Original Article
Primary renal synovial cell sarcoma in 2 cases

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Abstract: Synovial cell sarcoma is a soft-tissue tumor with a unique t(X;18) (p11.2; q11.2) chromosomal translocation. Generally, the tumor occurs in the proximity of joints of young adults with a poor prognosis. Primary synovial cell sarcoma arising from the kidney is extremely rare. Here we report 2 patients who were initially considered as renal cell carcinoma but were finally diagnosed as primary renal synovial cell sarcoma by histopathology and molecular studies. Urologists should be aware of the possibility of malignancy in cystic renal masses and consider a diagnosis of renal synovial cell sarcoma. The differential diagnosis is difficult, and molecular or genetic analysis should be applied for the diagnosis.

Keywords: Synovial cell sarcoma, histopathology, renal

Introduction

Synovial cell sarcoma is a malignant mesenchymal tumor which usually affects joints of young adults with a poor prognosis, but it can also involve other sites [1]. Primary synovial cell sarcoma originated from kidney is extremely rare. After first reported in 1999, cases have been sporadically described [2]. However, no more than 50 cases have been reported to date and the knowledge of this rare malignancy is still limited. It is quite difficult to differentiate this tumor from other renal neoplasms (sarcomatoid renal cell carcinoma, metatatic sarcoma, adult Wilms tumor, solitary fibrous tumors, etc), since they may have similar histological features. The diagnosis could be confirmed by detecting the chromosomal translocation t(X;18) (p11.2; q11.2) and the fusion gene SYT-SSX. Here we report 2 patients who were initially considered as renal cell carcinoma but were finally diagnosed as primary renal synovial cell sarcoma.

Case presentation

Case one

A 38-year old female was admitted to our institution complaining of left flank pain for 2-weeks. The physical examination revealed a mild percussion pain on renal region and there was no palpable mass in abdominal region. The routine blood test, liver and renal function was all within normal limits. Ultrasounography showed a solid-cystic lesion in the lower pole of the left kidney with heterogeneous echo pattern and unclear boundary. Computed tomography (CT) was performed for further diagnostic imaging. We identified a hypodense mass (80 mm in diameter) with slow contrast enhancement located in the lower pole of the left kidney, and involving the left proximal ureter and major psoas muscle (Figure 1A and 1B). There was no evidence of metastases. Surgery was performed under the general anesthesia. During operation, gross invasion of the major psoas muscle was observed and regional lymphadenopathy was also noted. Left radical nephrectomy along with regional lymphadenectomy was performed.

Macroscopically, the tumor located in the lower pole of the left kidney was of approximately 82*80*78 mm. The cut surface was grayish with focal hemorrhage and necrosis. Microscopical pathological evaluation revealed the tumor was composed of spindle-shaped cells and infiltrated to the Gerota’s fascia (Figure 1C). The left hilar lymph nodes were positive for...
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Figure 1. A, B. CT showed a hypodense mass, 80 mm in diameter, located in the lower pole of the left kidney. C. Histopathology showed that the tumor was composed of spindle-shaped cells microscopically.

Figure 2. A, B. MRI showed a solid-cystic lesion in the lower part of the left kidney. C. Histopathology showed that the tumor was composed of spindle-shaped cells microscopically.

malignancy. Immunohistochemical analysis demonstrated cells were positive for Vimentin, Bcl-2, EMA and focal positive for MIB-1 (60%) but negative for SMA, CD10, AE1/AE3, CK7, S-100, CD117, RCC, Des, GFAP, HMB45 and Melanin A. The diagnosis of primary renal synovial cell sarcoma was made. Post-operative adjuvant chemotherapy was offered using an anthracycline- and ifosfamide-based regimen; however, pulmonary and liver metastasis was observed 3 and 7-months after surgery respectively and the patient refused any further treatment. Unfortunately, the patient died 9-months postoperatively.

