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Case Report

Radiology Section

Primary Monophasic Omental Synovial Sarcoma in Nulliparous Female: A Case Report

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ABSTRACT

Synovial Sarcoma (SS) is an uncommon soft-tissue malignancy that primarily occurs near tendon sheaths and bone joints. Its primary intra-abdominal location is infrequent and characterised by non specific clinical signs. The mainstay of treatment remains wide local excision with negative margins. Hereby the authors report a rare case of monophasic omental SS in a 30-year-old nulliparous female who presented with chief complaints of an enlarging lump in the right abdominopelvic region over the past year and constipation for three months. The mass was non mobile and non tender. The rest of the physical and laboratory examinations were normal. She has a history of diagnostic laparoscopic excision (for the same complaint) of bilateral adnexal masses with right salpingectomy done three months ago in an outside set-up where histopathology diagnosed the mass as a low-grade fibromyxoid tumour. Presently, Contrast-enhanced Magnetic Resonance Imaging (CEMRI) revealed a large lobulated heterogeneous mass of 19.5×15.4×20 cm extending into mesorectal fat and encasing the sigmoid colon, rectosigmoid junction, uterus, and bilateral ovaries. The patient was taken for emergency surgery as she started displaying symptoms of acute obstruction. Debulking surgery (R2 resection) with ileostomy was performed. Histopathology and immunohistochemistry confirmed the diagnosis of monophasic SS. The patient was started on doxorubicin and has completed three cycles of chemotherapy. SS is an infrequent entity with high mortality rates. The main course of treatment is surgery with healthy resection margins. Long-term follow-up is required due to the high recurrence rate.

Keywords: Abdominal tumour, Debulking surgery, Ileostomy

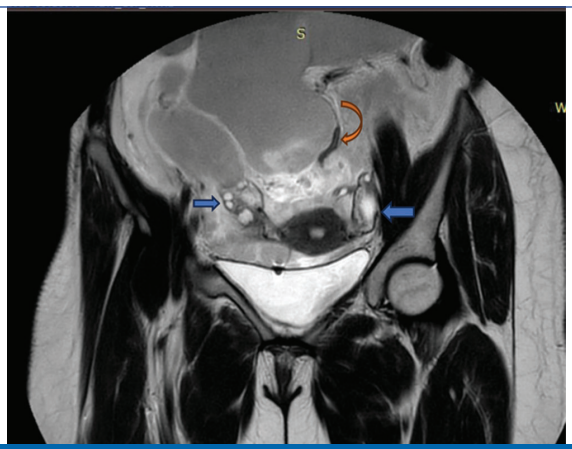
CASE REPORT

A 30-year-old female came to the hospital with the chief complaints of a gradually increasing painless abdominal mass for the last year and constipation for three months. She has a history of diagnostic laparoscopic excision of bilateral adnexal masses with a right salpingectomy done three months ago in an outside set-up for this same complaint. There, the histopathology diagnosed the mass as a low-grade fibromyxoid tumour. Thereafter, the patient was referred to the study institution.

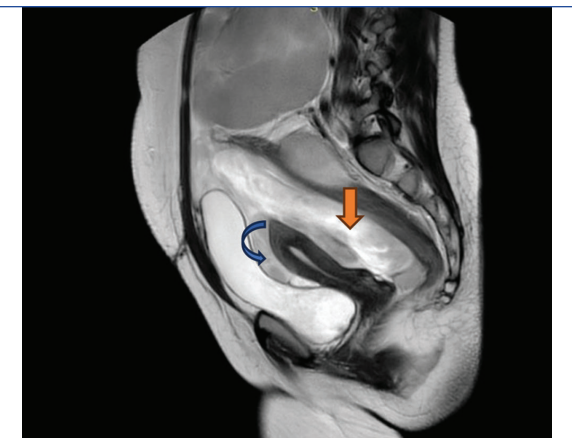
Physical examination revealed vitals within normal limits, an immobile firm non tender swelling (measuring 15×12.5 cm) in the right abdominopelvic region with no locoregional inflammatory signs. The abdomen was breathing normally, without any indication of peritoneal irritation. Complete blood count and biochemistry profiles were within the normal range. Tumour markers {Carcinoembryonic Antigen (CEA), CA125, alpha-fetoprotein, and beta-Human Chorionic Gonadotropins (HCG)} were normal.

Contrast-enhanced Magnetic Resonance Imaging (CEMRI) revealed relatively well-defined, lobulated lesions with a cumulative size of approximately (19.5×15.4×20.0) cm in the abdominopelvic region. The lesions showed heterogeneously hyperintense signal on T2 Weighted Image (T2WI)/Short Tau Inversion Recovery (STIR), iso to hypointense signal on T1WI.

