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Case Report

Symplastic leiomyoma: a rare clinicopatholgical diagnosis

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ABSTRACT

Although leiomyomas are common this case is being reported due to its uniqueness as it posed challenges at all levels of pathological rarity and management. Symplastic leiomyoma is an unusual benign variant of leiomyoma with less likelihood of malignant transformation. In our case report we present a 28-year-old nulligravida who presented to our OPD with Fibroid uterus detected on scan and myomectomy specimen done revealed a rare pathological diagnosis of symplastic leiomyoma and her followup.

Keywords: Symplastic, Leiomyoma bizarre, Atypical leiomyoma

INTRODUCTION

Although leiomyomas are common this case is being reported due to its uniqueness as it posed challenges at all levels of pathological rarity and management. Symplastic leiomyoma is an unusual benign variant of leiomyoma with less likelihood of malignant transformation. Patient counselling is important to alleviate the anxiety associated with such histologic reports. Atypical and bizarre leiomyoma synonymous with symplastic leiomyoma are smooth muscle tumors that contain cells with moderate to severe cytological atypia, with no cell necrosis and mitotic index fewer than 10/10 HPF. It was also diagnosed in different sites other than uterus such as the vagina, nasal cavity, and scrotum. Malignant transformation accounts for 0.2% of all cases of leiomyomas.

CASE REPORT

Mrs M, 28 years lady, married for 6 months, presented to OPD for opinion regarding fibroid uterus diagnosed on ultrasound on routine checkup. She had no history suggestive of abnormal uterine bleeding or abdominal distension. No history of weight loss, pain abdomen or dysmenorrhea, bowel or bladder complaints. On enquiring her mother had undergone hysterectomy in view of abnormal uterine bleeding-leiomyoma. Her sibling also has large leiomyoma.

On examination-moderately built and nourished. No pallor and pedal edema. Vitals stable. Breast, thyroid and spine normal. Per abdomen examination-An irregular, bosselated, abdominopelvic mass corresponding to 30 weeks gravid uterus size, extending transversly from right lumbar region to left lumbar region occupying all quadrants of abdomen, firm in consistency, non tender, lower border not made out.

Per speculum examination revealed a healthy cervix and vagina. Per vaginal examination showed firm posteriorly deflected cervix with left forniceal fullness: mass felt in the pouch of douglas, non-tender, uterus corresponding to 30 weeks size.

She was further evaluated. Blood investigations were normal. MRI pelvis done for fibroid mapping-revealed multiple fibroids-Left fundus-FIGO 5 transmuralisoechoic $8\times8.5\times7.8$ cm, midline anterior fundus-FIGO 4intramural isoechoic $3.4\times4\times4.2$ cm, right lateral corpus-FIGO 5 transmural-hyperintense $11.3\times10.5\times10.4$ cm, left lateral corpus FIGO 5 transmural-hyperintense $4.8 \times 4.4 \times 5.2$ cm and right lateral cervix-FIGO 8posterolateral right cervix $7.5 \times 6 \times 5.8$ cm-hyperintense.



Figure 1: MRI fibroid mapping showing multiple fibroid uterus.

She underwent laparotomy by midline subumbilical vertical incision- -myomectomy done through tunnelling incision technique after bilateral ureteric catheterisation under combined spinal epidural anaesthesia.

Intraoperatively Uterus was irregularly enlarged to 28 weeks gravid uterus size with multiple large fibroids-Fundal subserous fibroid FIGO V-8×8 cm, right lateral wall subserous fibroid FIGO V 10×10 cm, two intramural fibroids of 4×4 cm FIGO IV from anterior wall, along with multiple seedling fibroids. HPE revealed symplastic leiomyoma-Large hyperchromatic nuclei with chromatin clumps. Giant cells with multiple large nuclei and stroma with myxoid changes. There was no nuclear atypia/necrosis. Tumor was regular with low mitotic activity.



Figure 2: Intraoperative figure showing uterus with large leiomyoma.



Figure 3: Large fundal fibroid.



Figure 4: Enucleation of fundal fibroid during myomectomy.



Figure 5: Retrieved myomectomy specimens-five leiomyomas.



Figure 6: Spindle shaped neoplastic cells in fascicles (100x H and E), bizarre nuclear forms with giant cells.

Follow-up

Follow-up scan post 6 months post-surgery showed uterus AV $11 \times 6 \times 6$ cm with small seedling fibroids and Polycystic ovaries. HSG was done which showed normal cavity with free spill on both tubes. She is currently on management for subfertility.



Figure 7: TVS scan showing uterus with endometrial cavity post-surgery.



Figure 8: Hysterosalpingogram post surgery showing normal uterine cavity with free spill on both tubes.

DISCUSSION

Numerous histologic variants of uterine leiomyomas are described. One such variant is the symplastic leiomyomaathe term reserved for uterine leiomyomas with giant cells, nuclear atypia and, minimal mitotic activity (0-4/10 hpfs). The presence of atypia is well recognized. In 1966, Taylor and Norris first introduced the term 'atypical' uterine leiomyomas to denote the presence of cytologic atypia in uterine leiomyomas. Subsequently terminologies used for histologically similar lesions with cytologic atypia included 'bizarre leiomyoma' by Christopherson et al and, 'symplastic leiomyoma' by burns, suprarenal symplastic leiomyoma of the inferior vena cava, myxoid tumor of the uterus and right atrial myxomas benign metastatizing leiomyoma have been reported.¹⁻⁵ The case of symplastic leiomyoma had previously been reported as a single case. The criteria used in order to consider a smooth muscle tumour as a leiomyosarcoma were: a mitotic rate of two or more mitoses per 10 high power field/presence of necrosis. CD 74 and p53 are two of the most studied markers.

The following criteria were evaluated:⁶ Degree of cytologic atypia (none to mild or moderate to marked), presence or absence of coagulative necrosis and mitotic index (MI) (Table 1).

Table 1: Criteria for evaluating various categories of smooth muscle neoplasm of the uterine corpus.⁶

Leiomyoma	MI < 20MF/10 HPF
Leiomyoma with increased MI	No coagulative necrosis
	No atypia or no more than mild
	cytological atypia
Leiomyoma with	MI ≥20MF/10 HPF
increased MI,	No coagulative necrosis
but experience	No atypia or no more than mild
limited	cytological atypia
Atypical leiomyoma	MI <10/10 HPF
	No coagulative necrosis
	Diffuse moderate to severe
	cytological atypia
Leiomyosarcoma	Any MI
	Any degree of cytological atypia
	with coagulative necrosis
Epithelioid leiomyoma	MI <5MF/10HPF
	No coagulative necrosis
	No atypia/more than mild atypia
Epithelioid leiomyosarcoma	MI ≥5 MF/10 HPF
	No coagulative necrosis
	None /any degree of cytological
	atypia

The presence of hematopoietic or heterologous elements within an otherwise bland uterine leiomyoma or endometrial stromal tumor may give rise to diagnostic difficulties. The regularity of the tumor margins, low mitotic activity, and absence of nuclear atypia or necrosis should be made for the exclusion of malignancy. In the presence of massive lymphocytic infiltration of a leiomyoma, the clonality of the infiltrate may aid in differentiating it from malignant lymphoma. The pathogenesis and clinical significance of these rare neoplasms remain to be clarified.

CONCLUSION

Symplastic leiomyoma is an unusual variant of leiomyoma. Malignant transformation accounts for 0.2% of all cases of leiomyoma. The regularity of the tumor margins, low mitotic activity and absence of nuclear atypia or necrosis should be made for the exclusion of malignancy.

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