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PRIMARY OVARIAN LEIOMYOSARCOMA – A RARE CASE REPORT

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INTRODUCTION

Leiomyosarcoma, a rare malignant tumour originating from smooth muscle cells, constitutes a relatively small portion, approximately 10-20%, of soft tissue sarcomas. (1) While it can develop directly from smooth muscle cells or precursor mesenchymal cells, many leiomyosarcomas arise in the uterus, with the remainder emerging from various soft tissue sites, often associated with smooth muscle cells in vessel walls. Among these rare occurrences, leiomyosarcoma of the ovaries is an exceptionally infrequent pathology, with fewer than 20 reported cases in the existing literature. (2)

Primary leiomyosarcoma of the ovary (POLMS) predominantly afflicts women in the peri- and postmenopausal stages of life, presenting with non-specific symptoms like abdominal pain, bloating, and unintended weight loss, among others. Unfortunately, PLSO is frequently diagnosed at advanced stages, sometimes accompanied by metastasis, and currently lacks well-established diagnostic and management strategies. (3)

In efforts to enhance diagnostic precision, immunohistochemical and electron microscopy assessments have been employed. Nevertheless, surgery remains the cornerstone of treatment, while the role and methods of implementing adjuvant therapies remain contentious. (4) The prognosis for primary ovarian leiomyosarcomas is dire, with outcomes contingent on factors such as tumour stage, size, grade, and mitotic index, with recurrence predominantly observed in the abdominal and pelvic regions.

The exceedingly rare and aggressive nature of primary ovarian leiomyosarcoma, originating from smooth muscle cells within the ovary, sets it apart from its more common counterparts found in the uterus, gastrointestinal tract, or soft tissues. Given its scarcity, diagnostic complexities, and propensity for rapid progression, this malignancy presents an intriguing subject for clinical scrutiny and case reporting.

This case report embarks on a comprehensive exploration of the clinical presentation, diagnostic process, treatment strategies, and outcomes of a patient diagnosed with primary ovarian leiomyosarcoma. The rarity of this malignancy emphasizes the importance of documenting individual cases to expand our knowledge of its clinical behaviour, diagnostic markers, and therapeutic options. Moreover, by sharing this case, we aim to contribute to the limited body of literature available, potentially assisting healthcare professionals in early detection and improved management of future instances of primary ovarian leiomyosarcoma.

CASE REPORT

A 66-year-old post-menopausal female para 4 live 4 all four normal vaginal delivery presented with chief complaints of lower abdominal pain in the last 4 months, complaints of loss of appetite and weight loss in the last 6 months also had complaints of fullness and abdominal distension past 3 -4 months. There was a history of evening rise of temperature in the past 2 months, but no history of change in bladder habits, or complaints of recurrent episodes of loose stools on and off past 2 months. On examination, the patient was pale abdominal palpation was soft but diffuse tenderness was present no guarding or rigidity was noted Laboratory investigation showed reduced haemoglobin, elevated total count, and increased Serum CA 19-9 142U/ml CA 125 CEA within normal limits.MRI abdomen and pelvis showed a large heterogeneous peripherally enhancing lobulated solid lesion with central necrosis seen epicentered in the left adnexa and extending into the midline, suggestive of malignant ovarian neoplastic aetiology. MRI ORADS -IV/V. Ascitic fluid analysis showed a reddish colour with atypical cells. PET CT showed an irregular hypermetabolic mass with cystic areas in the left ovary, adherent to the fundus of the uterus, superior surface of the urinary bladder, right ovarian mass, anterior abdominal wall, ileal loops and proximal sigmoid colon, suggestive of carcinoma of the left ovary. The non-hypermetabolic cystic lesion in the right ovary contains fat suggestive of a dermoid cyst. Primary cytoreductive surgery (bowel resection with appendicectomy with omentectomy with total abdominal hysterectomy) was done. The specimen showed an

ovarian mass of 20 x 10 cm with necrotic encasing the small bowel loops with plaques of pus material in the mass, ileum involved with mesentery up to 15 cm along with right adnexal mass and appendix. The specimen was sent to HPE and was diagnosed with leiomyosarcoma of ovary FIGO stage IIIC. This was followed by 4 cycles of chemotherapy with Ifosphamide, Mesna, Adriamycin, and Cyclophosphamide. The patient was on regular follow-up, and post 3rd cycle of chemotherapy patient's condition started deteriorating. CT abdomen and pelvis showed evidence of distant metastasis noted in the anterior abdominal wall muscular and subcutaneous plane. Despite receiving 4 cycles of chemotherapy the patient ultimately succumbed to the disease due to the presence of distant metastasis and the poor prognosis of the tumor.

IMAGING FEATURES

MRI ABDOMEN AND PELVIS

A large heterogeneous peripherally enhancing lobulated lesion with central necrosis was seen epicentre in adnexa and extending into the midline with relation as described - likely malignant ovarian neoplastic aetiology.

