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Synovial Sarcomas Presenting as Abdomino-Pelvic Masses: Case Series

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Abstract: ***Synovial sarcoma is a rare high grade malignant sarcoma of mesenchymal origin1 with an incidence of 8-10% of all soft tissue sarcomas2. It is the fourth most common soft tissue sarcoma3. Although predominantly seen in adults less than 30 years but can occur at any age4 with a male predominance5.95% of synovial sarcomas occur in the extremities in near the large joints, 5-15% synovial sarcomas occur in head & neck, abdominal wall, retroperitoneum and mediastinum6-9. We present to you 2 rare cases of abdominal and pelvic synovial sarcomas.***

Keywords: Synovial Sarcomas, Abdomino, Pelvic Masses

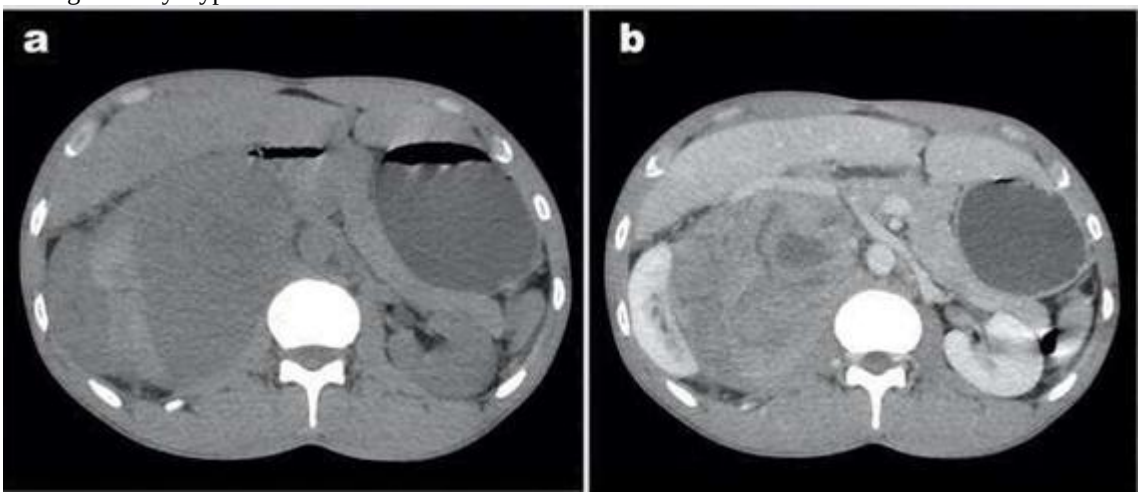
1. Case 1

A 25 year old male patient presented to IMS & SUM hospital OPD with chief complaints of abdominal pain and abdominal distention for 4 months. The pain was dull in nature with a gradual onset and not relieved on medications. There was no history of trauma, fever, vomiting, hematemesis, loose stools or Malena. On examination, the vitals were within normal limits (BP 126/80, PR 74/min). Routine CBC showed normocytic normochromic smear, RFTs were normal, LFTs were normal. On physical examination, the right hypochondrium was tender to palpation with a palpable right hypochondriac mass. Ultrasound examination was advised.

On ultrasound examination a large heterogenous lesion showing areas of hemorrhage and necrosis on doppler imaging with well-defined margins seen in right retroperitoneal region causing scalloping of margins of right lobe of liver was noted. The lesion was separate from right kidney while displacing the right kidney inferiorly and seemed to be adherent to infrahepatic IVC causing compression and displacement of IVC, right renal vein. Working differentials of high grade retroperitoneal sarcoma, unilateral adrenal hemorrhage was considered and advised for a CECT abdomen for further evaluation.

On CT examination, a large circumscribed oval shaped lesion with well-defined margins measuring 16 x 13x 12 cm seen in the right suprarenal area extending to the midline which is heterogeneously hypodense with internal areas of hyperdensity (HU 55-70) suggestive of areas of hemorrhage with no areas of calcifications or fatty attenuation on NCCT. On post contrast imaging the lesion showed heterogenous enhancement with significant central areas of non-enhancement suggestive of necrotic/cystic areas. The lesion was causing scalloping of posterior margin of right lobe of liver, compression of gall bladder, displacement of right kidney inferolaterally, displacement of head and uncinate process of pancreas anteriorly, displacement of celiac artery medially, compression and displacement of renal vein, renal artery, IVC anteriorly with angle of contact greater than 180° with IVC and renal vein at the level of renal veins. Right adrenal gland was not separately visualized from the lesion. There was subtle loss of interface with the right psoas muscle at the level of L1, L2 vertebral bodies. There was no evidence of intralesional vascularity, feeding vessels, retroperitoneal lymphadenopathy on postcontrast imaging. The visualized lumbar and dorsal vertebral bodies appear normal. Screening thorax appeared normal with no lesions. Working differential diagnosis of high grade soft tissue sarcoma, adrenal mass with hemorrhage was considered. Tru-cut biopsy was done from the lesions and 6 cores of 2 cm were obtained from the lesion.

