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**Case Report**

# Rare case of synovial sarcoma of anterior abdominal wall in a young Indian male

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## ABSTRACT

Synovial Sarcomas (synoviomas) are the fourth most common malignant soft-tissue tumours, and typically develop in para-articular locations of the extremities in close association with joint capsules, tendon sheaths, bursae and fascial structures. Other less common sites include the head and neck, abdominal wall, intra-abdominal cavity, and mediastinum. In this article, an interesting and rare case of a 25-year-old man with left upper abdominal lump is reported which was subsequently diagnosed as biphasic synovial sarcoma (spindle cell variety) of anterior abdominal wall.

**Keywords:** Abdominal wall, Synovial sarcoma

## INTRODUCTION

Synovial sarcoma is a rare soft tissue tumor with a high grade of malignancy developing mostly near large joints. The most common sites of origin are the thigh, knee, ankle, foot, and upper extremities, occurring young children to elderly but the peak incidence is observed in young adults[1] however incidence of sarcoma in adults is <10%[2], Surgery remains the mainstay of treatment along with the radiation, Sarcoma in abdominal wall as a primary is rare.

## CASE REPORT

A 25-year-old laborer from middle part of India had a lump in left lumbar region for 8 years which has gradually increased to present dimensions and had dull aching pain for 2 months. There was no history of fever, dysuria, hematuria or symptoms suggestive of adrenal, spleen or colonic origin. No abdominal operation was done beforehand. Patient had no adverse habits. On examination, vitals were within normal range and had no lymphadenopathy. On per abdomen examination, in left lumbar region a non-tender, 10*6 $cm^4$, ill defined, firm lump having superior margin 4 cms from left costal margin in mid clavicular line and moving with respiration was found. On contracting the abdominal wall muscles the lump became more conspicuous and margins better defined (Figure 1).

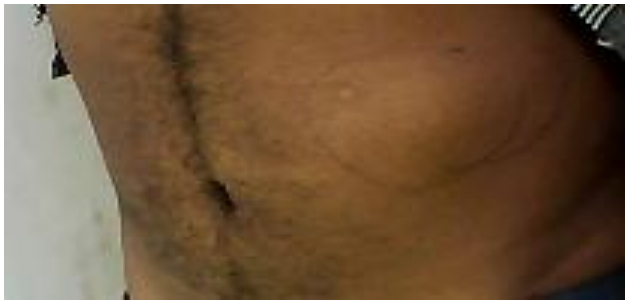

**Figure 1: Clinical presentation of a patient with lump in left lumbar region.**

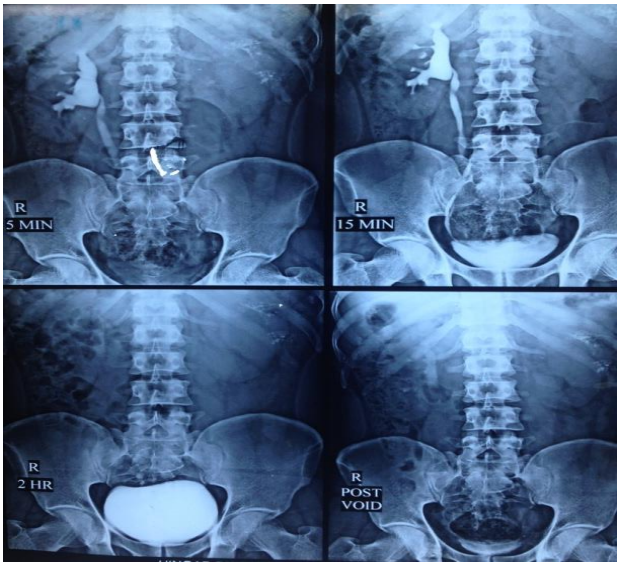


**Figure 2: Absence of contrast media by left kidney in IVP study.**

Laboratory investigations were within normal limits. FNAC of lump revealed moderately pleomorphic loosely cohesive cells arranged in papillary, acinar and diffuse growth patterns with abnormal mitosis but report was equivocal. Ultrasound abdomen showed 9.3*8.9 $cm^4$ sized well defined heterogeneous echotexture lesion with internal solid and cystic areas and calcification noted adjacent to left kidney. USG could not differentiate between intraperitoneal or retroperitoneal origin. It also revealed gross hydro nephrosis with dilated left renal pelvis (AP diameter of renal pelvis 48 mm) and thinned out renal parenchyma indicative of possibility of PUJ obstruction. Intravenous pyelogram revealed absence of contrast excretion on left side and normal excretion on right side (Figure 2).

