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[1-3]. Despite all the features, the nature of the tumour and natural history, the management of this tumour is still a matter of further investigation[4-5]. Synovial sarcoma (SS), or sarcoma of tissues adjacent to joints, is a rare type of STS, and represent 5 to10% 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 difaiculty to distinguish from other renal pathologies. We present a case of PRSS which was diagnosed by Histopathology.

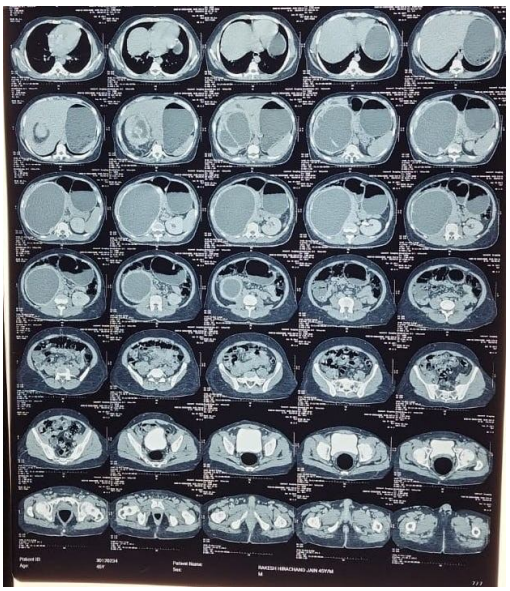

Fig 2 Mass Causing Gastric Outlet Obstruction

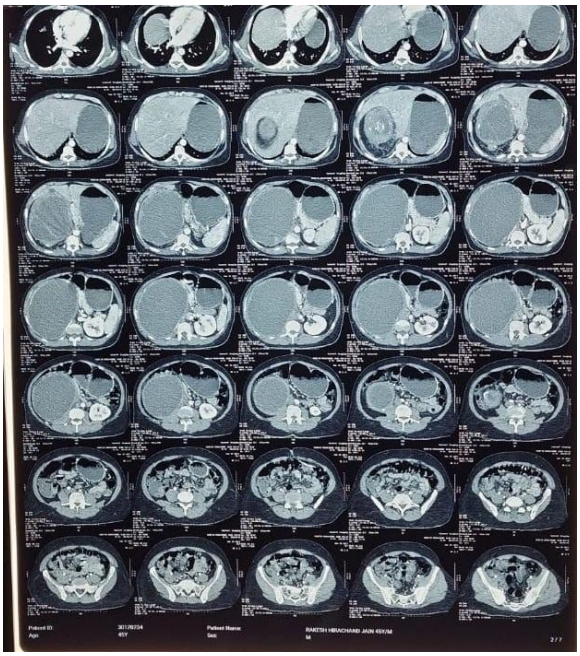

Fig 1 CT Image of Large Mass Arising from Renal Parenchyma

CASE PRESENTATION

A 43 year old gentlemen 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.

Clinical Examination revealed a 25 x 25 cm lump in the right Clank with smooth surface, ballot able, minimal movement with respiration and extending towards right iliac fossa, not crossing the midline.

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. **Metastatic work-up revealed it to be a localised tumour.**

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.

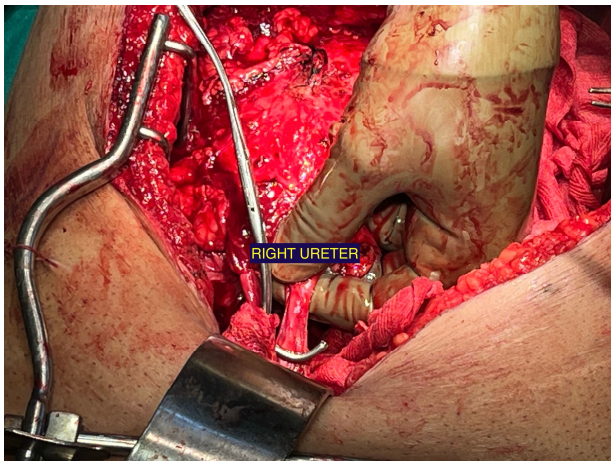

Fig 3 Right Ureter Identified and Transected

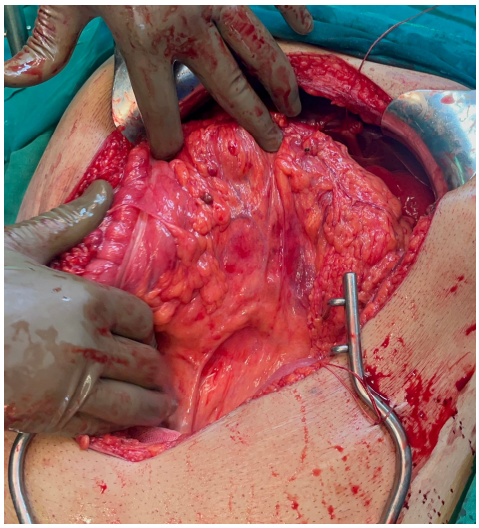

Fig 4 Adhesion of Viscera to Mass

Histopathology , revealed **a tumour composed of spindle cells arranged in storiform pattern, cells showing scanty cytoplasm with oval hyper chromatic nuclei, vascular tumour emboli detected with hemangiopericytomatous vasculature. Large areas of hyalinisation & foci of calciCication are evident.**