A fairly defined peripherally enhancing cystic lesion with internal enhancing septation and fat component noted in the right adnexa extending into the midline likely ovarian mature teratoma – MRI O- RADS – II/III.

PET CT

Irregular, hypermetabolic mass with cystic areas in the left ovary - suggestive of carcinoma of the left ovary. The mass appears densely adherent to the fundus of the uterus, superior surface of the urinary bladder, right ovarian mass, anterior abdominal wall, ileal loops and proximal sigmoid colon. Fat stranding in the omentum in the lower abdomen with no discrete peritoneal nodules or ascites. No evidence of lymph node metastasis or distant metastasis. Non-hypermetabolic cystic lesion in the right ovary containing fat - suggestive of dermoid cyst.

GROSS EXAMINATION (Fig 1 & 2)

The intraoperative right ovarian mass, capsule is ruptured and showed an ovarian mass of 20 x 10 cm with necrotic encasing the small bowel loops with plaques of pus material in the mass, ileum involved with mesentery up to 15 cm along with right adnexal mass and appendix. Small nodule on the anterosuperior segment of the liver 0.5 x 0.5 cm below the ligament of teres suggestive of haemorrhagic nodule. Left ovary capsule intact, serosa of bilateral fallopian tubes intact.

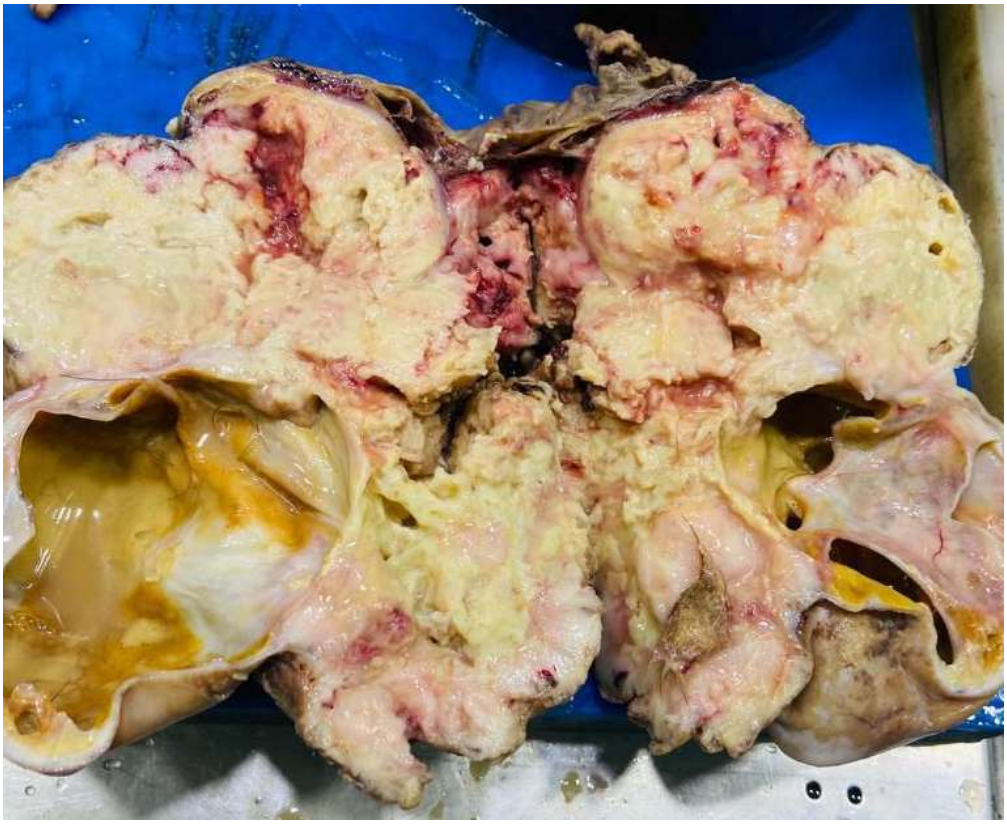

Fig1: Gross lesion in the right ovary

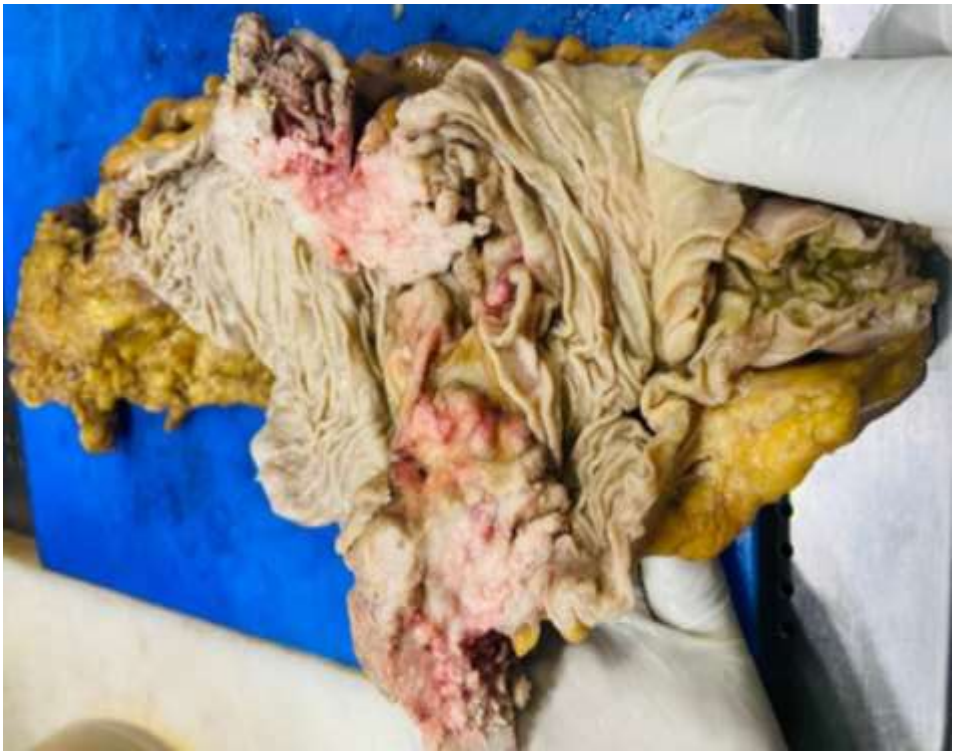

Fig2: Gross lesion in the omentum

MICROSCOPIC EXAMINATION (Fig 3-8)

Microscopically, the histological type of the tumour revealed teratoma with malignant transformation with high-grade spindle cell sarcoma in the right ovarian mass. The surface of the ovary is involved along with the surface of bilateral fallopian tubes. Additional findings include, the omental nodal showing a malignant tumour, and in the left ovary, serosa shows adherent tumour tissue.

In special staining, immunohistochemistry chemistry was performed which is negative for tumour cells in the markers 100 and MY OD1 and positive for SMA there is strong cytoplasmic positivity in 15%-20% of tumour cells to the final impression suggestive of features of teratoma with moles malignant transformation of right ovary suggest suggestive of leiomyosarcoma sarcoma.

