CASE REPORT
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Primary Renal Synovial Sarcoma: A Rare Case Report

Böbreğin Renal Sinoviyal Sarkomu: Nadir Bir Olgu Sunumu

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ABSTRACT

Synovial sarcoma (SS) is mainly derived from soft tissues. Primary renal SS is a very rare malignancy with around 60 cases reported in the literature. We report a renal mass which was undistinguishable from urothelial carcinoma clinically and pathologically but diagnosed as a primary renal SS at the definitive pathological diagnosis.

Keywords

Pathology, kidney, sarcoma, synovial sarcoma, kidney neoplasms, kidney tumor

ÖZ

Sinovyal sarkom (SS) esas olarak yumuşak dokudan gelişmektedir. Primer böbrek SS çok nadir bir kanserdir ve literatürde yaklaşık 60 olgu bildirilmiştir. Kesin patolojik tanısı primer renal SS olan klinik ve patolojik olarak ürotelyal karsinomdan kolay ayrılamayan renal kitle raporlanmıştır.

Anahtar Kelimeler

Patoloji, böbrek, sarkom, sinovyal sarkom, böbrek neoplazmları, böbrek tümörü

Introduction

Synovial sarcomas (SS) account for 5-10% of adult soft tissue sarcomas and occur mostly in the proximity of large joints (1,2,3). These tumors are rarely diagnosed in unexpected sites, including the thoracic and abdominal wall, head and neck region, retroperitoneum, bone, lung, or prostate (4,5). Primary renal SS is a very rare malignancy with around 60 cases reported in the literature and first described by Argani et al. (6) in 1999 and published by Argani et al. (1,6). Primary renal SS constitutes a subtype of the cases identified as embryonal sarcoma of the kidney and can clinically mimic an advanced renal cell carcinoma, making the correct diagnosis challenging. It is also difficult to differentiate pathologically from other spindle cell histologies of the kidney such as adult Wilms tumors, sarcomatoid renal cell carcinoma, hemangiopericytoma and undifferentiated carcinoma (7). It requires immunohistochemical (IHC) staining and cytogenetic analysis for diagnosis (8). More than 90% of cases of SS are seen the chromosomal translocation t(x;18) (p11;q11). CD99, smooth muscle actin, CD34, epithelial membrane antigen, cytokeratin, S100, and B-cell lymphoma 2 (BCL2) are used in IHC staining (9,10). We report a renal mass which was undistinguishable from urothelial carcinoma clinically and pathologically but diagnosed as a primary renal SS at the definitive pathological diagnosis.

Case Presentation

Forty seven years old man investigated for left flank and abdominal pain lasting for several months. Abdominal ultrasonography revealed a left renal mass and computed tomography (CT) reported a 90x70x60 millimeters solid mass. Open radical nephrectomy was performed with transperitoneal approach. Pathology was reported transitional cell carcinoma (tumor invaded the renal calyx, ureteral surgical margins was positive). Ureterectomy and bladder cuff excision was performed for the stump of ureter after 2 weeks. Pathology was reported as non-neoplastic tissue. Two months later CT was performed because of the mechanical ileus. Multipl metastatic lesions was revealed at the lung, para-aortic area, paravertebral area and around the spleen. The patient was operated, splenectomy was performed and retroperitoneal mass was resected. Histomorphological findings was found to be identical compared with first nephrectomy material. In examined section tumor mass is observed atypical spindle-shaped cells forming bundles and diffuse growth pattern. And also trabecular pattern areas were observed in myxoid tumors and hemangiopericytoma. Tumor is highly cellular appearance and comprised of cells containing several nucleolus. A panel of immunohistochemistry was performed periodic acid-Schiff (PAS), glicogene, CK7, CK19, reticulin, BCL2, CD99, Wilms

