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Primary Renal Synovial Sarcoma - A rare histology

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ABSTRACT

Renal tumours vary from a benign to highly malignant histology. Here we present a 54 yrs male with a heterogeneous large encapsulated cystic lesion in the right kidney upper pole showing areas of solid component and necrosis with no evidence of metastasis or paraneoplastic features.

Post Right Radical Nephrectomy IHC, showed positive reaction for BCL-2, CD99 and vimentin, suggesting Primary Renal Synovial Sarcoma. The patient was treated with ifosfamide based chemotherapy in view of unpredictable prognosis. We present this case for its rarity and good response to treatment and as a suggestion for differential diagnosis of Renal Cell Carcinoma.

Introduction

Renal masses until proven otherwise are considered to be malignant, more so if they are more than 4 cm in size. Malignant tumours other than RCC are rare, accounting for 6–10% cases. Renal sarcomas are much rarer (1–2%) and a majority of them are leiomyosarcoma and liposarcoma. Synovial sarcomas form about 5–10% of all the soft tissue tumours and are mostly seen in the extremities. Renal synovial sarcoma was first reported in 1999 by Faria et al.([1]). Only countable number of case reports are available with this histological subtypes as of today. Present-day confirmatory diagnosis is RT-PCR demonstration of SYT-SSX fusion gene transcript. The present case is an incidentally detected Primary Renal Synovial sarcoma presented like a well-defined RCC.

Case presentation

A 54yr male was incidentally detected with a right renal mass on routine health check-up with features suggestive of RCC.

A contrast-enhanced CT revealed a large mildly enhancing cystic lesion arising from the superoposterolateral aspect of the right kidney measuring about 20 cm × 15 cm, abutting the segment 6,7 of the liver with doubtful infiltration(Fig. 1). There was no evidence of lymph node enlargement. A provisional diagnosis of RCC was made and a metastatic workup initiated. The workup showed no evidence of metastasis or paraneoplastic features. Biochemical investigations were normal.

A radical nephrectomy was planned with possible hepatic resection if found infiltrating. A thoracoabdominal surgical approach was used to reach the mass. The mass was found to be confined to the kidney within the Gerota's fascia and mere indentation of the right lobe of the liver. An open right radical nephrectomy with regional lymphadenectomy was performed.

The histology showed areas of necrosis and haemorrhage with rich vascularity. It showed sheets of round cells with high nucleocytoplasmic ratio with prominent nucleoli. There was no metastasis in the regional lymph nodes, no perineural invasion and the margins of the specimen were free from malignancy(Fig. 2). Differential diagnoses of Blastemal Wilms and Sarcoma were made on histology. A final histological diagnosis was challenging and immunohistochemistry (IHC) was used for further clarity. The IHC evaluation tests showed a positive reaction for BCL-2, CD99, and vimentin (Fig. 3), which suggested the diagnosis of synovial sarcoma.

In view of rare histology and no clear guidelines in the management of this histological subtype,[2] the patient was subjected to adjuvant chemotherapy with Ifosfamide and adriamycin for 6 weeks. Though chemotherapy did not add anything clinically or biochemically measurable, the patient was doing well at 3 months and later lost for follow up.

Discussion

Primary synovial sarcoma of the kidney is the rarest type of renal

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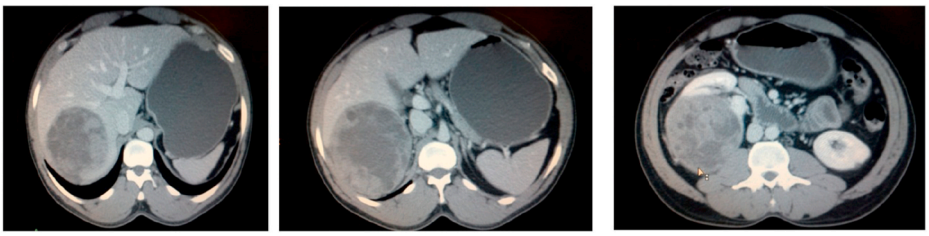

Fig. 1. Contrast-enhanced CT scan of the abdomen, showing a large heterogeneously enhancing mass involving the upper pole and inter-polar region of the right kidney, which appears to infiltrate the right lobe of the liver.

