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Giant Abdominal Synovial Sarcoma: A Case Report

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Abstract
We report in this article the case of a 29-year-old man with no prior medical history who was diagnosed with a giant abdominal synovial sarcoma, treated with initial chemotherapy in preparation for surgery. This article presents a rare sarcoma entity with poor prognosis in young patients requiring early management.

Keywords:*synovial sarcoma, sarcoma, surgery, radiotherapy, chemotherapy.*

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I. Introduction

Synovial sarcoma is a rare type of soft tissue sarcoma that affects young adult males. Its diagnosis is based on imaging, histology, and molecular biology. Treatment is primarily surgical, associated with radiochemotherapy. The prognosis remains poor, burdened by a high rate of local recurrence and metastasis.[1]

II. Clinical Observation

This is a 29-year-old patient with no prior medical history. The onset of symptoms dates back to 6 months with the occurrence of L5 sciatica, treated with anti-inflammatory drugs and analgesics. The patient reports no sphincter, urological, or digestive disorders, with the condition developing in a context of preserved general condition and absence of fever. Clinical examination finds the patient in good general condition, with a deep left abdominal mass, tense, irregular, and poorly delimited. Biology results are normal. An abdomino-pelvic CT scan reveals a left retroperitoneal neoplastic process invading the homolateral paravertebral soft tissues (fig 1,2)

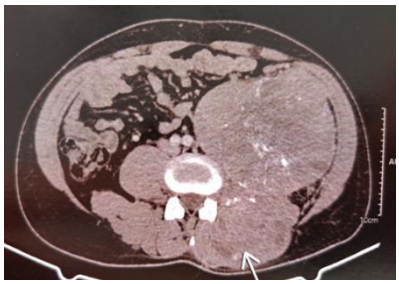

Figure 1

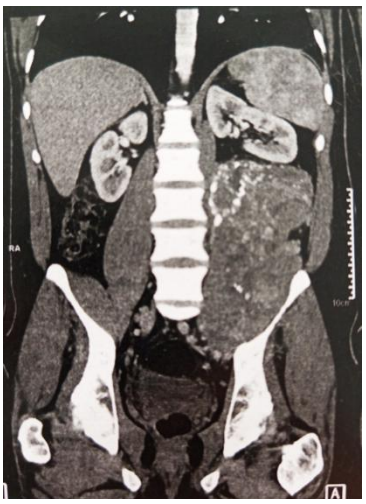

Figure 2

An abdomino-pelvic MRI details a voluminous left retroperitoneal process locally advanced (fig 3,4).