Case two

A 28-year old female was admitted to our institution because of a solid-cystic mass in the left kidney detected by an annual medical examination. The physical examination didn’t find any abnormal signs. Ultrasound revealed a solid-cystic lesion in the left kidney with solid-like component and thick septations, which were enhanced by contrast-enhanced ultrasonography. Magnetic resonance imaging (MRI) demonstrated a 67*61 mm space-occupying lesion in the lower part of the left kidney, which had a cystic signal along with an irregularly outlined solid component (Figure 2A and 2B). Suspecting a cystic renal cell carcinoma of the left kidney, we performed a radical nephrectomy. Neither gross invasion of the adjacent structures nor regional lymphadenopathy was observed during operation.

Macroscopically, the well-circumscribed renal neoplasm was approximately 70*65*60 mm. The cut-surface was tawny with cystic areas and also showed focal hemorrhage and necrosis. Microscopic pathological examination revealed a monotonous tumor composed of spindle-shaped cells (Figure 2C). Immunohistochemical analysis demonstrated cells were positive for Vimentin, Bcl-2, CD99 and focal positive for EMA, CD34, CAM5.2, MIB-1 (20%)
but negative for SMA, AE1/AE3, CK19, S-100, CD117, Des, GFAP, WT-1 and Melanin A. FISH test revealed a chromosomal translocation t (X; 18) (p11.2; q11.2) and the consequent fusion gene SYT-SSX, thus a definitive diagnosis of primary renal synovial cell sarcoma was achieved. Adjuvant chemotherapy was recommended but the patient refused. Until recently, the patient was free of local recurrence or metastasis 18 months after surgery.

Discussion

Primary renal synovial cell sarcoma, first described in 1999 [2], is an extremely rare tumor with uncertain histogenesis. Less than 50 cases have been reported in the literature until recently. It generally affects young individuals of both genders, with a slight predominance in males [3]. There are no specific clinical or imaging characteristics for the diagnosis. Clinical symptoms do not differ from other malignant renal tumors, such as flank pain, hematuria or even no symptoms and detected by medical examination. Ultrasound, CT or MRI findings are similar to those of any other primary renal cancer. Heterogeneous, enhancing masses with solid and cystic components can be observed by imagine studies, but none of them are diagnostic. The diagnosis of Primary renal synovial cell sarcoma always requires pathological confirmation.

The differential diagnosis includes sarcomatoid renal cell carcinoma, metastatic sarcoma, adult Wilms tumor, solitary fibrous tumors and hemangiopericytoma, etc [4]. Primary renal synovial cell sarcomas are typically positive for vimentin, Bcl-2, CD99, CD56, and are focally positive for EMA and cytokeratin [5, 6], but these immunohistochemical markers are non-specific. Recent studies demonstrated that the expression of TLE1 is strongly predictive of SYT gene rearrangement, making TLE1 a promising and more specific marker [3, 7]. Accumulated evidences have shown that synovial cell sarcoma is specifically associated with a unique chromosomal translocation between SYT gene on chromosome 18 [8], and definitive diagnosis of a renal synovial cell sarcoma requires confirmation of rearrangement involving the SYT gene.

The treatment includes surgical resection and chemotherapy; however, no consensus has been achieved on chemotherapeutic regimens until recently. Anthracycline-only chemotherapy did not show an improved survival [9], while anthracycline- and ifosfamide-based protocols achieved a small gain in survival [10]. The clinical benefit of adjuvant chemotherapy is still controversial. There is no convincing evidence on survival benefits with respect to adjuvant chemotherapy. Therefore, adjuvant chemotherapy is only recommended for cases that clinical advantages could be expected, such as younger patients and/or larger tumors [11]. We should be aware that renal synovial cell sarcoma is a rare malignancy with aggressive course and poor prognosis.

In conclusion, primary renal synovial cell sarcoma is extremely rare, with histomorphological and immunohistochemical features that may be confused with other tumors of the kidney. The differential diagnosis of such a tumor is therefore difficult, and molecular or genetic analysis should be used to confirm the diagnosis.

Disclosure of conflict of interest

None.

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