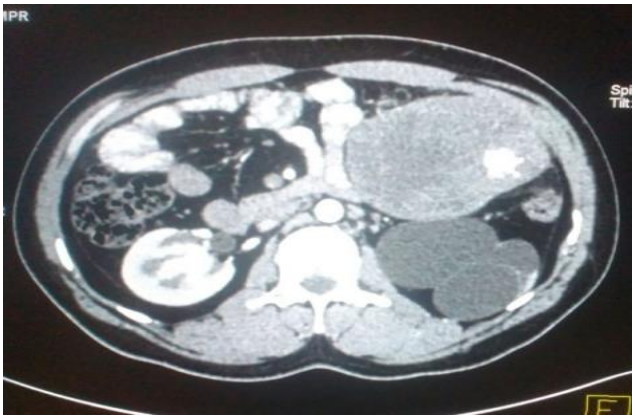

**Figure 3: CT scan view of lump and left hydronephrotic kidney.**

CECT scan of abdomen showed 10 *8.5*10 $cm^5$ soft tissue density mass lesion with internal cystic areas and foci of calcification invading anterior abdominal wall muscles with broad base towards wall, abutting renal pelvis posteriorly and causing displacement of bowel loops medially. Gross hydro nephrosis with dilated renal pelvis on left side is evident (Figure 3 and 4).

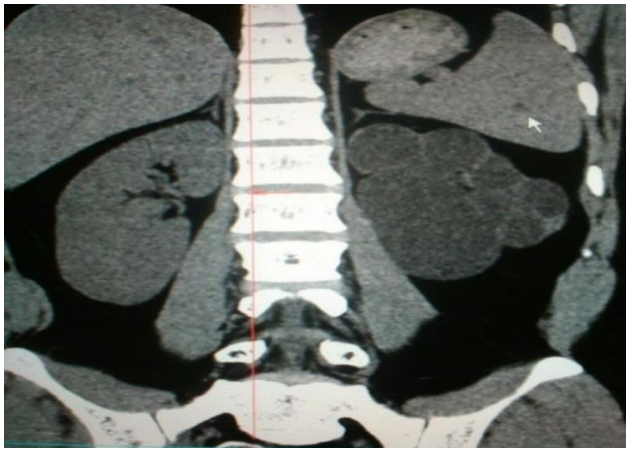

**Figure 4: CT Scan View of lump and left hydronephrotic kidney.**

DJ stenting of left ureter was done and decision of exploratory laparotomy and excision of mass was taken. Peritoneal cavity was accessed via left lumbar incision extending in left subcostal region.

