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Retroperitoneal Synovial Sarcome : A Rare Presentation

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ABSTRACT

Synovial cell sarcoma is the 4th most common type of soft tissue tumors in adolescents and young patients, with approximately 1 in 3 cases diagnosed in 1 million patients.Retroperitoneal synovial sarcoma usually appears as a nonspecific soft tissue mass that does not have specific imaging features differentiating it from other mesenchymal tumors. However general radiologic findings and anatomic location of the tumor may help the diagnosis. In addition, synovial sarcoma should be included in the differential diagnosis of retroperitoneal soft tissue mass detected in young adults. A 44-year-old Male with Retroperitoneal Synovial Sarcoma is presented.

Keywords: Right Iliac Fossa, Retroperitoneal Neoplasms, Sarcoma, Synovial, Neoplasm Staging, Sarcoma Synovial.

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INTRODUCTION

The term synovial sarcoma refers to the morphology that resembles developing synovium [1]. Eighty five to 95% of synovial sarcomas occur in the extremities near the large joints. Only 5-15% of synovial sarcomas affect the head and neck, mediastinum, abdominal wall, and retroperitoneum [1-4]. Primary retroperitoneal synovial sarcoma is extremely rare and has poor prognosis [5].

Case History

A 44year male presented with complaints of colicky, intermittent, progressing left lower abdominal pain for 15 days and sudden painful lump in left lower abdomen for 10 days. On Examination there was a smooth, non-mobile, ill-defined mass of 6x5 cm size, firm to hard consistency in left iliac fossa extending to the suprapubic region.

Radiology

Ultrasound of the abdomen revealed a Paravertebral retroperitoneal mass in the Left lower abdomen of size 5.6 x 4.3 cm. Colonoscopy was done and it was found to be normal till the transverse colon. MRI of the abdomen a lesion of size 9 x 4.6 x 6.3 cm in the Left Iliac Fossa; between L5-S1 vertebral bodies and the Left psoas muscle, extending along left common iliac arteries with Left common iliac vein compression.

Try-Cut Biopsy

Try-cut biopsy was done under Local Anesthesia and ultrasound guidance suggested Papillary malignant mesothelioma, Papillary serous tumor of mesothelioma or Synovial sarcoma.

Surgery

At operation, a single friable tumor overlying the L4-L5 vertebra & abutting the left external, left internal and common iliac vessels without any gross infiltration into the surrounding structures (Fig. 1) On cut section the tumor had a greyish pink appearance with areas of hemorrhage (Fig. 2). The tumor was completely excised with minimal tumor spillage and sent for Histopathological Examination.
The postoperative period was uneventful.

