

Primary renal synovial sarcoma- A Case report.

ABSTRACT

Primary Renal Sarcoma is a rare tumor comprising only 1% of all renal tumours. Synovial sarcomas are generally deep-seated tumors arising in the proximity of large joints of adolescents and young adults and account for 5–10% of all soft tissue tumours. Primary synovial sarcoma of kidney is rare and has poor prognosis. It can only be diagnosed by immunohistochemistry. It should be considered as a differential in sarcomatoid and spindle cell tumours. Here we present a case of a 43 year old male who presented to us with complaints of abdominal lump on the right side. Patient underwent exploratory laparotomy with right nephrectomy for renal mass. Histopathology confirmed it to be a Monophasic Synovial Sarcoma.

Introduction

Primary renal synovial sarcoma (PRSS) was initially described by Faria et al. in 1999 which was previously included in *embryonal* sarcomas of the kidney. Despite all the features, the nature of the tumour and natural history, the management of this tumour is still a matter of further investigation. There are only 51 cases of primary renal synovial sarcomas reported till date. Synovial sarcoma (SS), or sarcoma of tissues adjacent to joints, is a rare type of STS, and represent 5 to 10% of all STSs. SS is commonly found in the proximal limb of young adults and has a male predominance. Other unusual sites of occurrence include the head and neck, heart, lungs, and kidneys. Very few reports have tackled this tumor due to its rarity and difficulty to distinguish from other renal pathologies. We present a case of PRSS which was diagnosed by Histopathology.

CASE PRESENTATION

A 43 year old gentleman presented with symptoms of gastric outlet obstruction and abdominal lump on right side since 2 months. Patient gave no history of hematuria, fever, pain. There were no known comorbidities.

USG of the abdomen revealed a large 20 x 18 cm sub-hepatic mass. CECT Abdomen and pelvis revealed the right kidney replaced by a cystic structure measuring 17 x 14 x 20 cm with mild enhancing septa with surrounding fat stranding and prominent vessels. Right ureter is not visualised, gross stomach dilatation due to secondary mass effect.

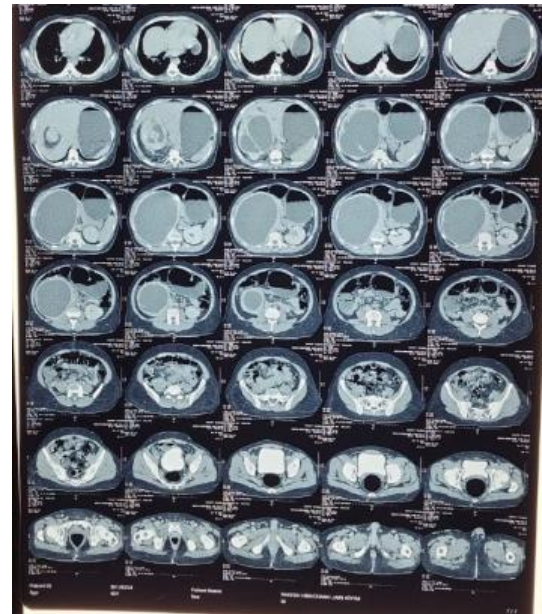


Fig 2 Mass Causing Gastric Outlet Obstruction



Fig 1 CT Image of Large Mass Arising from Renal Parenchyma

Patient underwent an exploratory laparotomy with right nephrectomy and the mass was excised in-toto. Intraoperatively, a large mass was noted arising from the right kidney with adhesion noted to the ascending and transverse colon, tumour was abutting the IVC. Careful adhesiolysis was done, right ureter was transected and the specimen was delivered and sent for HPE.