Histopathological examination showed monomorphic spindle cells without epithelial differentiation; suggested as spindle cell sarcoma. On Immunohistochemistry the specimen was positive for vimentin, CD56, STA T6, TLE1 and negative for S100, DOG1, SMA, Desmin, CD117, MYOD1; suggested Synovial Sarcoma.



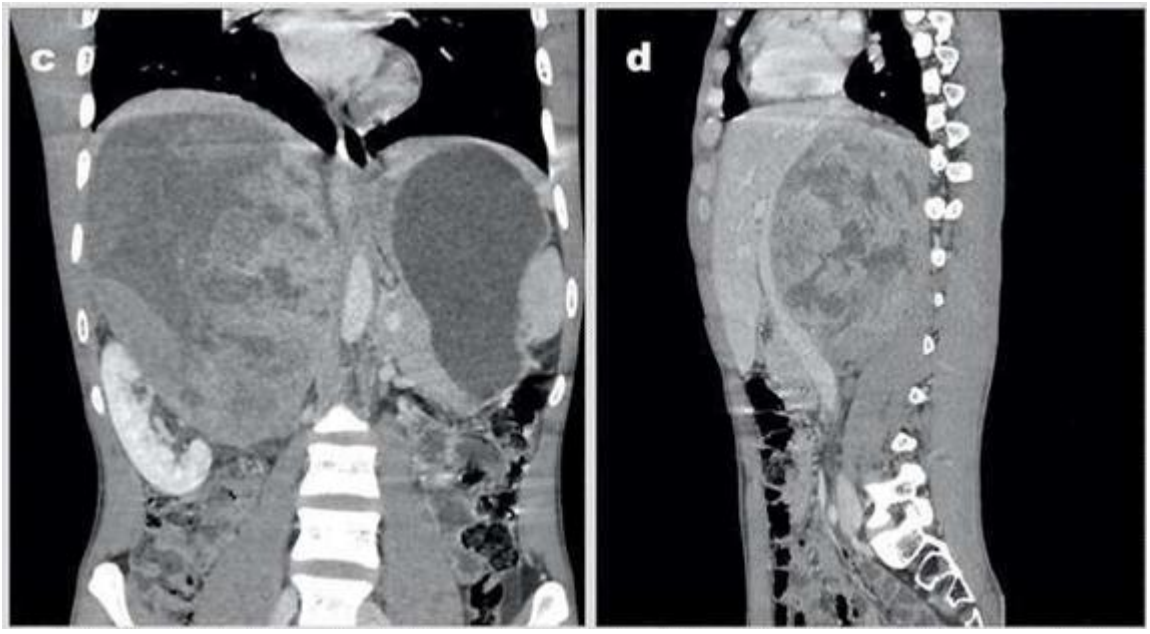


Figure 1: (A) NCCT axial image showing large hypodense lesion with hemorrhagic hypodense with hetrogenous areas of enhancement and central necrotic or cystic areas. (C) Coronal delayed post contrast image showing its suprarenal location. (D)Sagittal delayed post contrast post contrast image showing loss of interface with psoas muscle, anterior displacement of IVC