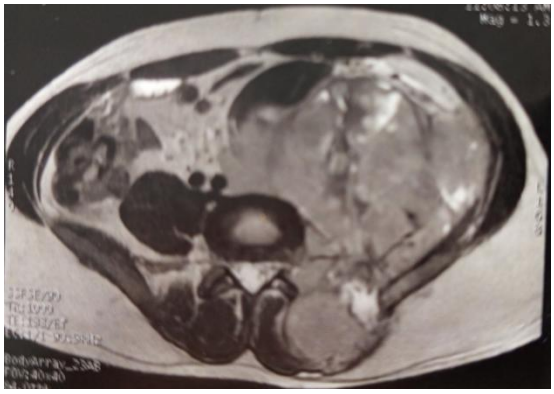

Figure 3

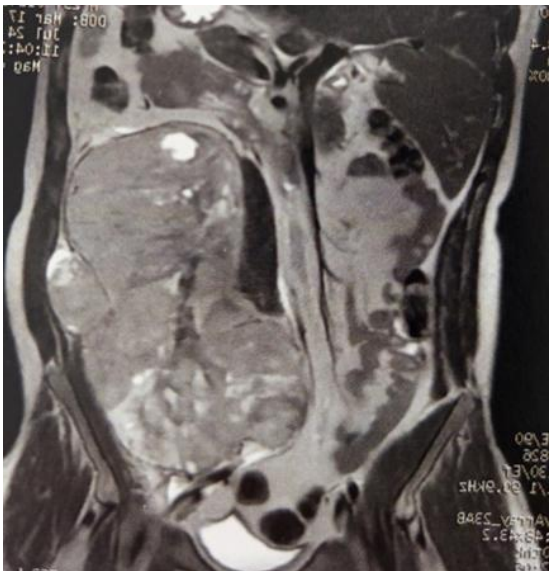

Figure 4

A radio-guided biopsy returned in favor of a biphasic synovial sarcoma grade 2 according to FNCLCC (3+0+1). An extension assessment was unremarkable. A multidisciplinary consultation meeting indicated chemotherapy based on doxorubicin + ifosfamide for tumor reduction followed by potential surgical excision.

III. Discussion

Synovial sarcoma represents 5 to 10% of soft tissue sarcomas, and can originate from all soft tissues independent of the synovial membrane. It affects young adults, with an incidence peak between 15 and 40 years, with a sex ratio leaning towards males and an incidence of 3/1,000,000/year [1]

This sarcoma is due to a specific chromosomal translocation, t(X;18) (p11.2;q11.2), which fuses the SS18 genes (formerly called SYT) and SSX (generally SSX1 or SSX2). The resulting oncogene induces malignant cell transformation.[1,2]

Patients typically present non-specifically with a painless or slightly painful mass near a joint, often of slow growth. Signs and symptoms may include: palpable mass, pain or tenderness at the tumor site, limited joint mobility, neurological symptoms if the tumor compresses nerves.[1]

Diagnosis relies on a combination of imaging, histology, and molecular biology. MRI is used to evaluate local tumor extension and mapping. CT scan is primarily requested for extension assessment, especially pulmonary and bone.[1,4]

Anatomopathological study reveals microscopy characterized by spindle cells, often arranged in biphasic patterns (epithelioid and sarcomatous zones) or monophasic, with fixation of anti-EMA, BCL-2, and CD99 antibodies in immunohistochemistry. RT-PCR or FISH detects the t(X;18) translocation.[3,5]

Treatment of synovial sarcoma is based on surgery, radiotherapy, and chemotherapy [1,2,5]:

- Obtaining negative surgical margins is crucial to reduce local recurrence risk; however, palliative surgery aimed at tumor reduction remains valid.
- Chemotherapy based on doxorubicin and ifosfamide is usually used for large tumors, locally advanced or metastatic cases.
- Radiotherapy is indicated neoadjuvantly to reduce tumor size or adjuvantly to prevent local recurrences.
- Targeted Therapies: such as tyrosine kinase inhibitors, are currently under evaluation.

Prognosis depends on several factors, including: Size (>5cm), site (trunk or abdomen), stage of discovery, metastasis especially pulmonary (76%). 5-year survival is between 50 and 70%. Half of the operated patients develop local recurrences and/or distant metastases, primarily in the lungs.[3]

IV. Conclusion

Synovial sarcoma is a tumor with poor prognosis; its diagnosis must be early to achieve good results. Surgery remains the only curative treatment. Advances in chemotherapy may help improve the prognosis.

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Legends:

Figure 1: Axial CT scan showing a large left retroperitoneal mass displacing the iliac head of the iliopsoas muscle inwardly, moderately and heterogeneously enhanced after contrast injection, containing liquid areas with necrotic appearance and multiple calcifications, without detectable fatty component, measuring 21 x 13 x 9.9 cm.

Figure 2: Frontal CT scan showing a process extending into the homolateral paravertebral soft tissues, extending from L3 to LS over 97.8 x 71 x 60 mm. Slightly protruding over the L4-LS foramen. Topographically, this mass is in relation to the lower pole of the kidney and the iliac wing.

Figure 3-4: Axial and frontal MRI scans showing a voluminous lesional process centered on the left retroperitoneum in isosignal T1 and T2, with T2 hypersignal foci and heterogeneous T1 signal, extended from L2 to S4, measuring 190 x 133 x 227mm in maximum dimension, heterogeneously enhanced after Gado injection.

CONFLICT OF INTEREST: The authors declare no conflict of interest.