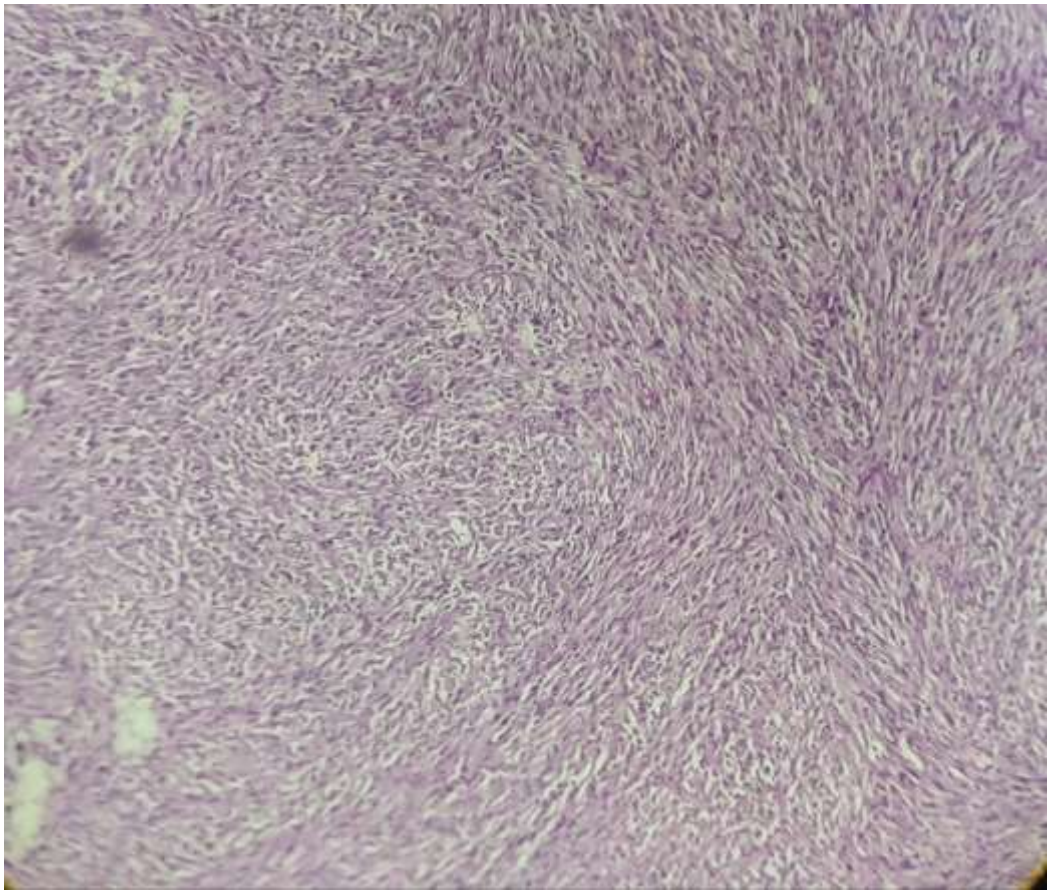

Fig3: High-grade spindle cell sarcoma- 100X

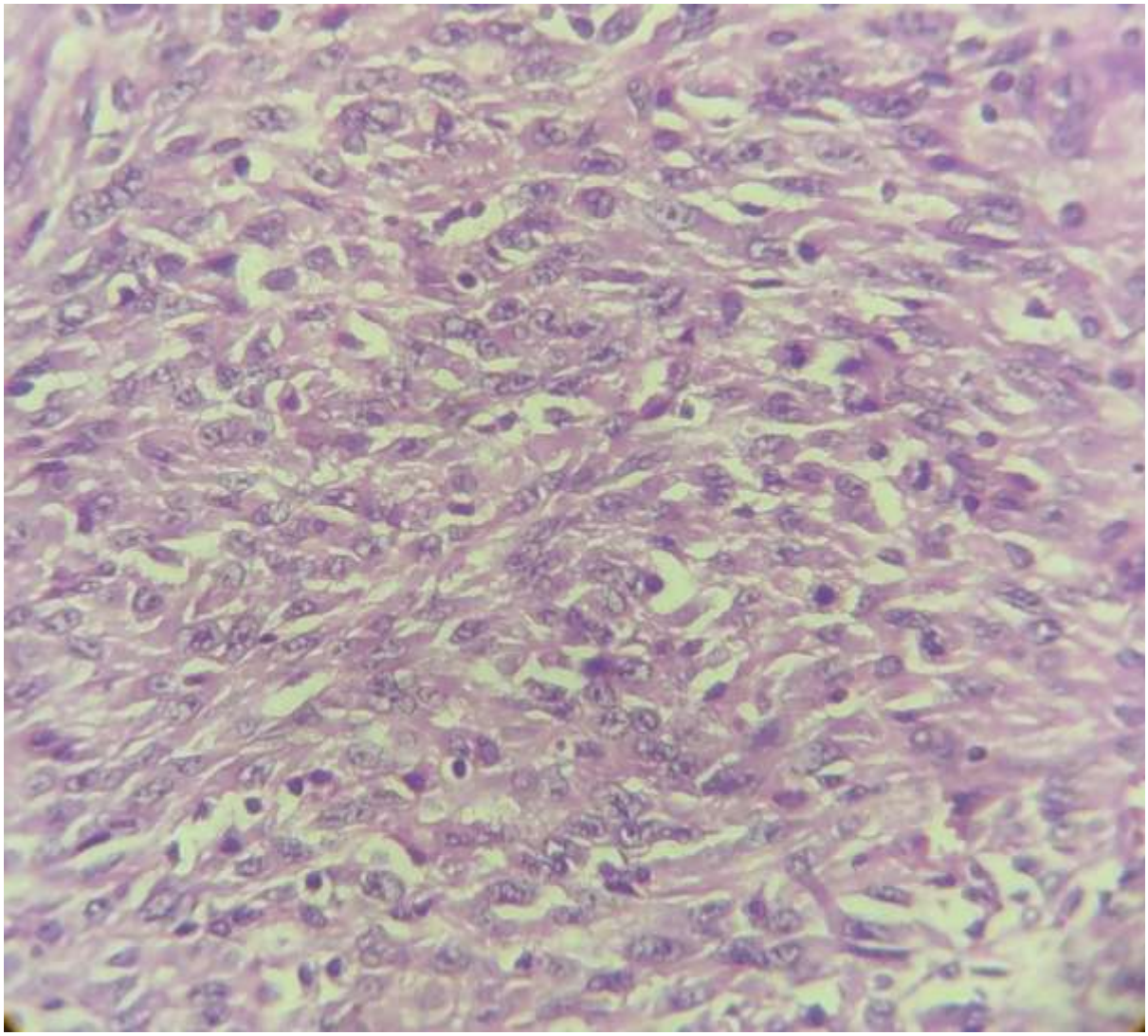

Fig4: High-grade spindle cell sarcoma- 400X

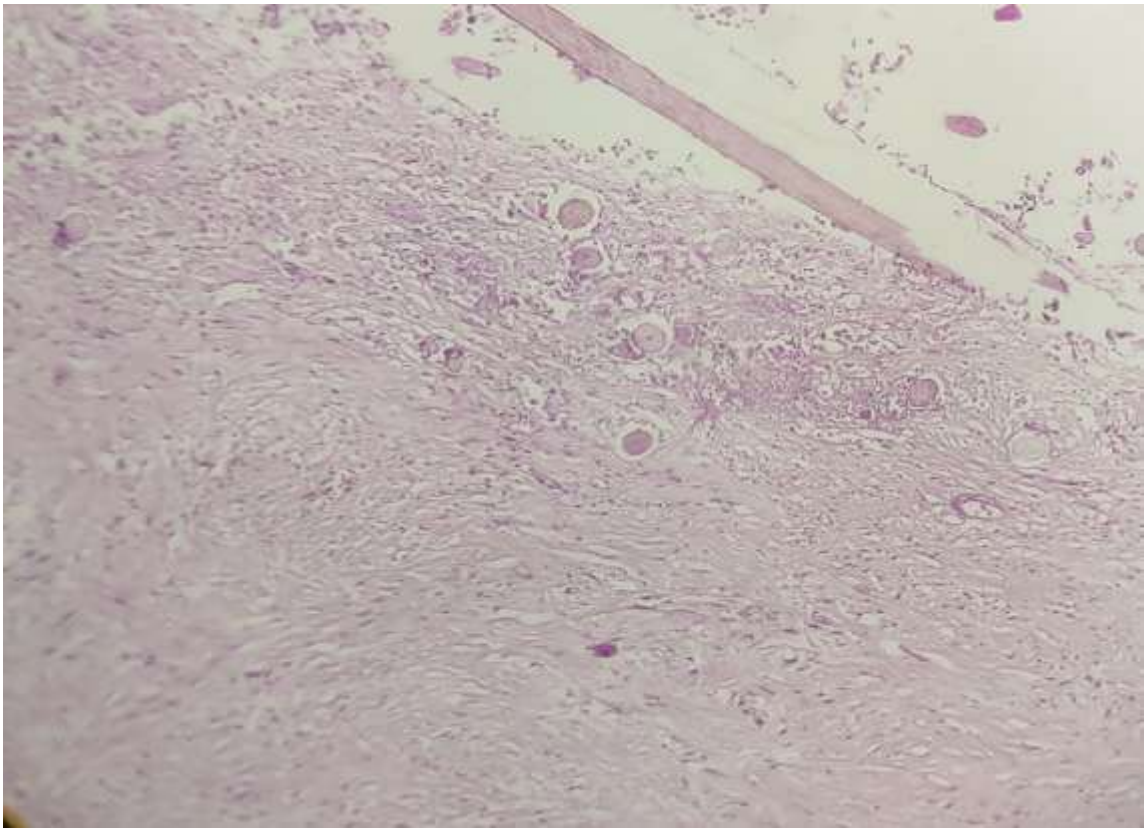

Fig5: Ovarian lesion with teratomatous component- 40X

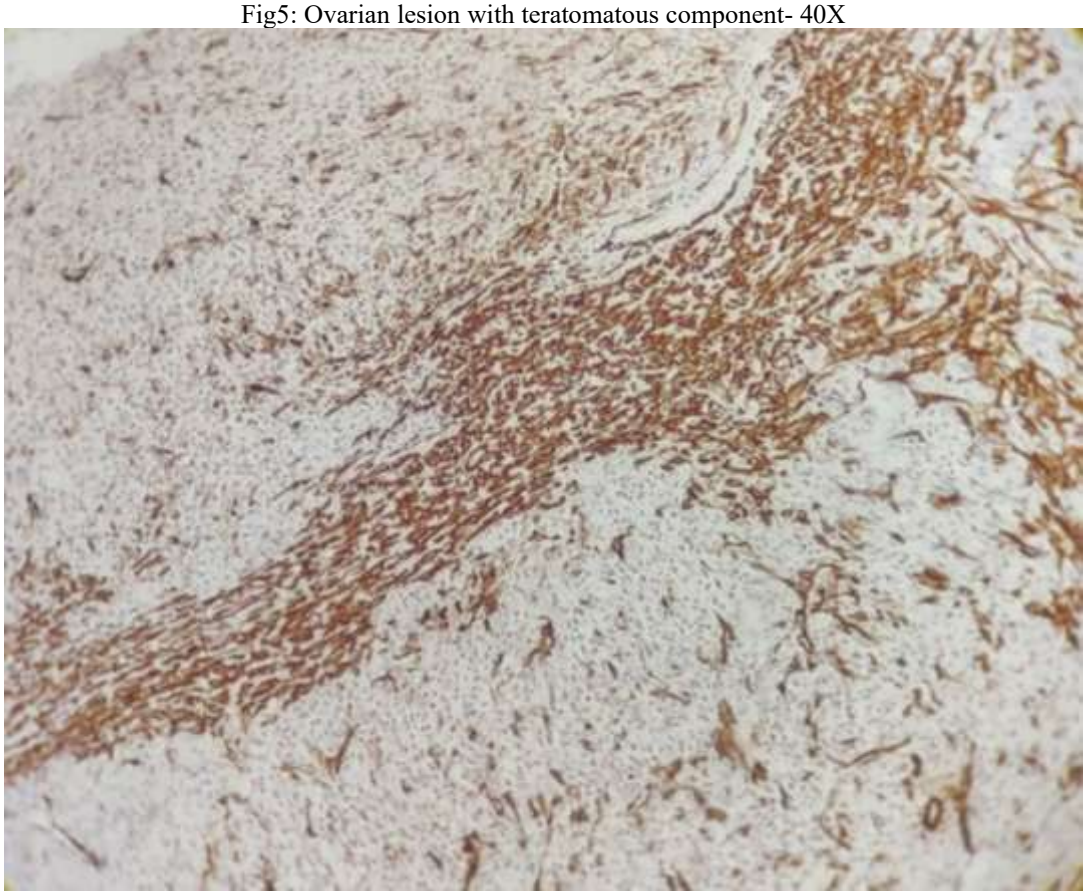

Fig6: IHC showing SMA positivity in tumour cells- 200X

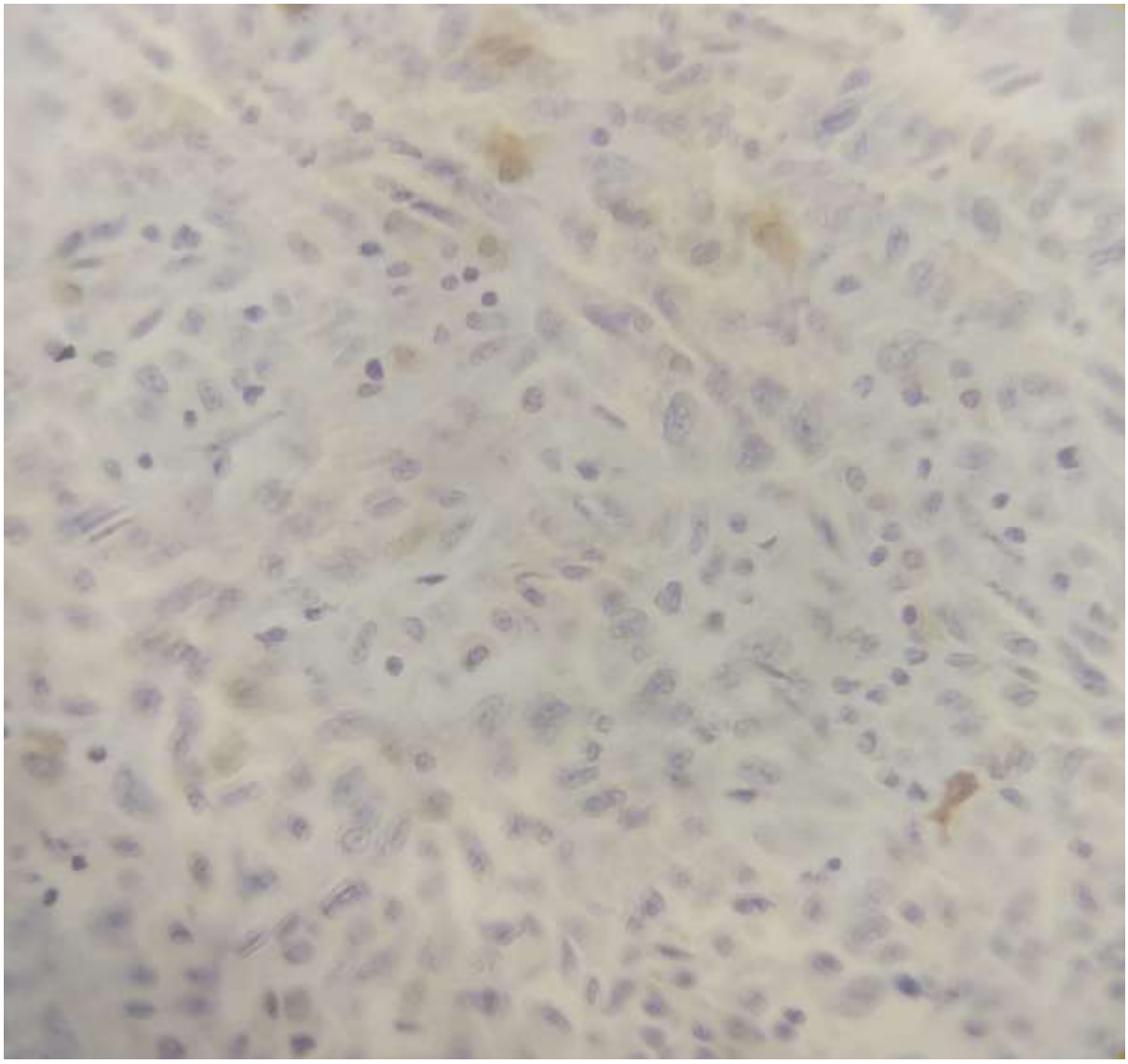

Fig7: IHC showing S100 negativity in tumor cells- 400X

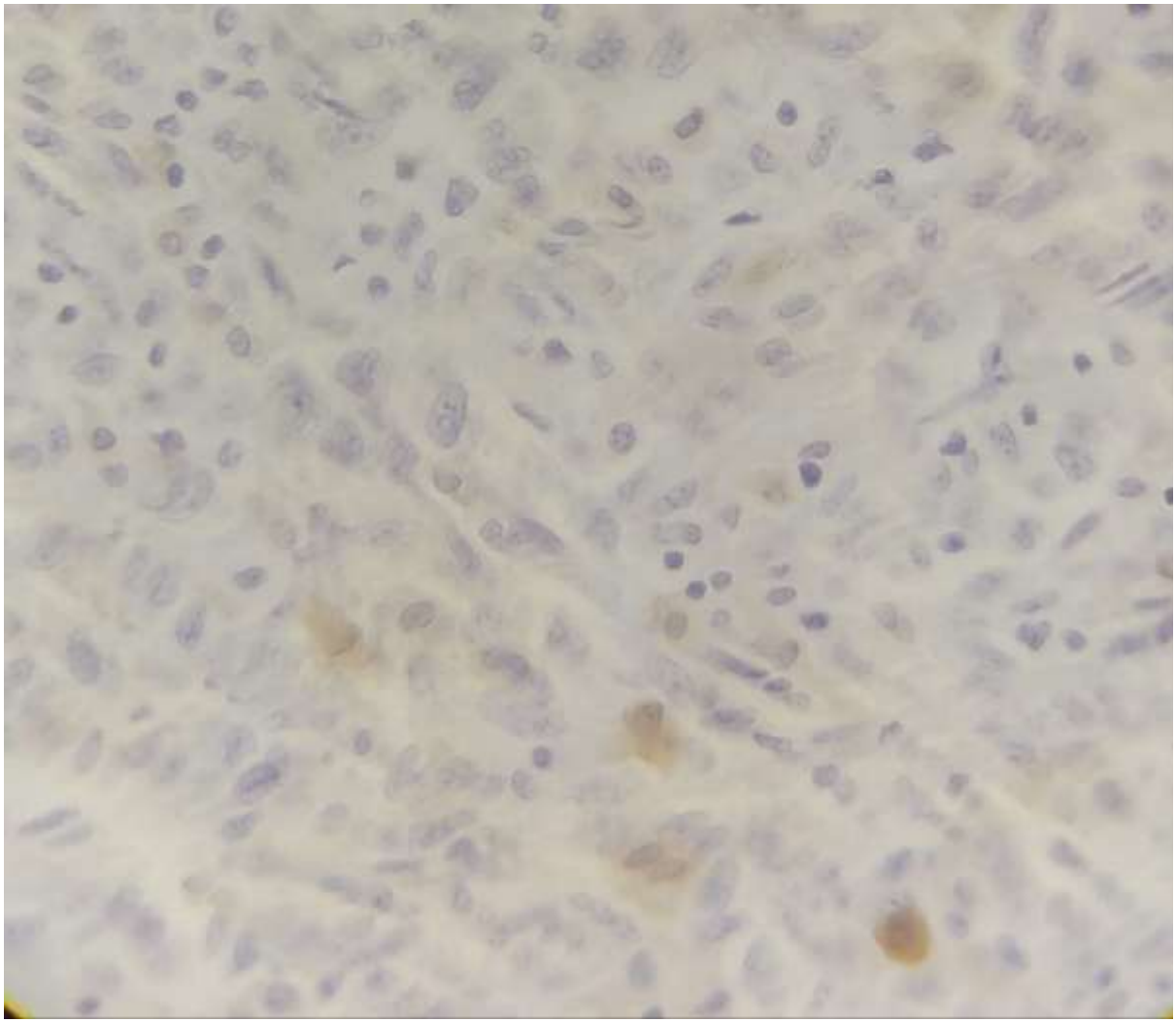