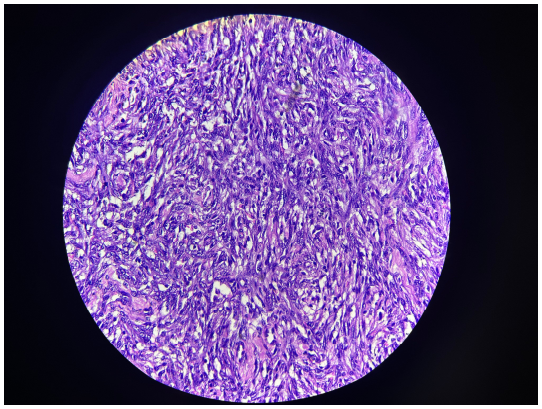

Fig 5 HPE Showing Bundles of spindle cells in clusters (H&E 40x)

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[1]. Other sarcomas that involve the kidney include rhabdomyosarcoma, malignant aibrous histiocytoma,

aibrosarcoma, 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 histocytochemistry 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.

PRSS generally presents in adults between the ages of 15 and 71 years with equal incidence in both the sexes. The clinical symptoms mostly include abdominal pain and hematuria as was seen in our patient. Despite the limited follow-up data and the small number of cases reported, this tumor has already proved to have a generally aggressive behavior.[1,3] Renal synovial sarcomas present as large masses ranging from 1 to 35 cm, usually between 5 and 20 cm in diameter. They are frequently associated with cysts with smooth walls. Cysts are lined by mitotically inactive polygonal eosinophilic cells with apically oriented nuclei (*hobnailed epithelium*). Microscopically, tumors are characterized by mitotically active, monomorphic plump spindle cells with indistinct cell borders growing in short, intersecting fascicles.[3–5]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 monophasic spindle cell variant consists of spindle cells with little or no evidence of epithelial differentiation. MSS appears frequently than the biphasic form, with both types sharing the same clinical, ultrastructural, and molecular features. Unlike the BSS patterns, the recognition of monophasic Cibrous 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, Cibrosarcoma, leiomyosarcoma, liposarcoma, and Ewing sarcoma.[1,4–7]

Synovial sarcomas are characterized by the translocation t(x; 18) (p11.2; q11.2). The breakpoint of this translocation fuses the *SYT* gene from chromosome 18 to one of three homologous genes, *SSX1*, *SSX2*, and *SSX4* on the X chromosome. Coexpression of two forms of SYT messenger RNA, I-SYT and N-SYT, has been described in both normal tissues and synovial sarcomas. Although both fuse with SSX, N-SYT fusion products predominate in normal tissues, while the I-SYT isoform is consistently overexpressed in synovial sarcomas. The SYT-SSX gene is thought to function as an aberrant transcriptional regulator. The nature of the chimeric gene appears to be of prognostic importance, as metastasis-free survival is signiCicantly higher in patients with *SYT-SSX2* compared to *SYT-SSX1* fusion genes. *SYT-SSX1* is associated with biphasic tumors, while *SYT-SSX2* is associated with monophasic tumors that lack glandular epithelial differentiation. However, not all reports support a worse outcome for patients with biphasic as compared to monophasic histology. In addition to these cytogenetic changes, Bcl-2 expression, as assessed by immunohistochemistry, has been reported to be an almost general constitutive alteration of synovial sarcomas 4,6,8. We also conCirmed expression of Bcl-2 protein which was negative, by using both genetic and Western blot analysis. The *Bcl-2* gene expression in a patient with SYT-SSX2 may indicate ineffectiveness of chemoradiation after surgery.[2,7,9] Ultrasound and computed tomography may be one of the means of diagnosis, but may not diferentiate between, renal synovial sarcoma and other sarcomas. It shows only areas of necrosis and haemorrhages which is suggestive of malignancy.[1,3] The treatment protocol of PRSS consists of adjuvant ifosfamide based chemotherapy programs in conjunction with radical nephrectomy. High-dose ifosfamide-based chemotherapy has been used as neoadjuvant and adjuvant therapy in patients with localized or advanced soft tissue synovial sarcomas, and favourable results have been reported.[2,3] Radiotherapy is also effective in patients with Bcl-2 negative expression as it was in our patient.[2,7]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 deainite consensus regarding the use of chemotherapy for patients with PRSS. More studies are required to develop a deainite 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 beneaicial in distant metastases. However, more studies are required for optimising treatment of this rare tumour for better prognosis.

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