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Case Report

Uterine Leiomyosarcoma: A Rare Entity

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Abstract

Uterine leiomyosarcoma is a rare uterine malignancy that arises from the smooth muscle of uterine wall. Presenting symptoms may be vague and mimic other benign uterine conditions. They are notoriously aggressive with poor prognosis. We report a case of a 35-year-old female, who presented with abdominal distension with pain and vomiting. A non-tender mass measuring 14X12 cms was felt on palpation in the same region. CT scan revealed a large, relatively well defined, hypodense pelvic mass in retro-uterine pouch. Patient underwent exploratory laparotomy with primary resection of tumour with pan-hysterectomy with pelvic lymph node resection. Histopathologically, it was diagnosed as pleomorphic sarcoma probably leiomyosarcoma of uterus which showed immunostain Smooth Muscle Antigen (SMA) positive and Epithelial Membrane Antigen (EMA) negative. It was Estrogen Receptor(ER) negative. The rarity as well as pathological diversity makes this study beneficial and this definitive reporting will be helpful in diagnosis of smooth muscle tumours with challenging histologic features.

Keywords: Uterine leiomyosarcoma, malignancy, smooth muscle, hysterectomy, pleomorphic, immunostain, SMA, EMA, ER.

INTRODUCTION

Uterine leiomyosarcoma is a rare uterine malignancy that arises from the smooth muscle of uterine wall [1]. It accounts for 1-2% of uterine malignancies with an estimated annual incidence of 0.04 per 100,000 women [1]. In majority of patients it presents with abnormal vaginal bleeding, palpable pelvic mass and pelvic pain [2]. They are notoriously aggressive with poor prognosis [3].

CASE REPORT

We report a case of a 35-year-old female, who presented with abdominal distension with pain and vomiting. Per abdominal examination showed fullness in the umbilical region, left iliac fossa and left lumbar region.

A non-tender mass measuring 14X12 cms was felt on palpation in the same region .CT scan revealed a large, relatively well defined, hypodense pelvic mass in the retro-uterine pouch.

Patient underwent exploratory laparotomy with primary resection of tumour with pan-hysterectomy with pelvic lymph node resection.

Gross morphology showed uterus with intramural ill-defined, irregular greyish white to greyish brown mass measuring 12x5x1cms, extending into subserosal area, received in bits and pieces.

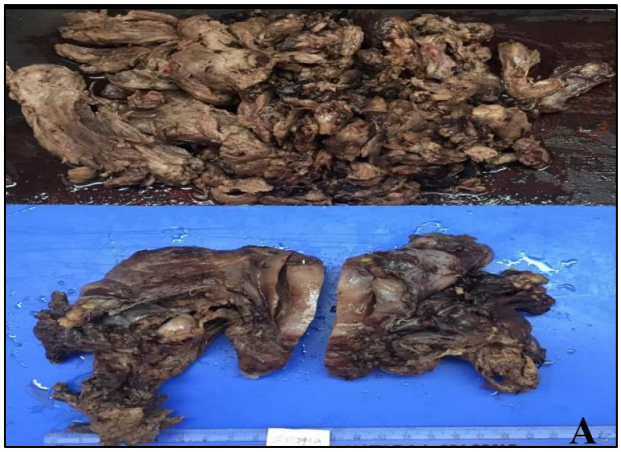
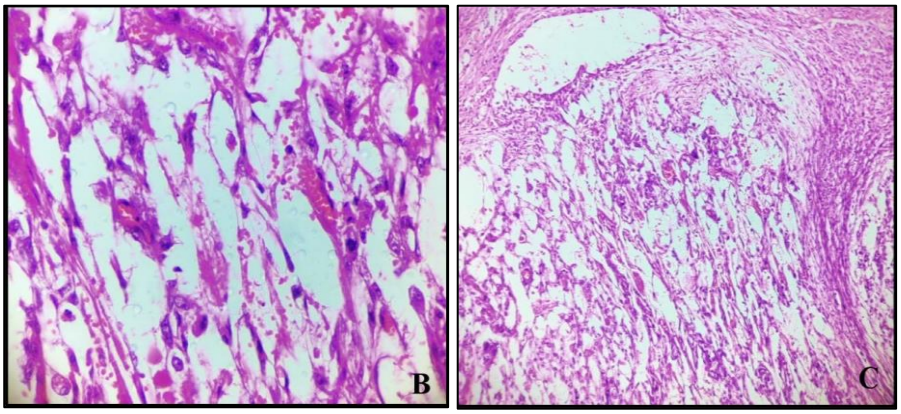


Figure A: Uterus with Intramural Ill-Defined, Irregular Greyish White to Greyish Brown Mass Measuring 12x5x1 CMS and Extending into Subserosal Area, Received in Bits and Pieces

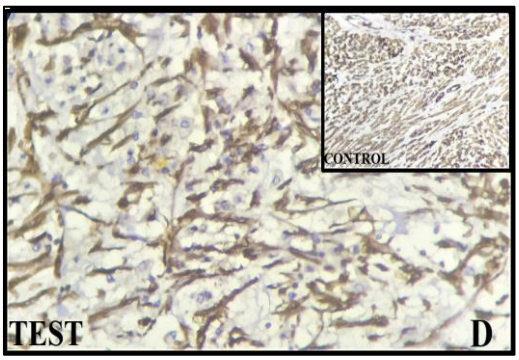
Microscopically, a malignant neoplasm comprising of tumor cells arranged in sheets, nests, organoid arrangement, cords and in singles was seen. The cells were pleomorphic spindle shaped with scant eosinophilic cytoplasm, had pleomorphic vesicular nucleus with prominent nucleoli. Scattered tumor giant cells were seen. Mitotic figures were sparse. Lymphovascular invasion was seen. Intervening stroma showed congested blood vessels, hemorrhage and chronic mononuclear inflammatory infiltrate. Areas of necrosis were noted.



