathologic examination was done. Grossly, an ill-defined yellowish soft tumor, $7 \times 5 \times 4$ cm was noted. Left renal hilar lymph nodes, thrombosis in IVC and right atrium were proved to be metastasis. Microscopically the tumor composed of fascicles of bizarre spindle cells (Fig. 5). The SYT-SSX fusion transcript associated with the $t(X;18)$ translocation were demonstrated by RT-PCR. The final diagnosis was primary synovial sarcoma of left kidney.

**Figure 1.** Sagittal sonogram of inferior vena cava with Doppler study shows inhomogenous echogenicity occupying the IVC and no flow signal can be found in IVC (arrows). Patent portal vein is demonstrated with flow signal. (arrow heads)

**Figure 2.** a. Axial T1-weighted (TR/TE: 150/4.2) MR image shows a soft tissue mass lesion occupying entire left kidney (T) and invading into the left renal vein (arrow heads) and IVC (arrow). b. Axial T2-weighted fast spin-echo with fat suppression sequence (TR/TE: 2000/86.8) reveals high signal intensity of this lesion (T). c. The lesion shows slightly inhomogenous enhancement on postcontrast dynamic T1-weighted axial fast spoiled gradient recalled (FSPGR) (TR/TE: 5.1/1.1) MR image (T).
Primary synovial sarcoma of the kidney is a rare neoplasm and was first reported as a unique entity in 2000 [1,2]. This disease can be confirmed as the development of cytogenetic and molecular analysis in histopathology [3,4,5]. Molecular analysis by RT-PCR to detect SYT-SSX fusion transcripts associated with the characteristic t(X;18) translocation of synovial sarcoma has allowed for the confirmation of this diagnosis.

The clinical symptoms and signs of this disease include hematuria, flank pain, abdominal pain [1,2,5]. However, it is difficult for the radiologists to differentiate renal synovial sarcoma with other primary renal tumors simply by imaging studies. In the fifteen cases of Argani P. et al. series, gross pathology of tumors was typically large and the presence of hemorrhage and necrosis was frequent. Most tumors exhibited smooth-walled cysts [1]. The abdominal CT of the two cases of D.-H. Kim et al. revealed large heterogenous masses arising from kidney with subcapsular hematoma. One of the tumors presented vena cava thrombosis [2]. The CT imaging revealed a large complex mass in the case of Bella et al. [5] In the case we presented here, the MRI images showed a heterogenous soft tissue mass lesion occupying the left kidney, with renal hilar metastatic lymphadenopathy, invasion into inferior vena cava and right atrium, and suspicious liver metastasis. The radiological findings were similar to the invasive renal cell carcinoma, sarcoma groups of kidney and Wilms tumor.

Although pathological study is the only way to confirm the diagnosis of primary synovial sarcoma of kidney, radiologist should keep this disease in mind due to its variable image patterns and similar appearance to other primary renal tumors. The presence of renal tumor with invasive behavior, including regional lymphadenopathy, renal vein invasion, and distant metastasis, in young adult, should alert radiologist the possibility of this unusual renal sarcoma. The

**DISCUSSION**

Figure 3. Coronal fast spin echo T2-weighted (TR/TE: 2550/82.9) MR image shows the tumor thrombus extending upward to the level of right atrium (arrow) and downward to the level of infrarenal IVC (arrow heads).

Figure 4. Postcontrast dynamic T1-weighted axial FSPGR (TR/TE: 5.1/1.1) MR image shows one enhancing mass about 3 cm in size located in S6 of right hepatic lobe (M). Patent portal veins (arrow heads) and occlusion of IVC (arrow) are also demonstrated.

Figure 5. Histological photomicrography reveals the tumor composed of fascicles of bizarre spindle cells. (H & E stain, 400x original magnification)
complete imaging study of whole abdomen is necessary. Besides, image-guided fine needle aspiration biopsy may be useful for the preoperative diagnosis [6]. The effort of imaging study may be helpful for figuring out the strategy of treatment including surgery and adjuvant chemotherapy.

REFERENCE

腎臓原發性滑液肉瘤：病例報告

崔祐宣¹ 林重榮¹ 王家槐¹⁴ 沈書慧¹⁴ 潘競成⁴ 張廷騏³⁴ 張政彥¹⁴

台北榮民總醫院 放射線部¹ 病理部² 泌尿外科³
國立陽明大學 醫學系⁴

腎臓原發性滑液肉瘤是一種罕見的腎臟腫瘤。t (18;X) 的染色體轉位是這個疾病的特徵。我們在此報告一個腎臓原發性滑液肉瘤伴隨有以下轉移的影像特徵：腫瘤佔據整個左腎，侵入左邊腎靜脈以及下腔靜脈，並且腫瘤栓塞向上延伸到右心房與向下到腎臍下段下腔靜脈。腎虛腔膨大的淋巴結也被發現。在肝臟發現有結節病灶，考慮腎轉移的可能。病理診斷經由 RT-PCR 顯示出與 t (X;18) 染色體轉位有關的 SYT-SSX 基因轉錄，確定為腎臍原發性滑液肉瘤。
腎臍原發性滑液肉瘤只能經由分子分析來確認。對於我們而言，很难單由影像檢查去與其他原發性腎臍腫瘤做鑑別診斷。當在年輕成年人發現腎臍腫瘤具有侵襲性的表現，如局部膨大淋巴結，腎臍侵犯，與遠端轉移時，提醒我們考慮這種不常見腎臍肉瘤的可能性。

關鍵詞：腎腫瘤，磁振造影；腎臍滑液肉瘤