Fig8: IHC showing Myo D1 negativity in tumor cells- 400X

DISCUSSION

Primary ovarian leiomyosarcoma (PLSO) is an exceptionally rare malignancy, posing significant challenges in both diagnosis and treatment due to the scarcity of literature and established guidelines. Most PLSO cases present as abdominal pelvic masses, leading to compressive symptoms resulting from pressure on adjacent visceral structures. Diagnosis heavily relies on histopathological examination and immunohistochemistry, while surgical debulking or cytoreduction remains the primary treatment. A multidisciplinary approach, involving tumour board presentations and discussions, is essential to determine the optimal surgical approach and the required surgical procedures.

This case highlights the diagnostic evaluation, treatment procedure, and histopathological confirmation of primary ovarian leiomyosarcoma in a 66-year-old female patient. The patient's non-specific symptoms, such as lower abdominal pain, loss of appetite, and weight loss, led to a diagnosis at an advanced stage, which is a common presentation for this rare malignancy. Such non-specific symptoms may also lead to a differential diagnosis that includes fibrosarcoma, rhabdomyosarcoma, and thecomas. (5)

In the case presented here, a combination of imaging techniques, MRI abdomen and pelvis, revealed a large heterogeneous peripherally enhancing lobulated solid lesion with central necrosis in the left adnexa and extending into the midline. Further PET CT revealed an irregular hypermetabolic mass with cystic areas in the left ovary, which is adherent to the fundus of the uterus, superior surface of the urinary bladder, right ovarian mass, anterior abdominal wall, ileal loops and proximal sigmoid colon. These imaging features are consistent with carcinoma of the left ovary, but given the non-specific nature of these findings, histopathological confirmation should be done after the excision of the tumour.