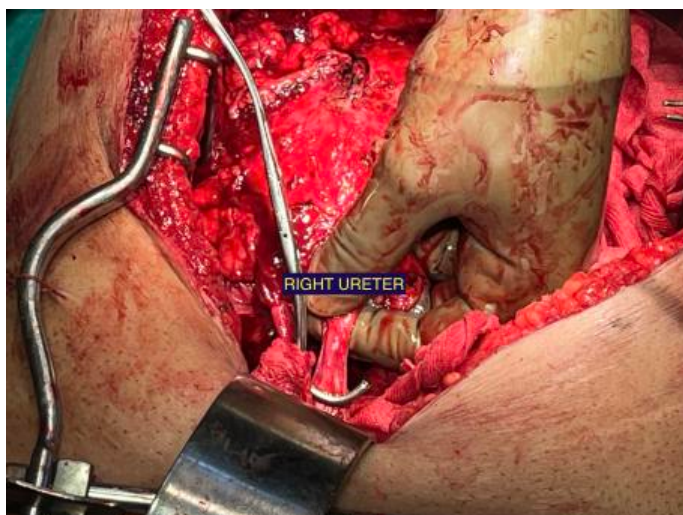


Fig 3 Right Ureter Identified and Transected



Fig 4 Adhesion of Viscera to Mass

Histopathology, revealed the tumour to be a Monophasic Synovial Sarcoma of the right kidney.

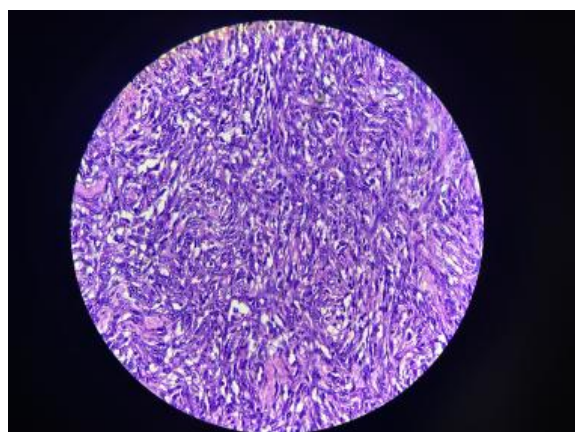


Fig 5 HPE Showing Bundles of spindle cells in clusters (H&E 40x)

DISCUSSION

Primary kidney sarcomas are rare neoplasms that account for 1% of all malignant renal tumors. Leiomyosarcoma is the most frequent type of renal sarcoma, comprising about 40–60% of all cases. Other sarcomas that involve the kidney include rhabdomyosarcoma, malignant fibrous histiocytoma, fibrosarcoma, angiosarcoma, hemangiopericytoma, Liposarcoma and

rarely synovial sarcoma. Synovial sarcomas are commonly seen in the proximity of large joints. They can be observed in unexpected sites, such as thoracic and

abdominal wall, head and neck region, including pharynx and larynx, retroperitoneum, bone, as well as visceral organs, such as lung, pleura, ovary or prostate. Immuno histochemistry and genetic analysis helps in establishing diagnosis of most of the tumors and in case of inadequate histopathology reporting and immunocytochemistry, genetic analysis should be considered.

Despite the limited follow-up data and the small number of cases reported, management of this tumor has already proved to be a difficult challenge. Synovial sarcomas are histomorphologically grouped into 3 types; monophasic synovial sarcoma (MSS), biphasic synovial sarcoma (BSS) and poorly differentiated synovial sarcoma (PDSS) which consists of 20% and has poorer prognosis. The recognition of monophasic fibrous synovial sarcoma and PDSS subtypes is often a diagnostic challenge for pathologists because they may easily be confused with other spindle to round cell sarcomas, especially malignant peripheral nerve sheath tumors, fibrosarcoma, leiomyosarcoma, liposarcoma, and Ewing sarcoma. A study reported tumor-free survival in 90% patients after a 37-month-follow-up with localized soft tissue synovial sarcoma. Other study reported complete remission using a doxorubicin and ifosfamide protocol in a PRSS patient developing metastases in the lung in the fourth month following radical nephrectomy. However, currently, there is no definite consensus regarding the use of chemotherapy for patients with PRSS. More studies are required to develop a definite plan of treatment.

CONCLUSION

PRSS is a rare tumour of kidneys which can be diagnosed on histopatholgy combined with immunohistochemistry and genetic analysis. Surgery is the mainstay of treatment. Radiotherapy can be useful as an adjuvant therapy in presence of local spread. Chemotherapy may be beneficial in distant metastases. However, more studies are required for optimising treatment of this rare tumour for better prognosis.

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