

## Unusual presentation of more common disease/injury

## Symplastic leiomyoma of uterus- a clinico-pathological dilemma

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**Summary**

The diagnosis of uterine smooth muscle tumours is usually not difficult. Occasionally, benign tumours with an unusual histopathology may cause some dilemma for clinicians who had not experienced such report before. A 50-year-old female patient was admitted to our gynaecology clinics with lower abdominal heaviness and urinary symptoms and undergone total abdominal hysterectomy with bilateral salpingo-oophorectomy with a probable diagnosis of cervical fibroid. Histopathological examination showed lipoleiomyoma uterus with focal symplastic features. This case report describes the clinic-pathological dilemma in managing the patient with symplastic leiomyoma of the uterus.

**BACKGROUND**

Although this case is of more clinical significance, the present case is being reported due to its uniqueness as it posed challenges at all levels of clinical diagnosis, pathological rarity and management. Clinical diagnosis was difficult, as on per vaginal examination, the firm mass was seen at the apex of the vagina, obscuring the cervix. This was secondary to mass expanding the lower uterine segment and cervix. Diagnostic curettage was impractical as there was no access to cervix. CT scan of pelvis showed the heterogenous mass with interspersed fat densities at the utero-vesical pouch, likely immature teratoma. MRI either as a diagnostic or therapeutic modality could not be availed due to her financial constraints. Intraoperatively, firm mass was seen expanding the cervix and the lower uterine segment like that of a cervical fibroid. Bilateral ovaries were found normal. In the histopathological report, a rare type of leiomyoma was described showing focal symplastic features. Again immuno-histochemical studies could not be performed due to financial constraints. The patient was explained the less likelihood of such leiomyomas undergoing malignant transformation. On reviewing the literature, very few case reports were found. Hence this case is being reported to share the experiences of the clinician in managing, counselling the patients when encountered with such rare histologic features in a very common condition like leiomyoma.

**CASE PRESENTATION**

A 50-year-old postmenopausal lady attended gynae clinic with lower abdominal heaviness along with urinary complaints like stress urinary incontinence and frequency of micturition bothering for 2 years. She had an obstetric history of three gestations resulting in vaginal deliveries. On general physical examination, she was obese weighing 75 kilograms making her body mass index of 29. Abdominal examination did not reveal any mass or organomegaly.

On per-speculum examination, cervix was obscured by a large globular mass. On bimanual pelvic examination, 6×6 cm firm, relatively fixed, non-tender mass was felt at the lower uterine segment and cervix. Uterus was found bulky. Bilateral fornices were free and non-tender. Cervical fibroid was the first differential diagnosis. Contrast-enhanced CT (CECT) of abdomen and pelvis showed 13×8.5×8 cm heterogenous vertically elongated well-defined mass with interspersed fat density seen in the utero-vesical pouch, likely immature teratoma. Haemogram revealed haemoglobin 14.4 grams, platelet count 2.9 lakhs, total leucocyte count 7100/cumm. Biochemical tests showed normal blood sugars, liver and renal function tests. Chest x-ray and electrocardiogram were normal. Urine routine examination, microscopy and cultures were normal. CA-125 was 14.35 units/ml. After consultation with the patient and relatives, exploratory laparotomy along with hysterectomy was planned. The patient underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy. Operative findings included large cervical firm mass measuring 11×9×8 cm and bulky uterus. Bilateral tubes and ovaries were grossly normal. Cut section of the cervical mass showed whorled appearance with areas of necrosis. Histopathological examination showed lipoleiomyoma of uterus with focal symplastic features. Basal endometrium and chronic cervicitis were the other findings in the uterus. Bilateral tubes and ovaries were unremarkable. Postoperative period was uneventful. The patient is on follow-up without any significant adverse events.

**INVESTIGATIONS**

CECT of abdomen and pelvis showed 13×8.5×8 cm heterogenous vertically elongated well-defined mass with interspersed fat density seen in the utero-vesical pouch, likely immature teratoma. Haemogram revealed haemoglobin 14.4 grams, platelet count 2.9 lakhs, total leucocyte count 7100/cumm. Biochemical tests showed normal blood