To address this complex scenario, surgical gastroenterology and gynaecological teams collaborated to plan a cytoreductive surgery. The cornerstone of treatment was primary cytoreductive surgery, which included bowel resection, appendicectomy, and omentectomy, alongside total abdominal hysterectomy.
Intra-operative exploration revealed a large necrotic mass of 20 x 10 cm in the right ovary, encasing small bowel loops with no significant enlargement of pelvic or para-aortic lymph nodes. Although the right ovarian lesion presented challenges during surgical excision, successful debulking was achieved through a comprehensive procedure, including total abdominal hysterectomy, bilateral scalping-oophorectomy, ileal with mesentery excision to 15 cm, omentectomy, and appendicectomy, followed by ilio-ileal anastomosis and haemostasis.
Histopathological examination confirmed a diagnosis of teratoma with malignant transformation, specifically the high-grade spindle cell variant of leiomyosarcoma.
Immunohistochemistry can prove valuable in the diagnosis of primary ovarian leiomyosarcoma (POLMS), aiding in the differentiation of its subtypes and, consequently, providing insights into its biological behaviour and eventual prognosis.
Further immunohistochemical staining is generally positive for SMA, desmin, vimentin, and caldesmon, weakly positive to negative for S100 protein, and negative for inhibin, EMA, and cytokeratin antibodies. (6)In this case, it showed S100-negative, Myo D1-negative, and SMA-positive results. The haemorrhagic ascitic fluid analysis further supported the diagnosis, highlighting atypical cells in a background of haemorrhage.
Primary leiomyosarcoma of the ovary is a rare malignancy, accounting for less than 1% of all adult malignancies. It typically originates from mesenchymal tissue, with embryonic mesoderm and neuroectoderm contributions. This cancer has a predilection for soft tissues and abdominopelvic organs compared to extremities. Leiomyosarcoma typically emerges after the third decade of life, peaking in the perimenopausal age group during the fifth decade. (4)
Recurrent occurrences of primary ovarian leiomyosarcoma (POLMS) are predominantly observed in the abdomen and pelvis, with less frequent instances documented in the lung, bone, liver, mediastinum, brain, preauricular lymph node, and scalp. (7) It is imperative to explore key clinical parameters such as response rates, metastatic tendencies, and treatment outcomes specific to primary ovarian leiomyosarcoma in comparison to metastatic leiomyosarcomas originating from other sites or leiomyosarcomas of different soft tissue origins. Collaborative efforts with specialists in sarcoma are essential to drive improvements in patient outcomes and further our understanding of this challenging malignancy.
Soft-tissue tumours present an ongoing challenge in the field of surgical pathology due to their complex biological behaviour and histogenesis. Immunohistochemistry plays a pivotal role in achieving precise diagnoses and subcategorization within this domain.(5) In the context of primary ovarian leiomyosarcoma (POLMS), which has an exceptionally poor prognosis, surgery remains the established treatment modality for this rare tumour. Nevertheless, the efficacy of chemotherapy and radiotherapy in managing POLMS remains unclear. Combining histological parameters such as tumour differentiation, necrosis, and mitotic activity with immunohistochemistry can enhance the accuracy of diagnosis. Given the rarity, heterogeneity, and diversity of soft-tissue tumours, documenting such cases is essential as they can contribute significantly to future research endeavours.
In conclusion, this case report sheds light on the non-specific presentation, diagnostic approaches, radical treatment, and histopathological confirmation of primary ovarian leiomyosarcoma in a 66-year-old female patient. It emphasised on the importance of accurate diagnosis through histopathological evaluation and the presence of immunohistochemistry markers along with comprehensive preoperative imaging which is crucial for appropriate management. Cytoreductive debulking surgery always remains the mainstay of treatment with adjuvant chemotherapy following the procedure to prevent recurrence or further metastasis further research and collaboration are necessary to improve our understanding of the rare tumour to optimise this treatment strategy and improve the patient outcomes.

CONCLUSION

This case report illuminates the intricacies of primary ovarian leiomyosarcoma (POLMS), an exceedingly rare and diagnostically challenging malignancy. It underscores the critical role of accurate diagnosis achieved through meticulous histopathological evaluation, complemented by immunohistochemistry markers and comprehensive preoperative imaging. Cytoreductive debulking surgery remains the cornerstone of treatment for POLMS, supported by the imperative need for adjuvant chemotherapy to mitigate the risk of recurrence or distant metastases. The presented case underscores the rarity and complexity of this malignancy, emphasizing the necessity for continued research and interdisciplinary collaboration to enhance our comprehension of POLMS and optimize treatment strategies, ultimately improving patient outcomes. This unique case serves as a valuable contribution to the limited body of knowledge surrounding primary ovarian leiomyosarcoma, offering insights that may guide future clinical management and research endeavours in the field of soft-tissue tumours.

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