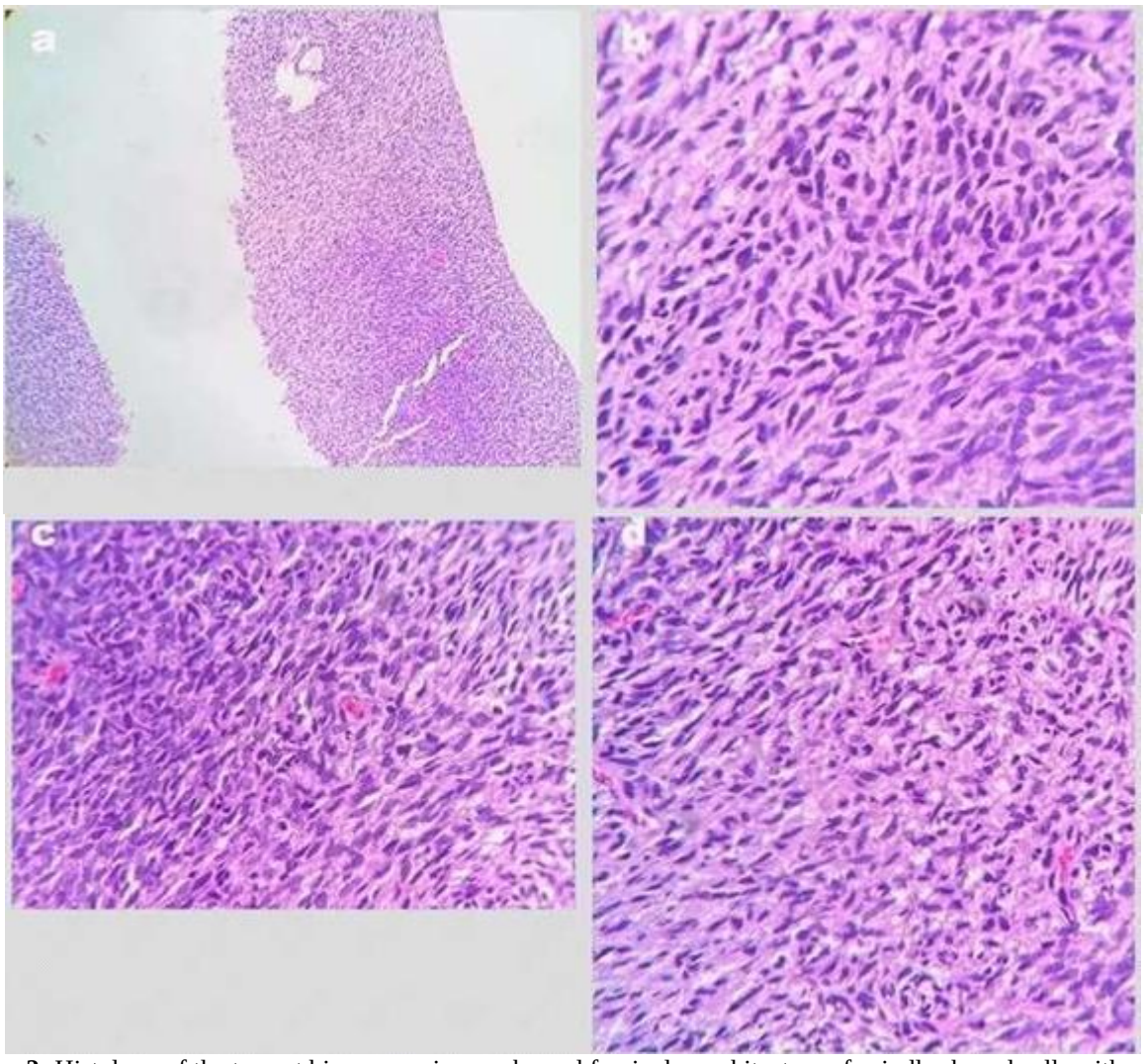


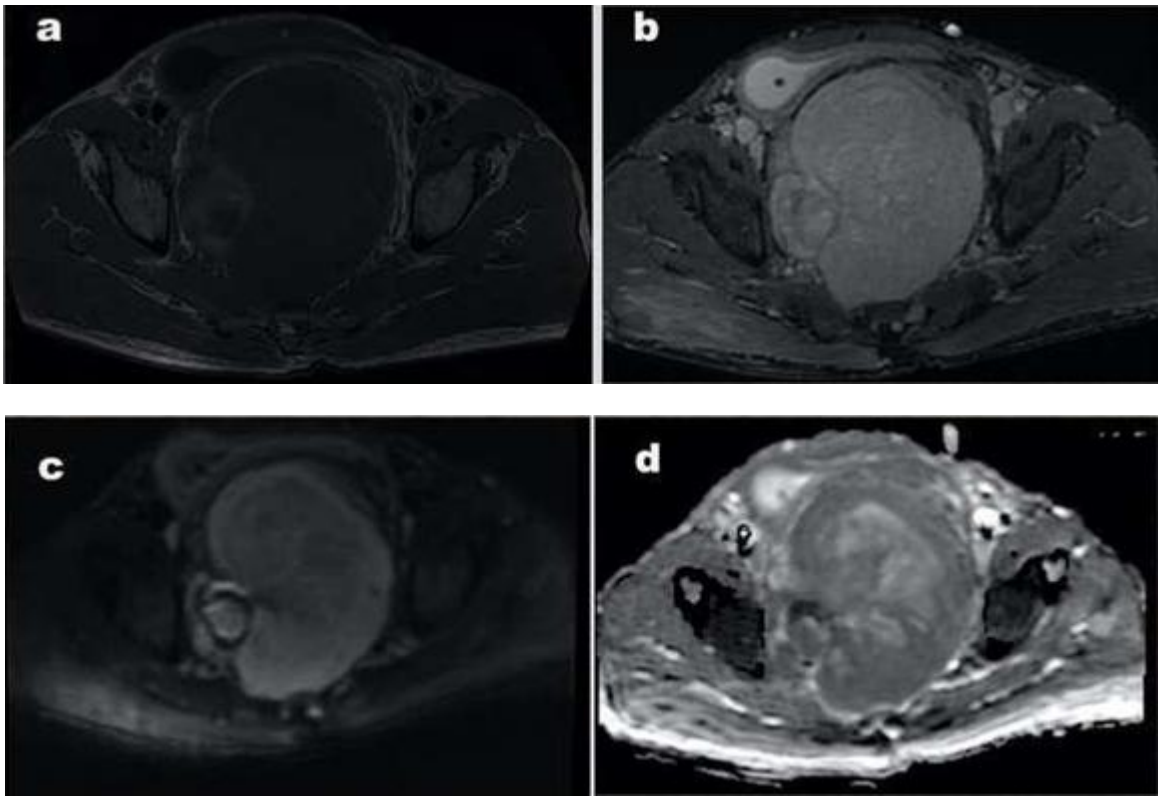
Figure 2: Histology of the tru-cut biopsy specimens showed fascicular architecture of spindle shaped cells with scanty amphophilic cytoplasm

2. Case 2

A 43 year old male patient presented to IMS & SUM hospital OPD with chief complaints of chronic constipation and abdominal distention for 6 months. The constipation was not relieved with regular medications. On examination, the vitals were within normal limits (BP 132/80, PR 76/min). Routine CBC showed normocytic normochromic smear, RFTs were normal, LFTs were normal. To rule out any organic pathology, MRI pelvis was advised as spatial resolution of MRI is better for pelvis imaging.

On MR examination, a large lobulated mixed signal intensity lesion showing hypo-hyperintensity on T1W images, hyperintensity on T2W images, diffusion restriction on diffusion weighted images, heterogeneous enhancement with areas of necrosis measuring x 11 x 16 cm seen in the presacral space extending into the mesorectal fat. It was causing compression & anterior displacement of the urinary bladder, anterolateral displacement of the rectum, rectosigmoid with maintained mesorectal fat plane. The fat plane with prostate was maintained. A nodular bulge of the lesion was seen in the right pelvic side wall muscle layer. There was no evidence of intralesional vascularity, feeding vessels, lymphadenopathy on postcontrast imaging. The visualized lumbar and dorsal vertebral bodies appear normal. Screening thorax appeared normal with no lesions. Working differential diagnosis of high grade soft tissue sarcoma was considered. Tru-cut biopsy was done from the lesions and 6 cores of 2 cm were obtained from the lesion.

Histopathological examination showed monomorphic spindle cells without epithelial differentiation and pleomorphism, hyperchromatism, increased mitosis with moderate amount of amphophilic cytoplasm; suggested as spindle cell sarcoma. On Immunohistochemistry the specimen was positive for vimentin, STAT6, TLE1, PanCK and negative for S100, DOG1, SMA, Desmin, CD68, MYOD1; suggested Synovial Sarcoma.