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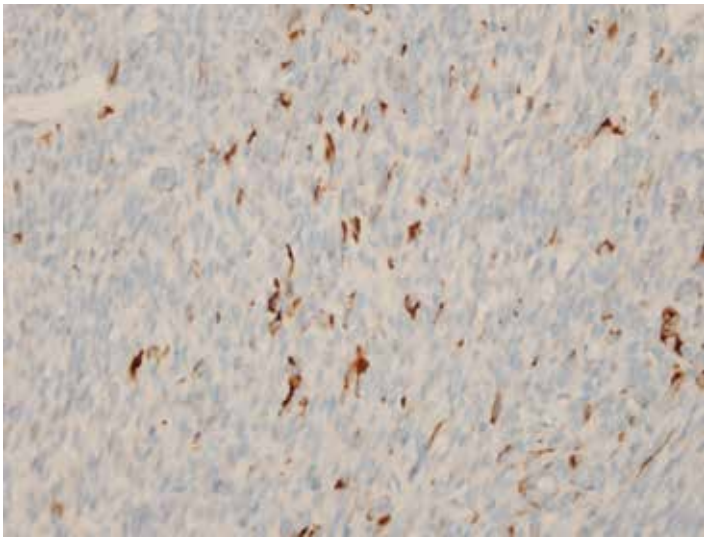

Figure 1. Histological appearance the cells are stained cytoceratin 7 with immunohistochemical (immunohistochemistery x400)

tumor-1 (WT1), desmin, caldesmon, synaptophysin, chromogranin, CD34, CD31, CK20 and S100. The tumor cells were positive for CK7, CK19, BCL2, actin and focally positive for reticulin (Figure 1). WT1 and CD99 had been stained with focal cytoplasmic granules diffusely positivity. There was no reaction to PAS, glicogene, desmin, caldesmon, chromogranin, synaptophysin, CK20, S100, CD34 and CD31. According to these findings a diagnosis of monophasic spindle cell SS was done. One months later, CT showed liver metastasis and a 15x10 cm mass that invades in the field of operation. A few lymphadenopathy (as 2.5x1 cm) and a 4.5x4 cm mass in the left adrenal area was also detected. A single dose doxorubicin was administered. Patient's general condition deteriorated and died after one month.

Discussion

Primary renal SS is a very rare tumor and comprises 1-3% of all malignant renal neoplasms (11). It has shown a gender ratio male to female: 1.7:1, a mean age at diagnosis of 37 years (ranging between 13 and 67) and mean tumor diameter of 11 cm (ranging 3-21 cm) (12). The diagnosis of SS are always problem, due to rarity and similar clinical presentation and imaging with other sarcomas. These tumors have 3 morphological variants: monophasic, biphasic and poorly-differentiated (10). The monophasic variant has difficulty in differentiating from other spindle cell sarcomas because of having only an epithelial or spindle cell component. The biphasic variant can be diagnosed with epithelial and spindle component. Poorly-differentiated subtype has undifferentiated round cells with hyperchromatic nuclei and frequent mitoses (13).

Diagnosis of SS is not possible without ancillary diagnostic techniques such as IHC and cytogenetic studies. Histopathological diagnosis is difficult. Cytogenetic studies have shown a characteristic t(x;18) (p11;q11) chromosomal translocation, over 90% of cases, as a diagnostic indicator of SS as well as cytogenetic or molecular methods have been used in order to detect it. Fluorescence in situ hybridization analysis are reported to be positive around 95% in the *SYT* gene translocation in SS but it is not apply to our case. IHC markers have been investigated in cases of SS but not to shown specific markers for diagnoses SS. WT1 expression is always found adult Wilms' tumor but not in primary tumors unlike in our case. Furthermore, malignant peripheral nerve sheath tumor is typically positive for S100, while primary renal SS are negative (14). The gold standart diagnostic study for SS is to demonstrate of *SYT* gene translocation (15,16).

The rate of metastasis on admission seems to be low. Firstly managed through surgery, there is no consensus about the role of chemotherapy on these cases, either as neoadjuvant or adjuvant therapy (7,12).

Clinically and histologically primer renal SS could not be easily diagnosed and it should be included in the differential diagnosis of a solid renal neoplasm.

Ethics

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Peer-review: Internal peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Mehmet Sinan Başay, Concept: Taha Numan Yıkılmaz, Okan Baş, Design: Taha Numan Yıkılmaz, Okan Baş, Ali İhsan Arık, Data Collection or Processing: Taha Numan Yıkılmaz, Okan Baş, İsmail Selvi, Analysis or Interpretation: Taha Numan Yıkılmaz, Okan Baş, Emine Bezer, Literature Research: Emine Bezer, Mehmet Sinan Başay, Writing: Taha Numan Yıkılmaz.

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