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PRIMARY OVARIAN LEIOMYOSARCOMA PRESENTING AS A COMPLEX ADNEXAL MASS – A CASE REPORT

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ABSTRACT
Ovarian Leiomyosarcoma is a rare entity accounting for less than 0.1% of all ovarian neoplasms.(1) We report a case of a 58 year old post menopausal lady who presented to us with complaints of abdominal distension for 6 months. Imaging studies revealed a 18x16x18 cm mass in the lower abdomen and pelvis. Patient underwent surgery and was diagnosed as Ovarian Leiomyosarcoma and is now being planned for adjuvant chemotherapy.

KEYWORDS: Ovarian leiomyosarcoma; Ovarian sarcoma; Uterine-type leiomyosarcoma; Postmenopausal woman; Ovarian neoplasm; Pelvic mass; Abdominal distension; Rare ovarian tumor; Adjuvant chemotherapy; Gynecologic oncology

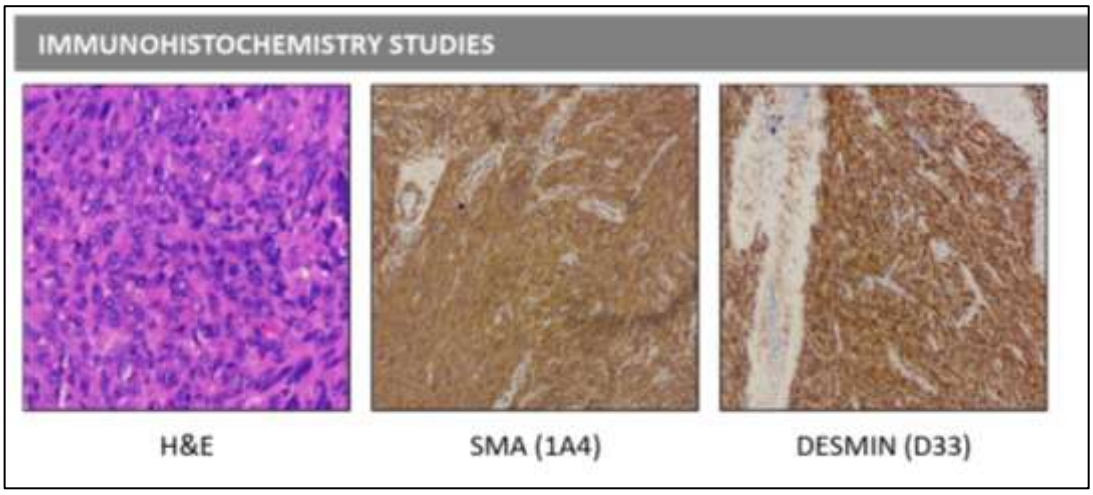
INTRODUCTION

Mesenchymal ovarian neoplasms include all primary ovarian tumours of connective tissue. They arise from the ovarian tissue rather than the epithelial or Mullerian cells(2). Ovarian leiomyosarcoma accounts for 0.01% of ovarian sarcomas which in turn accounts for around 1% of all ovarian neoplasms. (1)
They usually arise from malignant degeneration of ovarian leiomyoma or smooth muscles in blood vessels in cortical stroma and corpus luteum, muscular attachments of ovarian ligament, wolffian duct remnants, totipotential ovarian remnants or from a teratoma. (3)
The prognosis depends on stage, tumour size and mitotic index. On histology they appear as enlarged eosinophilic spindle shaped cells with variable pleomorphic nuclei arranged in bundles or sheets. On immunohistochemical analysis, tumour cells are positive for desmin, smooth muscle actin (SMA), H-caldesmon and rarely bcl-2. They also express Estrogen and progesterone receptors.(4)
Due to their rare nature optimal management is affected by the lack of clinical knowledge. They are usually managed by surgery followed by adjuvant chemotherapy and/or radiation.
Differential diagnosis include Krukenberg tumours (GI history, pan CK+), Mixed mullerian tumours (pan CK+), sarcomatoid form of sex cord tumours (Sex cord elements present), Undifferentiated carcinomas (pan CK+, caldesmon negative).

Clinical Case

Our case is of a 58 year old post menopausal lady with co morbidities of Hypertension and Rheumatoid arthritis on Methotrexate and Leflunomide, post hysterectomy in 2015 for Abnormal Uterine Bleeding caused by Leiomyoma (AUB-L) . The histopathological examination was suggestive of simple endometrial hyperplasia along with multiple leiomyomas of uterus with cystic, mucoid and hyaline degeneration. She presented to us now in the out patient department with complaints of abdominal distension for 6 months along with abdominal tightness for 2 days and occasional right iliac fossa pain. The symptoms were associated with loss of appetite and early satiety.
On examination she was found to have a 15x16 cm mass, solid cystic in consistency with restricted mobility along with bilateral forniceal fullness.
Sr CA 125 was 18.6 U/mL, Sr. CEA of 1.29 and Sr. Ca 19.9 was 5.2. She underwent imaging with a whole body PET CT scan which was suggestive of heterogeneously FDG avid heterogenous mass lesion with internal areas of enhancement and fluid attenuation in the lower abdomen, pelvis measuring 18x16x18 cm with an SUV max of 9.5. B/L adnexa were not seen separately from the lesion and the lesion was compressing the urinary bladder and displacing the bowel loops superiorly and laterally and had loss of fat planes with the sigmoid colon along with presence of mild ascites. Faint to non FDG avid peritoneal and omental fat stranding with intermittent nodular thickening was also noted.

