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Synovial Cell Sarcoma of the Kidney: A Case Report

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Abstract

Background: Primary renal sarcomas are rare neoplasm that accounts about 1% of malignant renal tumors. Prevelance of primary renal Synovial cell sarcoma is rare and comprise 1-3% of all malignant renal neoplasm. Synovial cell sarcoma overlaps with multiple spindle cell neoplasms affecting the kidney, this need immunohistochemical panel to can diagnose it. This paper reports a case of Renal Synovial cell sarcoma. We report a case of 32 year old female presented with presented by upper pole left kidney swelling. Computarized tomography (CT) revealed a heterogeneous, well marginated soft tissue mass 8x7 cm arising in the upper pole of left kidney with solid necrotic components and heterogeneous enhancement. Left radical nephrectomy was done.

Methods: The kidney was excised and gross examination revealed that upper pole of the kidney was replaced completely by grayish tan firm mass with cystic areas measuring 6x6x3 cm in diameter and shows areas of hemorrhage and necrosis. Microscopic evaluation and immunohistochemistry study were performed.

Results: The mass was Renal Synovial cell sarcoma.

Conclusion: Although Renal Synovial cell sarcoma is rarely diagnosed in kidney but it should be considered in the differential diagnosis of spindle cell tumors affecting the kidney and must be excluded by immunohistochemical studies as it has poor prognosis in the kidney.

Keywords: Renal Synovial cell sarcoma, Bcl2, CD99, Vimentin, EMA

Introduction

Spindle cell neoplasms affecting kidney are many and variable, most of them are malignant may be Sarcomatoid renal cell carcinoma, Adult form of Wilm's tumor, Malignant peripheral nerve sheath, Leiomyosarcoma, Hemangiopericytomarenal and Synovial cell sarcoma. Renal Synovial cell sarcoma is rare and to be diagnosed need panels of immunohistochemical markers to diagnose it. This paper reports a case of Renal Synovial cell sarcoma which is rare in kidney.

Case Presentation

A 32 year old female presented with a history of Left loin pain since one month, The patient has no family history of renal cancer. She reported no haematuria, weight loss or history of malignancy. Findings from physical examination were unremarkable except for left renal mass. The preoperative serum creatinine was 1.1 mg/dl. Abdominal ultrasonography revealed incidental LT Single upper polar renal mass which was confirmed at an abdominal computed tomography (CT) scan.

Abdominal computerized Tomography (CT) revealed a heterogeneous,

well marginated soft tissue mass 8x7 cm arising in the upper pole of left kidney with solid necrotic components and heterogeneous enhancement. There was no evidence of renal vien, inferior vena cava or right arterial thrombosis. No local invasion was identified. There was regional lymphadenopathy was identified. Clinical and Radiological diagnosis was consistent with renal cell carcinoma. The work up for metastasis yielded negative results.

The patient subsequently underwent surgical exploration of the mass with planning for radical nephrectomy with nodal dissection.

At surgical exploration, Pathologic evaluation by intraoperative frozen section was consistent with spindle malignant cell neoplasm. Negative surgical margins were reported for all excised kidney.

In the early postoperative phase, the patient needed intensive care unit for one day for follow up.

Pathologic Findings Gross Pathology

The specimen received weighted 230 g and measured 12x10x4 cm in diameter. Its cut section showed that upper pole of the kidney was replaced completely by grayish tan firm mass with cystic

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areas measuring 6x6x3 cm in diameter. Areas of hemorrhage and necrosis were seen.

Microscopic Pathology and Immunohistochemistry

Histopathological examination revealed the presence of neoplasm composed of solid monomorphic sheets of malignant spindle to oval plump cells showing pleomorphism, hyperchromatism, high nucleocytoplasmic ratio, clear to esinophilic cytoplasm with central vesicular nuclei and prominent nucleoli. Stroma showed rich vascularity with vessels showing hemangiopericytoma like pattern. Four mitosis per ten high power fields were noted. Areas of necrosis and hemorrhage were also observed. Renal capsule, Renal pelvis and ureter were free from infilteration. No lymphovascular invasion could be detected. Nine regional lymph nodes received with the specimen showed reactive hyperplasia (Fig1A).

