Rare case presentation of symplastic leiomyoma in nulliparous female

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ABSTRACT

We present a case of a 26–year-old female, nulligravida, who presented in outpatient department in our hospital with ultrasound suggestive of multiple huge fibroids at posterior wall and right lateral wall. Patient gave a history of myomectomy done in the past 2 years back. Histopathology was suggestive of benign leiomyoma. Ultrasound Doppler and magnetic resonance imaging were suggestive of multiple large size highly vascular fibroids in posterior and lateral wall of uterus of size $11 \text{ cm} \times 10 \text{ cm} \times 5 \text{ cm}$, compressing uterus and bladder with venous congestion. Intraoperatively, uterus was enlarged with multiple fibroids with sarcomatous changes in it. Largest of size $20 \text{ cm} \times 15 \text{ cm} \times 10 \text{ cm}$ with increased vascularity with ovaries and bowel adhered to uterus. Total abdominal hysterectomy with B/L pelvic lymph node dissection with omentectomy was done. Histopathology was suggestive of symplastic leiomyoma. Diagnosis was confirmed by immunohistochemistry as symplastic leiomyoma.

Key words: Myomectomy, sarcoma, symplastic leiomyoma

INTRODUCTION

Leiomyomas of the uterus (or uterine fibroids) are benign tumors that arise from the overgrowth of smooth muscle and connective tissue in the uterus. Histologically, a monoclonal proliferation of smooth muscle cells occurs. A genetic predisposition to leiomyoma growth exists. Calcified fibroids are often depicted on conventional radiographs of the pelvis. In some patients, magnetic resonance imaging (MRI) provides additional information. The role of computed tomography (CT) scanning is limited. Calcifications may be more visible on CT scans than on conventional radiographs because of the superior contrast differentiation achieved with CT scanning. Leiomyoma

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variants refer to a particular histological differentiation and growth pattern. Atypical and bizarre leiomyoma synonymous with symplastic leiomyoma are rare smooth muscle tumors that contain cells with moderate to severe cytological atypia, but cell necrosis is absent and mitotic index is fewer than 10/10 hpf. The features such as average age of the patient at presentation and maximum tumor size are identical to those of common leiomyoma, but clinical behavior and prognosis of these rare tumors depend on a number of mitotic figures.[1] Mitotic counts higher than 10 in such a tumor indicate a high malignant potential smooth muscle tumor (leiomyosarcoma).[2] Symplastic leiomyoma has been diagnosed in different sites other than uterus such as vagina, nasal cavity, and in scrotum. Rarely, uterine leiomyomas may undergo malignant degeneration to become a sarcoma. The true incidence of malignant transformation is difficult to determine because leiomyomas are common, whereas malignant leiomyosarcomas are rare and can arise *de novo*. The incidence of malignant degeneration is <1.0% and

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has been estimated to be as low as 0.2%. The preferred imaging modality for the evaluation of uterine fibroids is ultrasonography (US), specifically, transabdominal and transvaginal US. This case report describes the occurrence of symplastic leiomyoma in a young female with primary infertility.

CASE REPORT

A 26-year-old female, nulliparous married since 4 years presented to the outpatient department in our hospital with complaint of menorrhagia since two cycles with sonography report of private hospital showing multiple fibroids with largest being of 10 cm × 7 cm size. Patient also gave a history of open myomectomy done for removal of multiple fibroids in 2013 in corporation hospital largest being of size 20 cm × 10 cm in size.

On examination, patient was vitally stable. Her per abdomen examination was suggested of uterus of 22 weeks gestational size, firm to hard in consistency and solid with irregular margins, mobility restricted and nontender. Per speculum examination showed cervix pulled up and pinpoint. Per vaginal examination was confirmed the same abdominal findings with pulled up cervix and fullness in pouch of douglas.

Ultrasound findings suggestive of abdomen with Doppler was done which showed broad ligament fibroid with secondary sarcomatous changes with multiple omental deposits seen with increased vascularity.

MRI was also done showing multiple fibroids with largest being of $11 \text{ cm} \times 10 \text{ cm}$ compressing uterus and bladder and rectum with venous congestion seen [Figure 1].