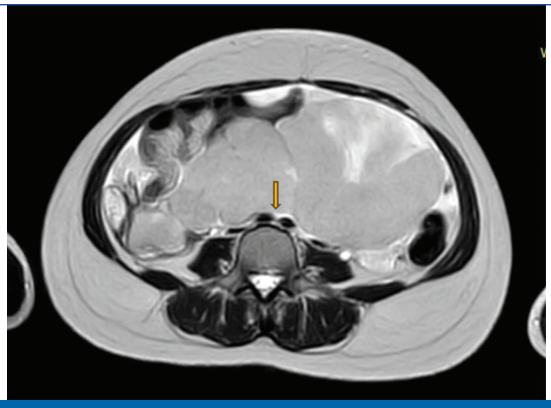
The lesions extended into mesorectal fat with encasement of the sigmoid colon and the rectosigmoid junction [Table/Fig-1]. Anteriorly, the lesions were extending and abutting the anterior abdominal wall. Inferiorly, the lesions encased bilateral ovaries and the uterus and extended into vesicouterine and rectouterine spaces [Table/Fig-2]. Posteriorly, there was an abutment of bilateral ureters, the aortic bifurcation, and bilateral psoas major muscles [Table/Fig-3]. Laterally and superiorly, the lesions were abutting and displacing adjacent bowel loops. Also, superiorly, the lesions reached up to the subhepatic region and were abutting the fundus of the gall bladder. The lesions showed restricted diffusion on Diffusion-weighted Imaging (DWI)/Apparent Diffusion Coefficient (ADC) [Table/Fig-4] with mild heterogeneous postcontrast enhancement.



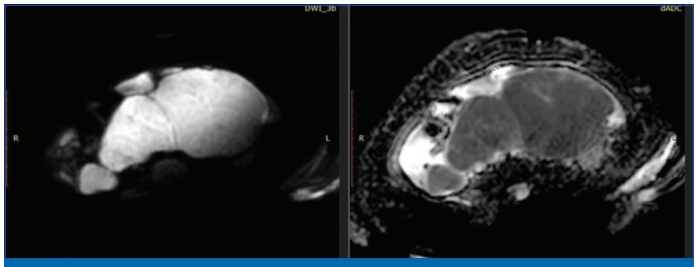
[Table/Fig-1]: T1 weighted coronal image showing mass abutting the sigmoid colon (curved arrow) Straight arrows: bilateral ovaries.



[Table/Fig-2]: T2 weighted axial image shows mass abutting. Aortic bifurcation (downward arrow).



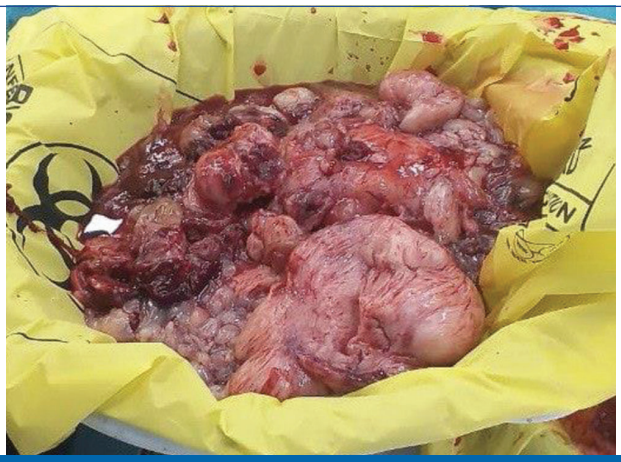
[Table/Fig-3]: T2 weighted sagittal image showing.
Mass infiltration into vesicouterine space (curved arrow) and the rectouterine space (straight arrow)



[Table/Fig-4]: DWI (right) and ADC (left) showing diffusion restriction.

The initial management plan based on the above radiological findings was to do an image-guided biopsy followed by systemic treatment. However, the patient was taken for emergency surgery as she started displaying symptoms of acute obstruction.

Intraoperative findings: Upon midline laparotomy, widely disseminated soft fleshy masses were scattered along all the peritoneal surfaces, including the parietal and visceral peritoneum. The diaphragmatic peritoneum was also involved. The abdominal and pelvic cavity was full of fleshy friable masses, part of which entrapped the uterus and sigmoid colon. Two large masses (6-7 cm in diameter) [Table/Fig-5] were seen arising from the peritoneum along the root of the mesentery. The greater omentum was nodular and fleshy. Piecemeal resection of the masses was performed, as the tumour was very scattered and fragile. The tumour masses were bleeding to the touch. Omentectomy was also performed along with R2 resection of the masses (R0 resection was not possible given the wide distribution of the disease along the distal colon and rectum complicated with haemodynamic instability). A diverting loop ileostomy was also done.



[Table/Fig-5]: Operated mass.