Overall, the features are consistent with malignant renal spindle cell neoplasm for differential diagnosis: Sarcomatoid renal cell carcinoma, Synovial cell sarcoma, Adult form of Wilm's tumor, Malignant peripheral nerve sheath, Leiomyosarcoma and Hemangiopericytoma.

Immunohistochemical study showed Strong positivity to Bcl2, Vimentin, CD99, Focal positivity to EMA and negativity to CK, CD34, SMA and S100 (Fig1B-I).

Morphologic and immunohistochemical features are compatible with the diagnosis of Synovial cell sarcoma (SCS) of Left kidney.

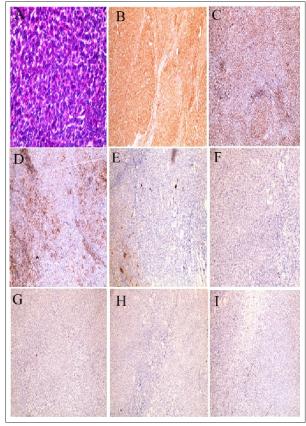


Figure legends

Fig1: Microscopy and immunohistochemistry of the renal mass Fig1A: Monomorphic sheets of malignant spindle to oval plump cells with frequent mitotic figures separated by blood vessels with hemangiopericytoma like pattern

Fig 1B: Bcl2 positive tumor tissue.

Fig 1C: Vimentin positive tumor tissue.

Fig 1D: CD99 positive tumor tissue.

Fig 1E: EMA focally positive tumor tissue

Fig 1F: CK negative tumor tissue.

Fig 1G: SMA negative tumor tissue

Fig 1H: S100 negative tumor tissue

Fig 1I: CD34 negative tumor tissue.

Discussion

Synovial cell sarcoma (SCS) is a clinically and morphologically well defined, but uncommon entity which is known to occur at unusual sites as kidney [1]. Histologically, Synovial cell sarcoma is subclassified into three types as biphasic SCS, monomorphic SCS and poorly differentiated SCS variants. PDSCS comprises approximately 20% of cases with the poorest prognosis [2].

Primary renal sarcomas are rare neoplasm that account about 1% of malignant renal tumors [3]. Prevelance of primary renal Synovial cell sarcoma is rare and comprise 1-3% of all malignant renal neoplasm [4]. The first case of primary renal Synovial cell sarcoma was reported in 1999 and at 2000, Argani et al., published another case of renal SCS [5].

As spindle cell morphology SCS overlaps with multiple spindle cell neoplasms, its differential diagnosis would be Sarcomatoid renal cell carcinoma, Adult form of Wilm's tumor, Malignant peripheral nerve sheath, Leiomyosarcoma and Hemangiopericytoma. All of these differential diagnosis may show immunophenotypic overlap, So these entities may be differentiated by a panel of antibodies. The presence of both mesenchymal and epithelial markers is suggestive of SCS at any site [6].

In our case the tumor tissue shows focal positivity to EMA, Although EMA isn't a specific marker for SCS but its presence in poorly differentiated sarcoma can suggest SCS, also its positivity with other positive markers expression as vimentin, CD99 and Bcl2 and histopathological features confirms the diagnosis of SCS [7]. Positivity for TLE1 is also needed for more confirmation of SCS as it is more specific marker [8].

The simultaneous use of a panel of antibodies as CK, S100, SMA and CD34 excludes the possibility of other differential diagnoses. Negativity for CK excludes both Sarcomatoid renal cell carcinoma and Adult form of Wilm's tumor, also negativity for S100, SMA and CD34 excludes possibility of Malignant peripheral nerve sheath, Leiomyosarcoma and Hemangiopericytoma respectively.

To diagnose the case as it is primary renal SCS, the possibilities of distant metastases as secondary extension of retroperitoneal SCS must be ruled out by clinical and radiological data [9]. Definitive diagnosis of primary renal SCS requires cytogenetic study by either reverse transcriptase polymerase chain reaction or applying FISH testing [10].

Conclusion

Although Renal Synovial cell sarcoma is rarely diagnosed in kidney but it should be considered in the differential diagnosis of spindle cell tumors affecting the kidney and must be excluded by immunohistochemical studies as it has poor prognosis in the kidney.

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