Patient was taken for exploratory laparotomy. *In situ* findings were suggestive of uterus with multiple fibroids

Figure 1: Magnetic resonance imaging of symplastic leiomyoma

with largest being of 20 cm × 15 cm × 10 cm size, with irregular surface. Bilateral ovaries were bulky and adhered to uterus. Multiple bowel adhesions were seen. No omental deposits were seen. The surface of mass was highly vascular [Figure 2].

Total abdominal hysterectomy with bilateral salpingooophorectomy with B/L pelvic lymph node dissection with omentectomy was done [Figures 3 and 4]. On final histopathology report (HPR), diagnosis of symplastic leiomyoma was made which was confirmed on immunohistochemistry. It showed minimal mitotic activity which ruled out malignancy.

HPR was done suggestive of symplastic leiomyoma [Figure 5].

DISCUSSION

Leiomyomas of the uterus are one of the most common pathologic abnormalities of the female genital tract. Their occurrence increases with age, and they are found in 20-50% of women older than 30 years. Although found elsewhere in the body, leiomyomas most frequently occur in the myometrium. Uterine leiomyomas are commonly referred to as myomas, fibromyomas, or "fibroids" because of their firm, fibrous character, and high content of collagen. Approximately, 175,000 hysterectomies are performed annually for leiomyomas. Uterine leiomyomas result in masses associated with a variety of gynecologic problems, the most prominent of which are the asymptomatic pelvic mass or abnormal vaginal bleeding. The last two decades have seen rapid advancements in the diagnosis and treatment of uterine leiomyomas. The introduction of pelvic US, sonohysterography, hysteroscopy, and MRI have made it possible to detect small myomas not clinically suspected as a cause of abnormal bleeding. It is also possible to make a definitive diagnosis of a pelvic mass as a uterine myoma short of performing laparotomy or laparoscopy. Understanding the



Figure 2: Intra operative picture of huge leiomyoma

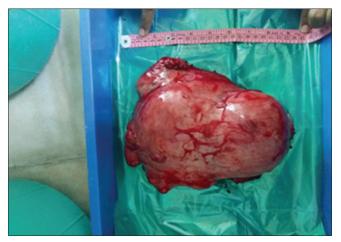


Figure 3: Gross specimen of leiomyoscarcoma



Figure 4: Cut section of leiomyoma

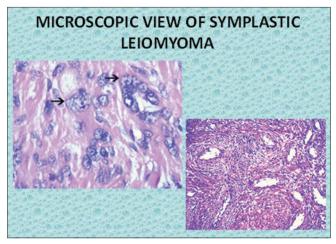


Figure 5: Microscopic view of symplastic leiomyoma

relation of sex steroid hormones and uterine leiomyomas has provided the foundation for using gonadotropin-releasing agonists, which have facilitated treatment of leiomyomas by hysteroscopy, laparoscopy, and the liberal use of vaginal hysterectomy. With the introduction of the antiprogesterone RU486 and other drugs that can be used in lieu of surgery, the

21st century will be marked by dramatically new treatments of leiomyomas of the uterus.[4]

Uterine sarcoma is a malignant uterine tumor, a very rare complication of leiomyoma that is composed of part or all sarcomatous (mesodermal) elements. They however account for a minority of all uterine malignancies. Most of the cases reported are of the postmenopausal age group and are usually present with postmenopausal bleeding or pelvic mass.^[5] Uterine rhabdomyosarcoma sometimes present in teenage age group with complications and life-threatening events.^[6]

They can be broadly classified as pure or mixed:

- Mixed
 - o Malignant mixed Mullerian tumor of the uterus: ~50–70% (most common of sarcomas)
 - o Mixed uterine leiomyosarcoma and endometrial stromal sarcoma
- Pure
 - o Uterine leiomyosarcoma: ~33–50%
 - o Endometrial stromal sarcoma: 10%
 - o Fibrosarcoma of the uterus: Rare
 - o Rhabdomyosarcoma of the uterus: Rare^[6]
 - Liposarcoma of the uterus: Rare.

In general, they are aggressive lesions characterized by early dissemination.

CONCLUSION

Atypical (symplastic) leiomyoma is a rare tumor of uterine smooth muscles. Histomorphological features of atypical (symplastic) leiomyoma closely mimic leiomyosarcoma, and immunohistochemical markers are certainly of help to exclude malignancy in case of dilemma.

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Conflicts of interest

There are no conflicts of interest.

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