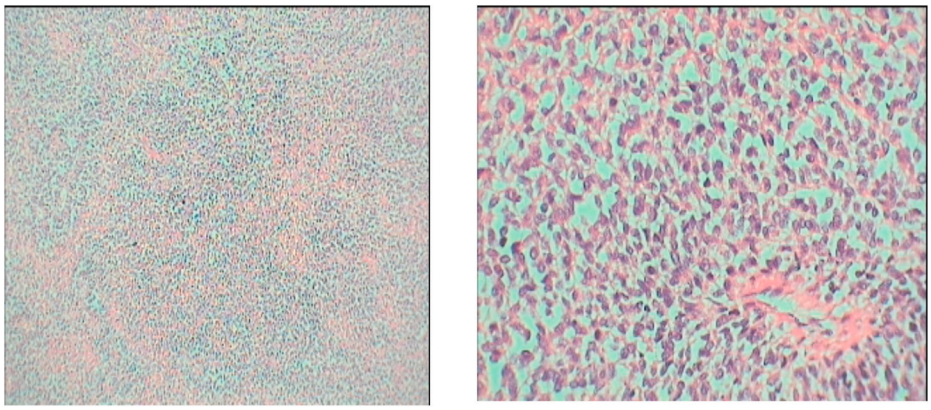

Fig. 2. The low power and high power images respectively, of the tumour showing sheets of round cells with high nucleo-cytoplasmic ratio and prominent nucleoli.

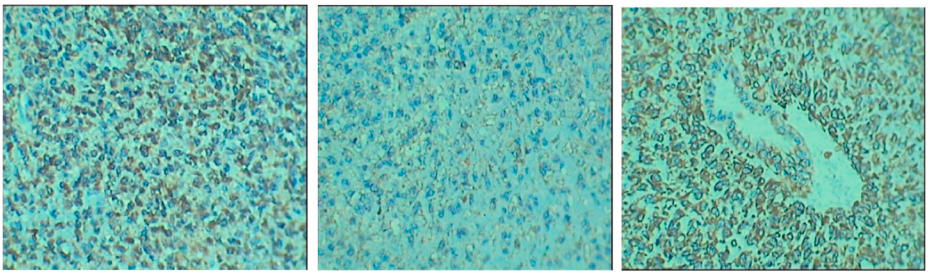

Fig. 3. Three images of the immunohistochemistry of the tumour tissue. The first image on the left-hand side shows the positive reaction to BCL-2, the next image showing a positive reaction to CD99 and the third image showing a positive reaction to Vimentin.

sarcoma, accounting to less than 1% of Renal malignancies. Leiomyosarcoma represents 40–60% of the described renal sarcomas, followed by rhabdomyosarcoma, histiosarcoma, chondrosarcoma and osteosarcoma, liposarcoma, angiosarcoma and hemangiopericytoma. The usual presentations in synovial sarcomas of the kidney that have been reported are well defined large masses with morphology varying from irregular outline to smooth cystic lesions([3]). Generally, it affects young individuals, of both genders, between 20 and 50 years, presenting with a clinical picture that is similar to any renal tumours. Presently, there are no clinical or imaging characteristic to indicate a confirmatory diagnosis of synovial sarcoma([4]). Furthermore, there is no standard treatment protocol for synovial sarcoma to date. In many cases, surgery followed by ifosfamide-and/or adriamycin-based chemotherapy have been used.[5]

Despite its rarity and non-specific presentation, clinicians should consider synovial cell sarcoma in the differential diagnosis of renal masses, especially when histopathology is non-diagnostic. In this case, an adjuvant chemotherapy regimen with ifosfamide and adriamycin have been used in view of an unpredictable clinical course.

Conclusion

Primary Renal Synovial Sarcoma is rare among renal tumours amounting to less than 1% of all the renal tumours. Since a histological diagnosis based on the clinical data and imaging studies is impossible, keeping an open mind is essential for surprising histology as this case.

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