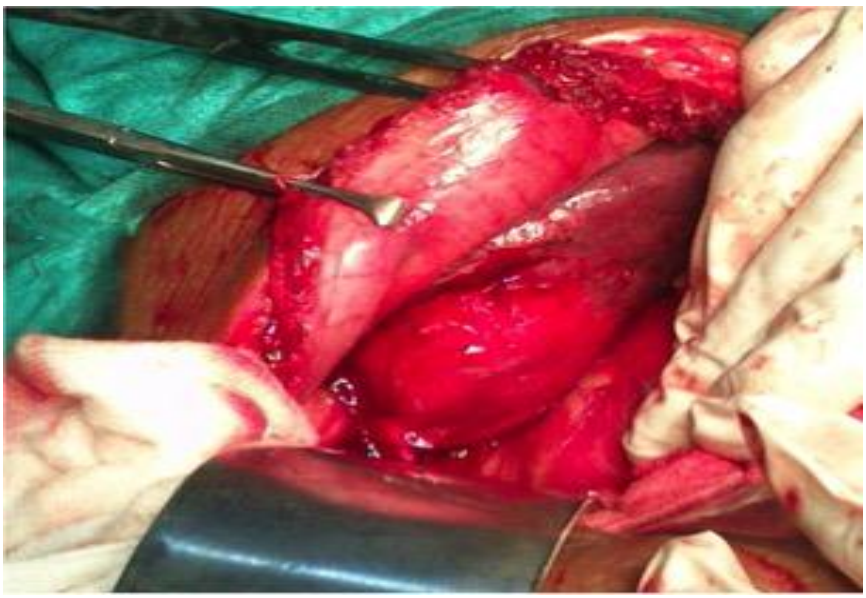

Figure 1

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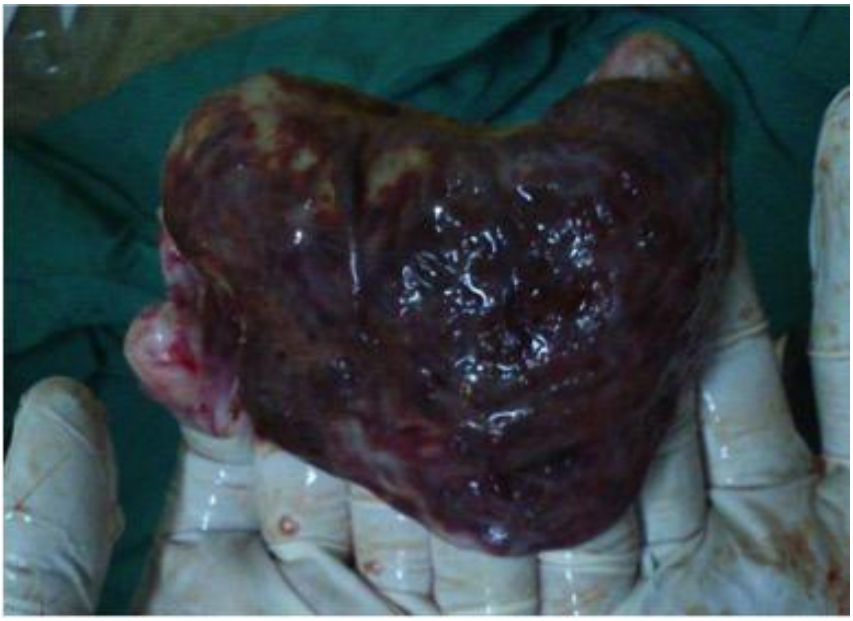

Figure 2

Final Histopathology

High grade Sarcoma with Biphasic Pattern.

Immunohistochemistry

Biphasic Synovial Sarcoma.
Tumor Markers PANCK, Vimentin, EMA, CD99 and BCL2 were Positive.

The patient is currently undergoing Chemotherapy and planned for further Radiotherapy. Follow-up is planned for 5 years.

DISCUSSION

The nature of differentiation in synovial sarcoma is a controversial topic. Potential origins of the synovial sarcoma include normal synovium, primitive pluripotent mesenchyme and arthrogenous mesenchyma. Usually synovial sarcoma occurs in the vicinity of the joint capsules most commonest being the Knee, bursae, and tendon sheaths.

But the occurrences of the tumor in various extra articular sites such as the mediastinum, abdominal wall, retroperitoneum, intraperitoneum, and esophagus have been reported [2-4]. Synovial sarcoma is most prevalent in adolescents and young adults between 15 and 40 years of age. A primary retroperitoneal sarcoma has been defined as a tumor arising in the retroperitoneal space with an origin of mesodermal structures exclusive of bony, renal, visceral, adrenal, and pancreatic tissues [5].

Retroperitoneal synovial sarcoma usually appears as a nonspecific soft tissue mass that does not have specific imaging features differentiating it from other mesenchymal tumors [2, 3, 6]. However radiologic findings and anatomic location of the tumor may help the diagnosis. In addition, synovial sarcoma should be considered when retroperitoneal soft tissue mass is found in young adults. On CT, these tumors are hypo-dense and may show an irregular enhancement in the periphery with a poor enhancement in the central area, reflecting the necrotic, cystic, and hemorrhagic changes [6]. In the region of 30% of cases show intra-tumoral calcification and extensive calcification suggest a favorable prognosis [3, 6]. CT is still recommended as the best imaging method for assessing the local extent of the primary tumor and is a useful tool in the planning of appropriate therapy as well as evaluation of tumor response to ongoing treatment [6].

Histologically there are two types of synovial sarcomas: biphasic and monophasic. Biphasic type is an admixture of epithelial cells and spindle cells. Monophasic type is composed of either only epithelial cells or spindle cells [1-4]. Both types have a mortality rate of 40%. The known poor prognostic factors are frequent mitotic figures (more than 10 or 15 mitoses per 10 HPF) and extensive tumor necrosis, and least favorable for the poorly differentiated (small cell) type. Favorable factors are young age of the patient (15 yearsr or younger) and tumor size smaller than 5 cm, and distal rather than proximal location in the extremities [7].

Surgical ablation remains the mainstay of management of retroperitoneal sarcomas, but complete resection rate of is approximately 50% [5]. The recurrence rate ranged from 28% to 36% even with adequate surgical and adjunctive therapies [3].

CONCLUSIONS

Retroperitoneal presentation of Synovial Sarcoma is very rare and Surgical Excision followed by Chemotherapy and Radiotherapy is the definitive treatment but it is still associated with a high mortality and poor prognosis.

We report this case due to its rarity

Ethical Statement

We warrant that the patient's rights and confidentiality have been well protected in all aspects and he consented to the study described in the Work. All relevant ethical safeguards have been met in relation to patient protection.

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