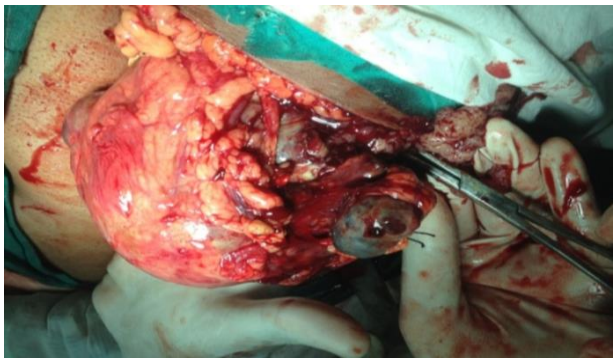

**Figure 5: Intraoperative photograph of broad-based tumor with abdominal wall.**

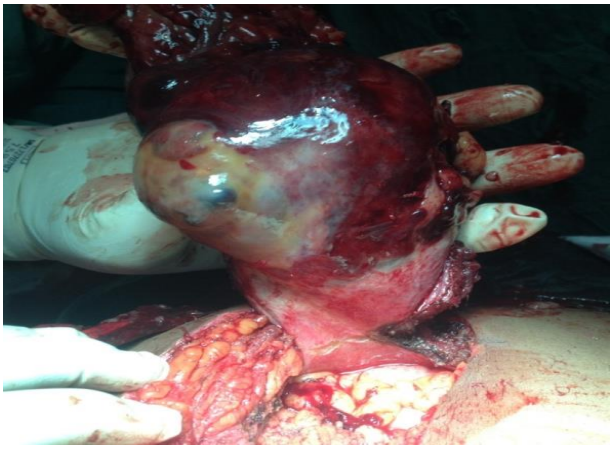

**Figure 6: Intraoperative photograph of encapsulated tumor with smooth surface.**

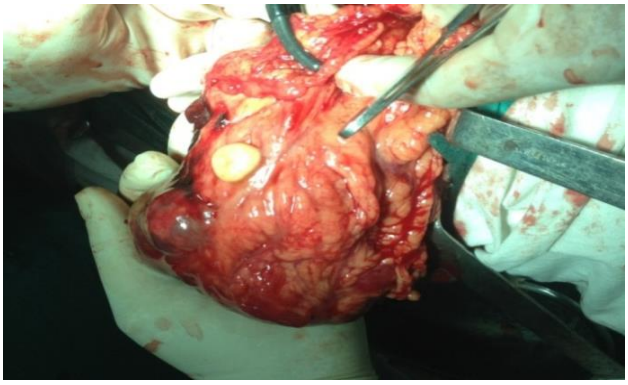

**Figure 7: Intraoperative photograph of cystic areas.**

A mass measuring 12*8*6 $cm^5$ with narrow base was found arising from transverses abdominis and internal oblique muscles with adhesions to omentum. Omental adhesions were released and mass were excised in whole with a surgical margin of 2 cm of normal muscle layer all around. Drain was kept in left paracolic gutter and abdomen was closed in layers without tension and no mesh was kept. On gross examination, mass had a variable consistency (Figure 5, 6, 7). Cut section of mass showed multiple necrotic areas with foci of calcification (Figure 8 and 9).