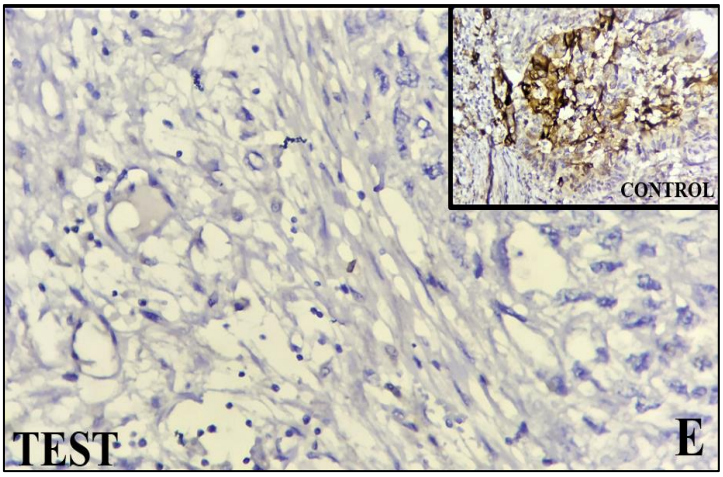
Photomicrograph B and C: Pleomorphic Spindle Shaped Cells with Scant Cytoplasm, Pleopmorphic Vesicular Nucleus and Prominent Nucleoli (B – 40X and C – 10X, H&E)

Histopathologically, it was diagnosed as pleomorphic sarcoma – suggestive of leiomyosarcoma of uterus.

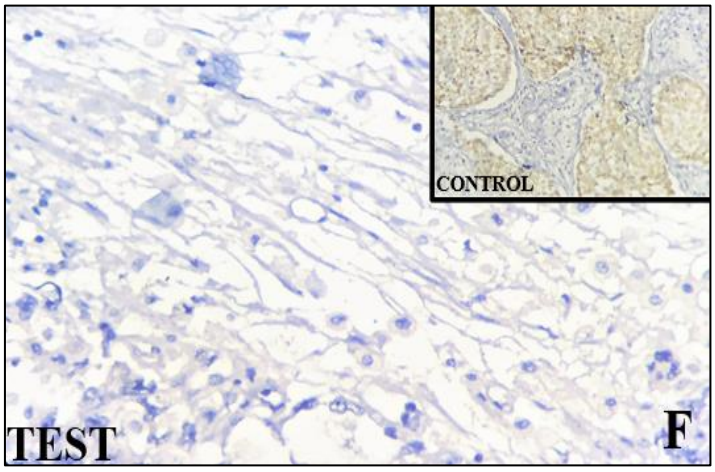
Immunohistochemistry was done for confirmation, which showed immunostain Smooth Muscle Antigen (SMA) positive and Epithelial Membrane Antigen (EMA) negative. It was Estrogen Receptor(ER) negative.



Photomicrograph D: SMA Control-Uterine Fibroid (Inset) SMA Test–Showing Positive



Photomicrograph E: **EMA Control - Colon (Inset) EMA Test – Showing Negative**



Photomicrograph F: ER Control- Fibroadenoma (Inset) ER Test – Showing Negative

Final Diagnosis: Uterine Leiomyosarcoma.

DISCUSSION

Uterine leiomyosarcoma is the most common type of uterine sarcoma [4]. It is an extremely aggressive malignancy associated with a poor overall prognosis. Presenting symptoms may be vague and mimic other benign uterine conditions [2]. These rare mesenchymal tumors are characterized by cytologic atypia, a high mitotic index, and tumor necrosis on histologic inspection [4]. Use of immunostains like Smooth Muscle Actin (SMA) highlights the smooth muscle origin and a reduction of ER immunoexpression is associated with an increase of the malignant potential, as seen in this case [5]. The diagnosis of uterine leiomysarcoma means poor outcome for most of the patients affected, and complete hysterectomy without tumor laceration and with clean tumor margins is the definite goal of surgical therapy for these patients [3].

Novelty

The rarity as well as pathological diversity makes this study beneficial.

CONCLUSION

Uterine leiomyosarcoma is a rare tumour. It originates from the myometrium or myometrial vessels [1]. Women affected may vary in age, but are most often diagnosed in their perimenopausal years [4]. It is difficult to differentiate uterine leiomyoma from leiomyosarcoma before surgery as the diagnostic tools are not reliable. Thus, the diagnosis is confirmed by the histopathological examination of the tumor mass done after surgery [2]. Immunohistochemistry is an extremely useful tool in diagnosis while supporting the high degree and aggressiveness of this malignancy [5]. This definitive reporting with combination of immunohistochemistry (IHC) study will be helpful in diagnosis of smooth muscle tumours with challenging histologic features.

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Conflict of Interest: None.
Source of Funding: None.

Ethical Consideration

As the study required only routine histopathology slides and blocks with no additional financial requirements, and consent being taken from

the patient at the time of hospital admission regarding utilization of data for research purpose, the protocol did not need formal ethical clearance from review board.

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