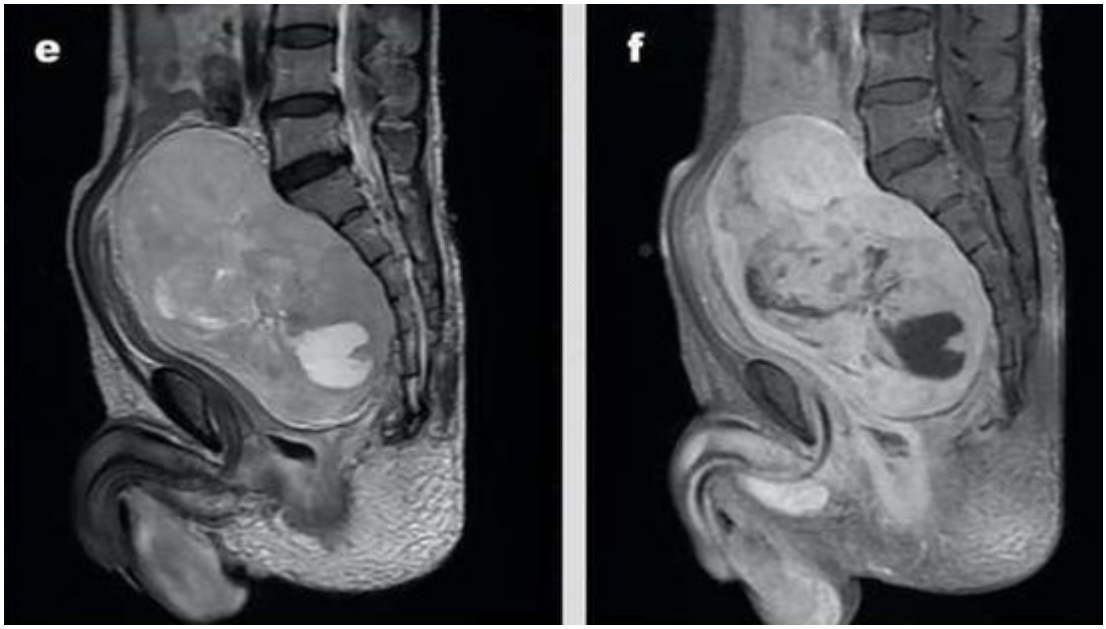


Figure 3: (a) Axial T1W (b) Axial T2W (C, D) Axial Diffusion weighted images DW1 & ADC (E) Sagittal T2W (F) Sagittal Post Contrast T1W images showing large heterogeneously enhancing lobulated lesion with areas of necrosis in presacral space showing diffusion restriction and causing compression and displacement of urinary bladder, rectum & rectosignoid with maintained flat plane of UB, prostate & mesorectal fascia