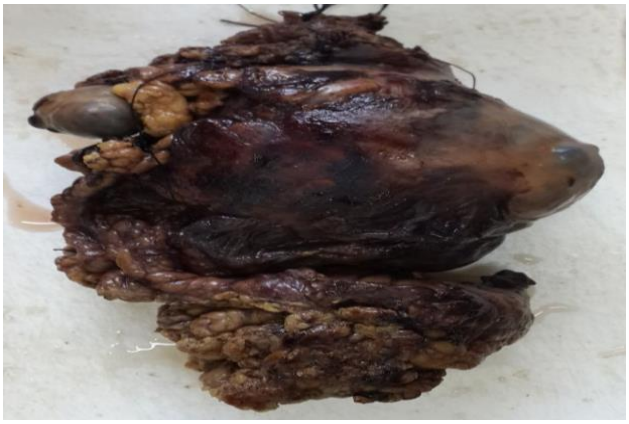

**Figure 8: Cross sectional view of specimen hemorrhagic areas.**

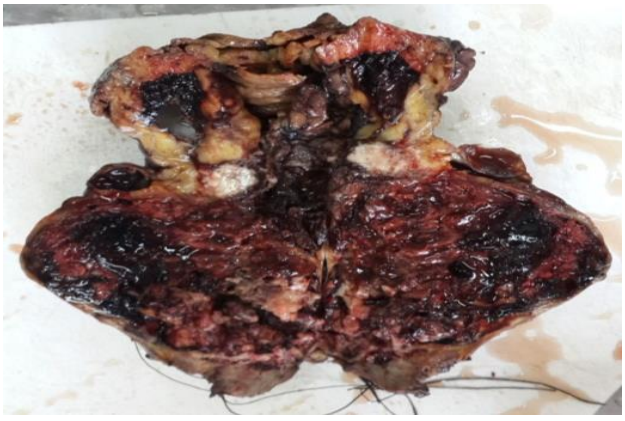

**Figure 9: Specimen areas of calcifications.**

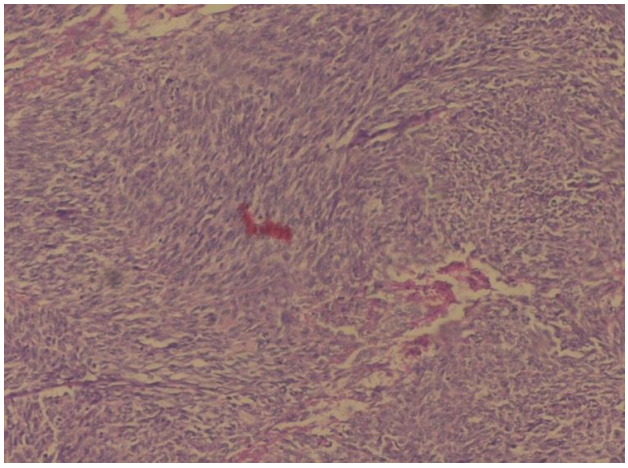

**Figure 10: Microscopic photograph of specimen sheets of spindle cells with epithelial components.**

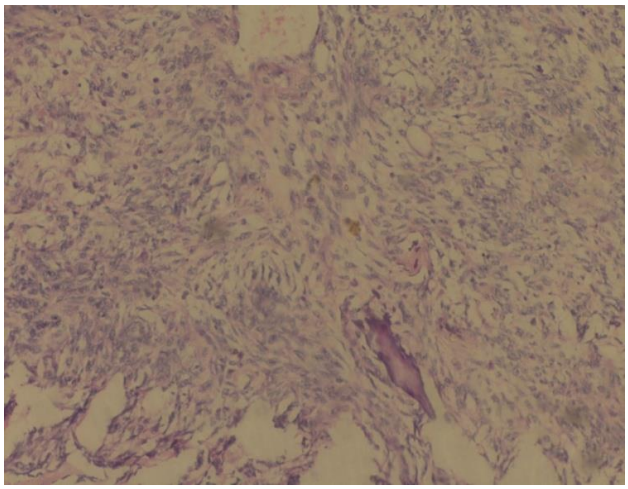

**Figure 11: Microscopic photograph of similar specimen showing spindle cells.**

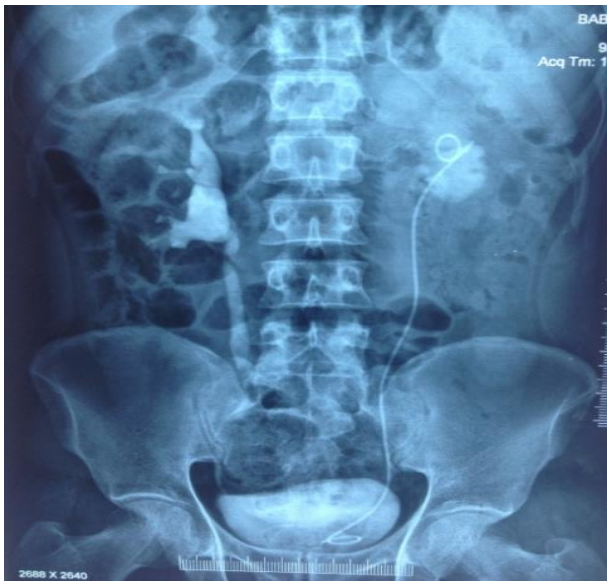

**Figure 12: Post-operative IVP excretion of dye from left kidney.**

Histopathological examination of mass showed oval to spindle cells in a myxoid stoma suggesting a diagnosis of synovial sarcoma (Figure 10,11). Immunohistochemically staining showed positivity to vimentin, S100, AE1 markers. Post op period of patient was uneventful. Postoperative IVP showed excretion of contrast from left kidney in contrast to pre-op findings (Figure 12). DJ stent was removed. After a follow, up of 6 weeks scar was healthy and there were no signs of recurrence (Figure 13). Patient underwent 6 months of chemotherapy and post chemotherapy period was uneventful.

