

Uterine Leiomyosarcoma: A case Report

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Received: 10 Feb 2019

Accepted: 15 May 2019

DOI: <https://doi.org/10.3126/njog.v14i1.26633>

ABSTRACT

Leiomyosarcoma is a rare but aggressive tumor with poor clinical outcomes in compared to other uterine cancers regardless of its stage. The preoperative diagnosis of leiomyosarcoma is seldom made as the patients present with the symptoms similar to that leiomyoma following hysterectomy and myomectomy. Herein, we present a case of a 50 years old lady operated for broad ligament fibroid whose histopathology and immunohistochemistry report revealed leiomyosarcoma.

Keywords: : fibroid, histopathology, leiomyosarcoma.

INTRODUCTION

Leiomyosarcoma (LMS) is a malignant mesenchymal tumor that is derived from the smooth muscle lineage and accounts for about 3% of uterine malignancy. The median age for women with leiomyosarcoma is 43 to 53 years that is somewhat lower than other uterine sarcomas and premenopausal patients have a better chance of survival.¹ This malignancy has no relationship with parity. Incidence increases with age, African-American race, and prolonged use of tamoxifen of over 5 years.^{6,7} Incidence of sarcomatous change in benign uterine leiomyoma is 0.13-0.81%. There is a history of pelvic radiation in 4% of patients.¹⁻³ Diagnostic evaluation of leiomyosarcoma and distinguishing benign leiomyoma from its malignant counterpart is a clinical challenge as the symptoms are often similar.⁷ Abnormal uterine bleeding, pelvic pain or abdominal bloating may present in both disease processes. The standard treatment for early and advanced uterine leiomyosarcoma is hysterectomy and bilateral salpingo-oophorectomy and complete cytoreduction of the tumor en block. The mean survival rate for patients with uterine leiomyosarcoma is 47% (range=20-63%).¹

CASE

A 50 years old para 3 lady presented with chief complaints of stress urinary incontinence, lower abdominal mass and cessation of menstruation for 3 months, associated with weight loss and easy fatigue.

Per abdomen examination revealed a hard mass of 22 weeks size, mobile side to side, lower pole was not accessible and was non tender. On per speculum examination, cervix appeared normal and deviated to right side. On bimanual examination, no groove was felt between the mass and uterus. Ultrasonography showed a mixed echogenic mass of size 15.7x11x11.5 cm occupying whole of the uterus.

Total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. Intraoperative finding revealed a huge mass of about (15x10) cm in the left broad ligament with smooth surface, regular margin, extending up to the left pelvic side wall, up to the cardinal ligament downward and uterosacral ligament posteriorly. Uterus was about 6 weeks size and was pushed to right side by the mass. However, bilateral tubes and ovaries appeared normal. The fibroid weighed 1.5 kg and in cut section, fatty degeneration was present, uterine cavity and cervical canal appeared normal. In the formalin fixed sample, areas of necrosis were seen [Fig-1].

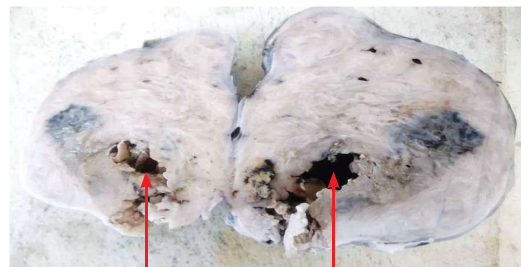


Fig-1: formalin fixed samle showing the areas of necrosis

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Histopathology examination revealed undifferentiated tumor cells with marked pleomorphism, intervening desmoplastic stroma with dense chronic inflammatory cells infiltrates with extensive areas of hemorrhage and necrosis, and atypical mitotic figures 1-3/10hpf noted. However, there was no lymphovascular involvement [Fig-2].

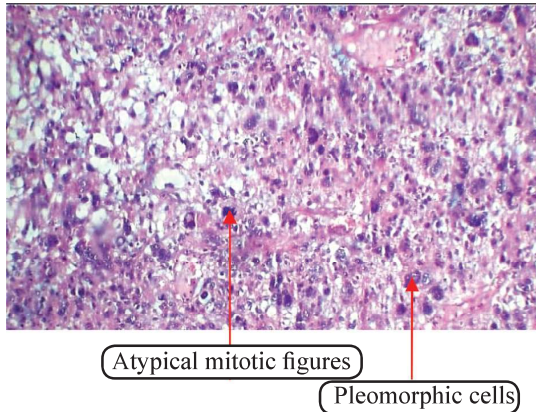


Fig-2: Pleomorphic variant of leiomyosarcoma

The diagnosis of leiomyosarcoma was further confirmed by immunohistochemistry, which showed positive staining for smooth muscle specific isoform of actin [Fig-3].