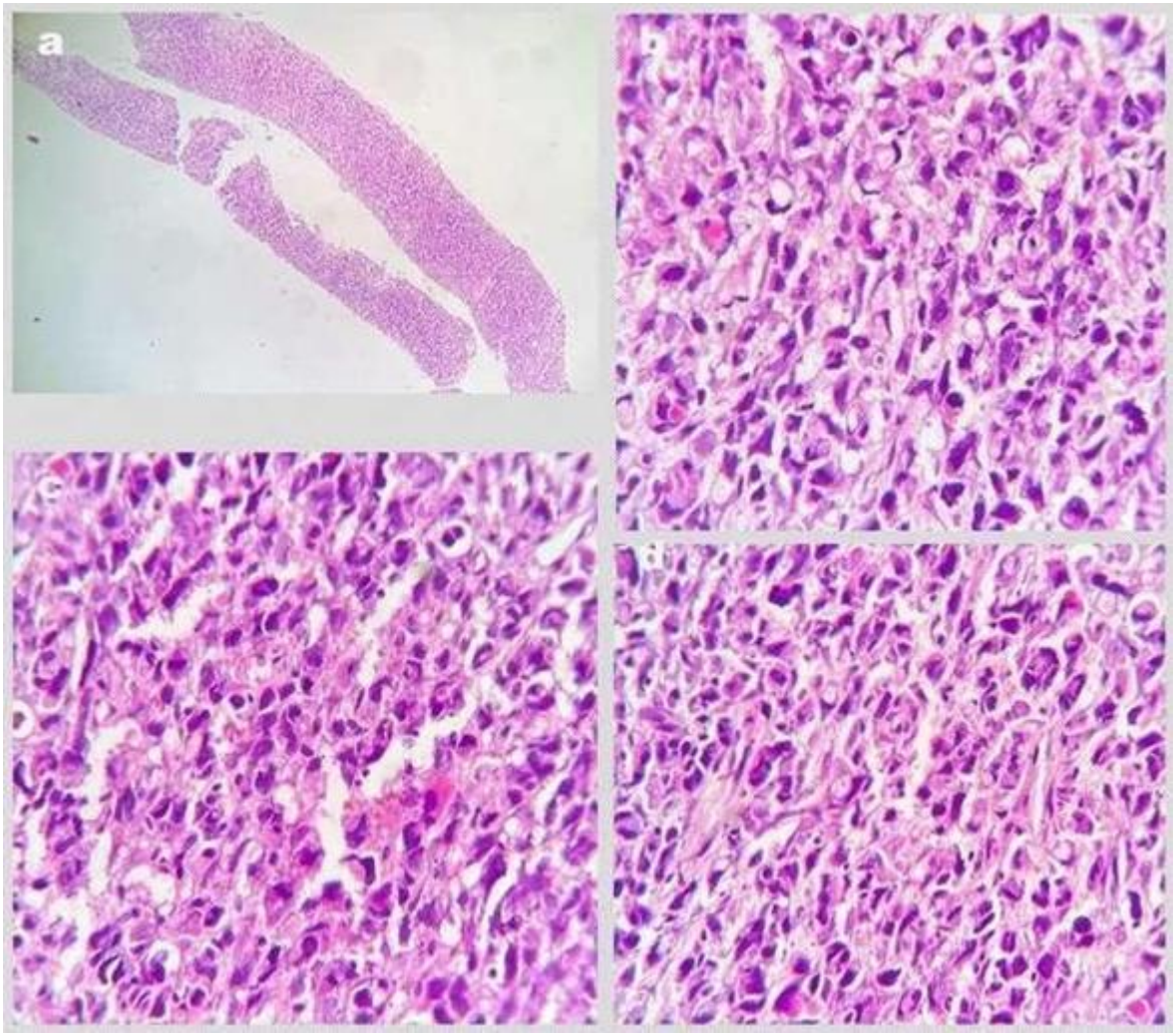


Figure 4: Histology of the tru- cut biopsy specimens showed fascicular architexture of of spindle shaped cells with scanty amphophilic cytoplasm, hyperchromatism, increased mitosis

3. Discussion

Synovial sarcoma is a rare high grade malignant sarcoma of mesenchymal origin1 with an incidence of 8-10% of all soft tissue sarcomas[2]. It is the fourth most common soft tissue sarcoma[3]. Although predominantly seen in adults less than 30 years but can occur at any age 4 with a male predominance 5.95% of synovial sarcomas occur in the extremities in near the large joints, 5-15% synovial sarcomas occur in head & neck, abdominal wall, retroperitoneum and mediastinum[6-9]. Synovial sarcomas are likely originating from normal synovium, primitive pluripotent mesenchyme.

On imaging, synovial sarcoma is indistinguishable from other soft tissue sarcomas and often misdiagnosed as benign due to its well circumscribed appearance, slow growth potential10. On CT imaging synovial sarcomas are large (>10 cm) heterogeneously hypodense lesions with areas of haemorrhage, necrosis, cyst formation. About 30% of synovial sarcomas have intratumoral extensive calcifications [8, 11]. Calcifications strongly suggest synovial sarcoma but our case didn't not show any intratumoral calcifications. On post contrast imaging there is hypoenhancement of the lesion with peripheral subtle enhancement. As there are no distinct imaging findings differentiating synovial sarcoma from other soft tissue sarcomas, the differential of synovial sarcoma should always be in the differential in case of retroperitoneal soft tissue lesion in an young adult12. Diagnosis is usually conferred by histopathology, immunohistochemistry, molecular and cytogenetics.

Histologically, can show monophasic (monomorphic spindle cells only), biphasic (monomorphic spindle cells and epithelial cells) and poorly differentiated morphologies. However clinical prognosis of monophasic not different from biphasic morphology except for poorly differentiated type[5].

Synovial sarcomas are very aggressive lesions with grave prognosis.70% mortality rate within 3 years from diagnosis5. Poor prognostic factors include lesions greater than 5 cm at diagnosis, older patients, proximity to critical structures, poorly differentiated morphological type, advanced clinical stage, high mitotic activity, tumor necrosis. Favourable prognostic factors include smaller lesions, younger (<15 years), calcifications[5, 12].

Treatment include complete wide resection of the lesion but due to its large size complete resection with negative margins is difficult to achieve and associated with high recurrences. So adjuvant chemotherapy, radiotherapy are indicated but benefits are yet to be ascertained[13].

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