**Table 1** Criteria for evaluating various categories of smooth muscle neoplasms of uterine corpus used by<sup>7</sup>

Smooth muscle tumours with usual differentiation	
Leiomyoma	MI <20 mf/10 hpf
Leiomyoma with increased mitotic index	No coagulative necrosis No atypia or no more than mild cytologic atypia
Leiomyoma with increased mitotic index but experience limited	MI >/=20 mf/10 hpf No coagulative necrosis No atypia or no more than mild cytologic atypia
Atypical leiomyoma with low risk of recurrence	MI <10 mf/10 hpf No coagulative necrosis Diffuse moderate to severe cytologic atypia (1/46 failed)
Leiomyoma with atypia but limited experience	MI </=10 mf/10 hpf No coagulative necrosis Focal moderate to severe cytologic atypia
Leiomyosarcoma	Any MI Any degree of cytologic atypia with coagulative necrosis (29/39 failed) Usually there will be significant cytologic atypia
Leiomyosarcoma	MI >/=10 mf/10 hpf No coagulative necrosis but with diffuse moderate to severe cytologic atypia (4/10 failed)
Smooth muscle tumours with myxoid stroma	
Epithelioid leiomyoma	MI <5 mf/10 hpf No coagulative necrosis No atypia or no more than mild cytologic atypia
Epithelioid leiomyosarcoma	MI >/=5 mf/10 hpf No coagulative necrosis None or any degree of cytologic atypia Or Any MI Any degree of cytologic atypia with coagulative necrosis

MI, mitotic index.

sugars, liver and renal function tests. Chest x-ray and electrocardiogram were normal. Urine routine examination, microscopy and cultures were normal. CA-125 was 14.35 units/ml.

### DIFFERENTIAL DIAGNOSIS

- ▶ Immature teratoma
- ▶ Cervical fibroid.

### TREATMENT

Total abdominal hysterectomy and bilateral salpingo-oophorectomy.

### OUTCOME AND FOLLOW-UP

The patient is being treated as for a case of benign leiomyoma.

### DISCUSSION

Numerous histologic variants of uterine leiomyomas have been described. One such variant is the symplastic leiomyoma—a term reserved for uterine leiomyomas with giant cells, nuclear atypia and minimal mitotic activity (0–4/10 hpf). The presence of atypia is well recognised. In 1966, Taylor and Norris first introduced the term ‘atypical’ uterine leiomyomas to denote the presence of cytologic atypia in uterine leiomyomas. Subsequent terminologies used for histologically similar lesions with cytologic atypia included ‘bizarre leiomyoma’ by Christopherson *et al* in 1971 and

‘symplastic leiomyoma’ by Burns.<sup>1,2</sup> Suprarenal symplastic leiomyoma of the inferior vena cava,<sup>3</sup> myxoid tumour of the uterus and right atrial myxomas,<sup>4</sup> benign metastasising leiomyoma<sup>5</sup> have been reported.

The case of the 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 or presence of necrosis. CD74 and p53 are two of the most studied markers. The immunohistochemical profiles and genetic aberrations of the examined cases suggest that uterine leiomyosarcoma could arise from the pre-existing leiomyoma-like areas that often have a symplastic or cellular morphology. As the frequency of leiomyosarcomas is only 0.1–0.3% of the frequency of leiomyomas only rare leiomyomas progress to leiomyosarcoma. As cellular and symplastic leiomyoma-like areas were over-represented in uterine leiomyosarcoma-associated leiomyoma-like areas, leiomyomas with this morphology may be more prone to malignant transformation than usual type leiomyomas.<sup>6</sup>

Bell, Kempson and Hendrickson analysed 213 problematic cases and devised a classification scheme to distinguish between the various categories of smooth muscle neoplasms of the uterine corpus. The following summarises their findings.

The following criteria were evaluated,<sup>7</sup>

- ▶ Degree of cytologic atypia (none to mild or moderate to marked)
- ▶ Presence or absence of coagulative necrosis
- ▶ Mitotic Index (MI) (table 1)

The presence of hematopoietic or heterologous elements within an otherwise bland uterine leiomyoma or endometrial stromal tumor may give rise to diagnostic difficulties. Regularity of the tumour margins, low mitotic activity and absence of nuclear atypia or necrosis should be established for the exclusion of a malignancy. In the presence of massive lymphocytic infiltration of a leiomyoma the clonality of the infiltrate may aid in differentiating it from a malignant lymphoma. The pathogenesis and clinical significance of these rare neoplasms remain to be clarified.<sup>8</sup>

### Learning points

- ▶ Symplastic leiomyoma is an unusual variant of leiomyoma.
- ▶ Less likelihood for malignant transformation.
- ▶ Patient counselling is important to alleviate the anxiety associated with such histologic reports.

**Competing interests** None.

**Patient consent** Obtained.

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