Histopathologically, it was diagnosed as SS (monophasic, FNCLCC (Fédération Nationale des Centres de Lutte Contre le Cancer) grade 3). [1]. Sections show tumour tissue disposed of in sheets and intersecting fascicles with a haemangiopericytomatous vascular pattern. The tumour was predominantly composed of spindle cells with pale eosinophilic cytoplasm and elongated to ovoid, moderately pleomorphic nuclei with finely dispersed chromatin. Mitoses (24-25/10 HPF) are evident. Tumour tissue was positive for vimentin, TLE-1, and Bcl-2, and was negative for Epithelial Membrane Antigen (EMA), CD-34, S-100, desmin, and DOG-1. The Ki-67 proliferation index was 40-45%.

Further workup included whole-body Positron Emission Tomography-Computed Tomography (PET-CT) scan for staging and 2-D echo. After that, palliative chemotherapy with single-agent doxorubicin (D1-D3) every three weeks was started. A multidrug regime could not be initiated given the poor patient status. The patient has completed all three cycles of chemotherapy and currently is doing well.

DISCUSSION

The SS is a rare and aggressive tumour first described in 1893 [2] and represents up to 10% of all soft-tissue malignancies. Contrary to its name, SS is not related to synovial tissue and is considered a tumour of unknown origin [2]. It most commonly develops in the extremities, particularly in the knee (70%), although many other sites can be affected, including the retroperitoneum, abdominal wall, chest wall, head and neck, and subcutaneous tissues. Primary intra-abdominal omental SS has rarely been reported [3]. It occurs at any age, but it is more common in adolescents and young adults aged between 15 and 35 years [2].

The most common presentation is usually a painless mass. However, the symptoms depend on the site and size of the mass [4]. The gross appearance depends on the rate of growth and the site of the tumour. Slow-growing tumours are well-circumscribed, firm, round, or multinodular lesions. Cystic changes may be prominent. Rapidly growing tumours are poorly circumscribed with a variegated, friable appearance. There may be areas of haemorrhage and necrosis within the tumour [4].

Histologically, there are three main subtypes: biphasic SS, monophasic SS, and poorly-differentiated. Monophasic SS is common (up to 60% of SS) and is traditionally composed of spindle cells harbouring a fascicular pattern. Biphasic SS is made of two components: mesenchymal spindle cells with epithelial components arranged in a glandular pattern. This subtype represents nearly 25% of all SS. Poorly-differentiated SS exhibits generally epithelioid morphology with severe nuclear atypia and a high mitotic rate [4]. SS displays a rich vascularisation with a hemangiopericytomatous pattern of vessels [2,5]. Immunohistochemically, the spindle cell population of SS can show focal or widespread positivity for EMA (most sensitive) and for CK7 and CK19, which are nearly 100% specific [5]. The characteristic t(X;18) (p11.2; q11.2) translocation is a cytogenetic hallmark of SS that presents in nearly all SS and does not occur in other forms of sarcomas [6].

Radiologically, there is no specific presentation. Ultrasound is usually the first imaging modality, concluding with a solid hypoechoic soft-tissue mass or heterogeneous with more echoic areas and irregular margins. Due to its incapacity to recognise malignant features, further investigations are required. MRI is the leading imaging technique for diagnosis and assessment of local invasion (bone involvement, neurovascular encasement, and muscular invasion), thus estimating the aggressiveness of the tumour and planning the surgical course [7].

The MRI of SS appears isointense to slightly hyperintense on T1-weighted images and reveals a mosaic of mixed low, intermediate, and high signal intensity on T2-weighted images because of the admixture of solid components, cystic degeneration, haemorrhage, myxoid stroma, and fibrous tissue. The main differentials for intra-abdominal SS are leiomyosarcoma, malignant schwannoma, gastrointestinal stromal tumour, and solitary fibrous tumour. The most common CT appearance of SS is mixed soft tissue with attenuation equal to or slightly less than that of muscle. The heterogeneous areas may represent necrosis or haemorrhage of the tumour. Marked heterogeneity and enhancement are highly suggestive of SS on both CT and MRI [3]. The final diagnosis

is always based on histopathology and immunohistochemistry. Like other soft tissue sarcomas, SS metastasises mainly to the lung. Unlike most other soft tissue sarcomas, however, SS carries a small risk of spread to lymph nodes [4].

The main course of treatment in non metastatic SS is wide surgical excision, including the tumour, and a wide resection with negative margins (R0) is recommended. In practice, resections may be grossly complete but have microscopically positive margins (an R1 resection) or be macroscopically incomplete (an R2 resection) [8]. If resection margins are not respected, the risk of local relapse is high [7].

Throughout literature reviews, many prognostic factors have been reported (age, specimen size, resection margins, histological subtype, tumour grade, and fusion type) but only tumour size superior to 5 cm has been linked more than others to a poor outcome [7]. Since SS is associated with an elevated local recurrence and metastatic risks, long-term follow-up is needed. Despite various methods of treatment and long-term surveillance, disease-free survival at 10 years does not exceed 65% [7].

CONCLUSION(S)

The main course of treatment is surgery with healthy resection margins. However, adjunctive radiation treatment and chemotherapy, either alone or in combination, may decrease local relapse and control metastases. Long-term follow-up is required due to the high recurrence rate.

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