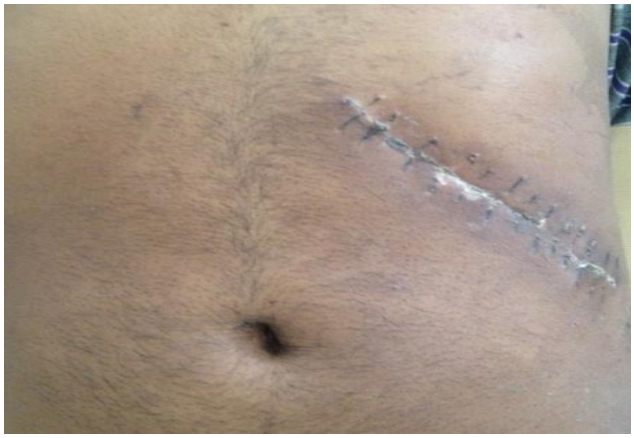

**Figure 13: Post-operative scars.**

## DISCUSSION

Synovial sarcoma is a malignant mesenchymal tumour which occurs in close association with joint capsules, tendon sheaths, bursae and fascial structures. It is generally accepted that synovial sarcoma is derived from primitive mesenchymal cells, not synovial cells. Synovial sarcoma predominantly occurs in the extremities, head, neck and trunk. It frequently arises in the 2nd to 5th decades. Other rare sites of occurrence are neck (retropharyngeal area), anterior abdominal wall, abdominal cavity, retroperitoneum, mediastinum, blood vessels and nerves.[5] Abdominal wall synovial sarcomas are quite rare with only 44 cases having been reported in literature between 1950 and 2005.[3]

Cytogenetic studies show 90% of cases show translocation of chromosome t (X; 18) (p11.2; q11.2) and fusion of SYT gene on chromosome 18 with either SSX1 (67% of cases) or SSX2 (33% of cases) on chromosome X, though the prognostic significance between two fusion types remains controversial.[4] High degree of association is seen with hereditary syndromes and carcinogens. In 50 % of cases x rays may show equivocal findings. Synovial sarcoma predominantly present on X-ray films as round or oval, lobulated swellings or masses of moderate density, without involvement of bony structures. In cases with associated bone erosion, often an indolent non-aggressive appearance is reported. Aggressive bone invasion and destruction is less common, occurring in approximately 5%. Calcification indicates good prognosis and is present in up to 30% of cases on radiography as evident in this case. Computed tomography usually presents with a heterogeneous soft tissue mass with attenuation similar or slightly less than that of muscle. Areas of necrosis or haemorrhage presenting as lower density areas are common. Contrast enhanced CT shows heterogeneous enhancement in 89%-100% of cases. MRI is the optimal radiological modality for assessing the extent and characteristics of synovial sarcomas.[3] Histopathology classification of tumour is of 3 types namely biphasic, monophasic and poorly differentiated forms. Biphasic has both sarcomatous and epithelial components. Monophasic variety has only sarcomatous component in majority of cases. Pure forms of epithelial monophasic components are rare. The poorly differentiated forms are rare but increasingly recognised and are characterised by greater degree of atypia and mitotic activity and carry a poor prognosis.[5] The immunohistochemically panel for confirming the diagnosis of synovial sarcoma consists of cytokeratin AE1/AE3, keratin 7, 14, 19, desmin, vimentin, S 100 (occasional).[3]

National Comprehensive Cancer Network (NCCN) guidelines recommend total surgical resection with at least a 1 cm margin or intact fascial margins, with preoperative or postoperative radiation and anthracycline or an alkylating agent-based chemotherapy.[6] Five years survival for intermediate to high grade sarcoma is 36-76%. Local recurrence is seen in 30-050% of cases and is linked to inadequate margins and aggressive histopathology.[7] Various adverse prognostic features in Synovial Sarcoma include metastasis, invasiveness, positive surgical margins, high histological grade, and poor differentiation. Good prognostic factors still controversial and debatable include SYT-SSX2 gene fusion, monophasic Synovial Sarcoma, size less than 5 cm, less than 15 years of age, and distal extremity location.[3,8,9]

## CONCLUSION

Synovial sarcomas although a rare entity can occur in anterior abdominal wall and can alter the anatomy and physiology of other intra-abdominal organs owing to their size i.e. kidney in this case.

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