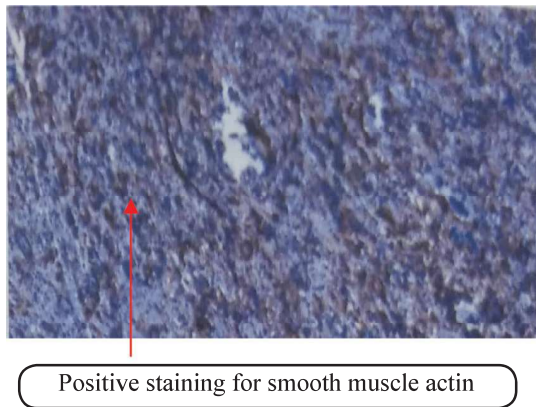


Fig-3: positive staining for smooth muscle actin

After confirming the diagnosis of leiomyosarcoma stage IIA, the patient was then referred to oncology department.

COMMENTS

Leiomyosarcoma comprises of approximately 30% of uterine sarcomas and about 3% of all uterine malignancies which can spread to pelvic blood vessels and lymphatic, contiguous pelvic structures, abdomen, and to distant organ, most often to the lung.^{1,3}

Although very rare, these are aggressive tumors with poor clinical outcome and risk of recurrence.⁴

It usually occurs in peri or postmenopausal age group with median age being 52 years, as in our patient who was perimenopausal age of 50 years. The presenting symptoms of leiomyosarcoma is usually of short duration (mean being 6 months) accompanied by pain, pressure sensation, abnormal uterine bleeding or a low abdominal mass and not specific to the disease. The principal physical finding is the presence of a pelvic mass.¹⁻³ similarly in this case, the patient presented with abdominal mass and a short clinical period of 3 months.

Diagnosing leiomyosarcoma preoperatively is a challenge for all clinician as there are no specific criteria that differentiate leiomyoma from leiomyosarcoma, even if diagnostic imaging and endometrial sampling have been performed preoperatively. There are no scientifically validated screening instruments that diagnose a LMS, the diagnosis of LMS is purely histological.⁴

Immunohistochemistry will stain positive for smooth muscle actin, desmin and caldesmon; and immunopositivity for p16 and p53 with a high Ki-67 proliferation index. As in this case, the presenting symptoms and clinical findings of the patient inclined us towards the diagnosis of leiomyoma and the final diagnosis was made postoperatively after histopathology and immunochemistry (positive staining for smooth muscle actin) reports like other published case reports.⁴⁻⁶ For LMS, there are 3 criteria for microscopic diagnosis: more than 5 mitoses/10 HPF, the presence of tumor necrosis, and moderate to severe cytologic atypia.⁵

Several clinical and pathological characteristics contribute to the prognosis of leiomyosarcoma. Specifically advanced age (>55 years), high tumor grade, larger tumor size (>5 cm), high mitotic index ($\geq 15/hpf$), and omission of bilateral oophorectomies. In addition to these, tumor morcellation are also observed to increases the local recurrence in the abdomen and pelvis which is associated with a shorter survival period.⁷

Primary treatment of leiomyosarcoma comprises of total abdominal hysterectomy with bilateral salphingo-oophorectomy. Radiation may be useful for local recurrence and chemotherapy for advanced and

recurrence of disease, although the survival benefit may not persist and long term survival or chance of cure remains low.^{8,9} The 5 year survival rate for stage I leiomyosarcoma is 50-55% and for stage II to IV is 8-12% only.^{9,10}

CONCLUSIONS

Since clinical presentation of leiomyosarcoma is similar to that of leiomyoma, when women presents

with rapidly enlarged lower abdominal mass with pain and pressure symptoms, leiomyosarcoma should always be kept in mind. Surgery is the mainstay of treatment. Confirmation of the diagnosis of leiomyosarcoma is only after histopathology and immunohistochemistry.

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