Patient underwent staging laparotomy along with right salpingooophorectomy with bilateral pelvic lymph node dissection and total omentectomy and peritoneal wash. Post op Histopathology was suggestive of spindle cell neoplasm involving right ovary. Capsular breach present. All 17 lymph nodes negative. Omentum, right subdiaphragmatic biopsy, right pelvic peritoneal biopsy, Left subdiaphragmatic biopsy, and left pelvic peritoneal biopsies were negative for malignancy. Peritoneal wash cytology was negative for malignancy.
On Immunohistochemical examination tumour cells were positive for SMA, Desmin, CD34(focal weak), H caldesmon and negative for Myo D1, S100, SF-1, MDM2, CK and Inhibin and had a Ki67 index of 25%.
Based on HPE and IHC final diagnosis of Ovarian Leiomyosarcoma was made.



Patient has been further planned for adjuvant chemotherapy with Adriamycin and Ifosfomide along with Mesna.

DISCUSSION

Primary Ovarian Leiomyosarcoma (POLMS) is most commonly encountered in postmenopausal women with a median age of 52.9 years(5). Patients present mostly with non specific symptoms such as abdominal discomfort, pain and distension, palpable lump in the abdomen and difficulty in urination. Laboratory tests may reveal anaemia. Tumour markers may be normal and are non specific(6). Imaging tests are helpful in determining the location, size, morphology and internal structure of tumour. Definitive diagnosis is made by histopathological examination along with IHC. Microscopically, the presence of at least two of the following three criteria is required for diagnosis: coagulative necrosis, cellular atypia, and a mitotic index of ≥10/HPF (7)
Spindle cell morphology with areas of necrosis can be seen in the above image. IHC for smooth muscle actin (SMA) positivity is seen (D).

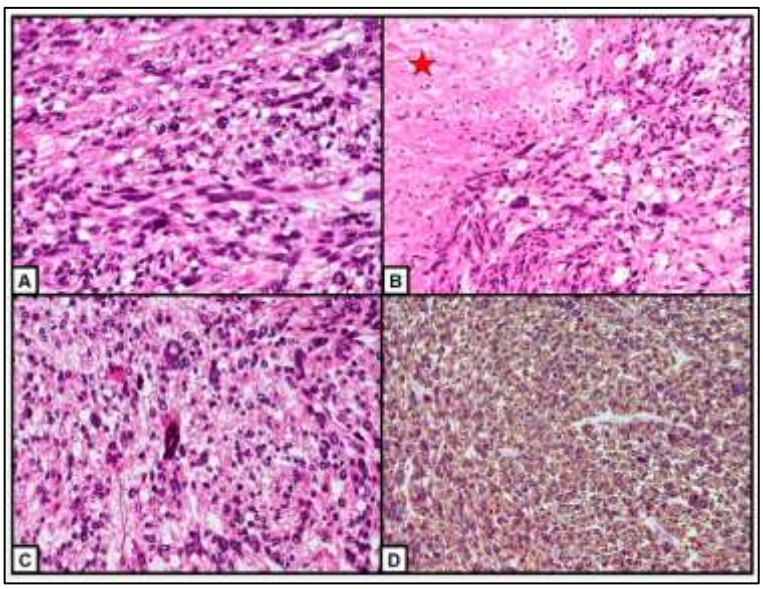


Image credits: Mandato, V.D.; Torricelli, F.; Mastrofilippo, V.; Palicelli, A.; Costagliola, L.; Aguzzoli, L. Primary Ovarian Leiomyosarcoma Is a Very Rare Entity: A Narrative Review of the Literature. Cancers 2023, 15, 2953. https://doi.org/10.3390/cancers15112953 (4)

Due to the rarity of these tumours there are no clear guidelines on the management. For staging of ovarian Leiomyosarcoma, the International Federation of Obstetrics and Gynaecology (FIGO) recommends the same staging system as for epithelial ovarian tumours.
The Gynaecological Cancer InterGroup (GCIG) recommends Total Abdominal Hysterectomy and Bilateral Salpingoophorectomy as the surgical approaches to POLMS.(8) In a literature review by Mandato et al, patients who underwent hysterectomy with salpingoophorectomy and lymphadenectomy followed by adjuvant chemotherapy seemed to have a better prognosis. (4) But a multivariate analysis by the same authors showed that the stage of the disease was the main prognostic indicator irrespective of the modality of treatment. There is no conclusive evidence that adjuvant treatment with chemotherapy or radiation improves outcomes but surgery along with lymphadenectomy followed by adjuvant chemotherapy or radiation may provide some survival advantage. (4)
In most of the literature available, surgery remains the main modality of management with no clear evidence proving the utility of adjuvant treatment. It may be possible that patients with a longer survival may have a low grade disease. Due to the rarity of these tumours, the estimated 5 year survival of POLMS is around 30-54%(8).

CONCLUSION

Primary Ovarian Leiomyosarcoma (POLMS) is a rare and aggressive tumour arising from the soft tissue of the ovaries or the connecting ligaments.
Due to its rarity the optimal management still remains a challenge. Staging is similar to epithelial ovarian tumours.
Surgery still remains the cornerstone of treatment with benefit of adjuvant therapy still under evaluation.
More clinical studies are required for the better understanding of this rare entity.

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