Case Report

All uterine enlargements are not fibroids!

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ABSTRACT

Endometrial stromal sarcomas are rare mesenchymal tumors observed in pre- and peri-menopausal age group. The diagnosis is made postoperatively, but a high index of suspicion is needed. The usual presentation is lump in abdomen and abnormal vaginal bleeding. Endometrial sampling, ultrasound, and magnetic resonance imaging help in preoperative diagnosis. Total abdominal hysterectomy with bilateral salpingo-oophorectomy is the main line of management. This article describes its occurrence in a young woman, who presented without the usual clinical features of lump or abnormal vaginal bleeding. The diagnosis was established postoperatively and was confirmed with specialized tests such as immunohistochemistry and mutation studies.

Key words: Endometrial stromal sarcoma, pelvic tumor, uterine sarcomas

INTRODUCTION

Approximately, 2%–3% of women develop uterine cancer during their lifetime. Ninety-seven percent of all uterine cancers arise from the glands of the endometrium and are known as endometrial carcinomas. The remaining 3% of uterine cancers arise from mesenchymal uterine components and are classified as sarcomas. Endometrial stromal sarcomas (ESSs) are rare malignant tumors of the uterus, and most of the information available in literature is based on small series or case reports.

The annual incidence of ESS is 1–2 million women. Compared to other uterine malignancies, ESS affects younger women. ESS are uncommon mesenchymal tumors of the uterus which are composed of cells closely resembling normal proliferative endometrial stromal tissue.[1]

Traditionally, ESS was classified into low-grade and high-grade ESS based on mitotic rate. High-grade tumors, however, have little resemblance to original endometrial stroma. Therefore, high-grade tumors are presently classified as undifferentiated endometrial or uterine sarcoma. In this classification, the differentiation between low-grade and undifferentiated tumors is not made on mitotic count but the presence of nuclear pleomorphism and necrosis.[1] Permeative, infiltrative growth into the myometrium and the presence of vascular invasion are the main characteristics of ESS.[2] ESSs are hormone sensitive tumors. A state of hyperestrogenemia could act as a growth stimulus.

CASE REPORT

A 29-year-old single woman presented with complaints of heaviness in lower abdomen for 5 months. She had no menstrual complaints. On examination, she was underweight, pale with a mass arising from pelvis of uterine size of 20 weeks, nontender, immobile. Her Hb was 4 gm%. Her ultrasonography revealed multiple heterogeneous areas with calcified hyperechoic foci occupying the fundus, body and lower segment of the uterus. Magnetic resonance imaging reported bulky uterus and cervix completely replaced by large heterogeneous mass lesion with areas

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Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.197859

Cite this article as: Wani RJ, Shah SA, Khatib YS, Khade AL. All uterine enlargements are not fibroids!. Clin Cancer Investig J 2016;5:476-8.

of necrosis measuring 17 cm × 12.5 cm × 8 cm invading full thickness of myometrium with loss of junctional zone and invasion of serosa. She investigated and advised to go to a center with oncology backup. She insisted on care with us and underwent exploratory laparotomy with total abdominal hysterectomy (TAH) with bilateral salpingo-oophorectomy (BSO) with lymph node dissection. Intraoperatively the uterus was friable, and foul smelling. Specimen was cut open, and mass was extending down to the cervix.

The tumor on histopathology was suggestive of a high-grade sarcoma [Figure 1] involving full thickness of the myometrium. Uterine serosa, cervical cut margins, and omentum showed tumor deposits. On immunohistochemistry (IHC), the tumor cells were strongly and diffusely positive for CD10 [Figure 2], dot-like positivity for Mic-2 and weak expression for Fli2. CD10 is a marker for ESS and Mic-2 is a marker for atypical Ewing's tumor/primitive neuroectodermal tumor (PNET).

As the tumor showed dual positivity, translocation studies for EWSR1 and SYT were done to rule out PNET. Translocation studies for both the above markers were negative. Hence, the final diagnosis was given as ESS. She had a stormy postoperative course and was discharged after 3 weeks. She followed up at a specialized cancer center for chemoradiation.

DISCUSSION

Endometrial stromal tumors are composed purely of cells resembling normal endometrial stroma. They are divided into three types on the basis of mitotic figures, myometrial and vascular invasion, and observed differences in prognosis: (1) endometrial stromal nodule (ESN), (2) ESS, and (3) high-grade or undifferentiated sarcoma.[3]

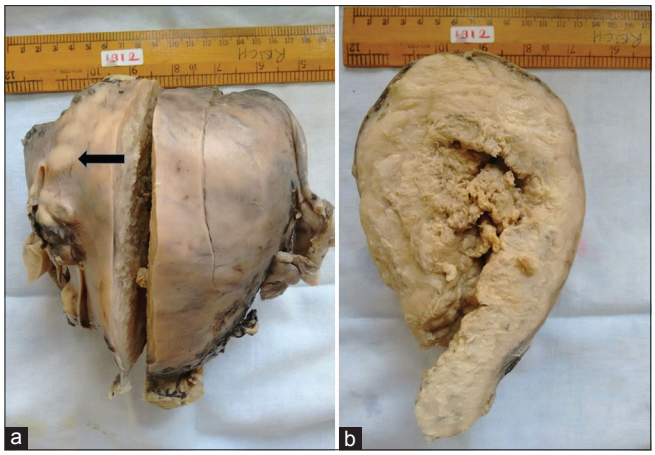

Figure 1: Gross photo of uterus with (a) enlarged uterus with serosal nodules (arrow), (b) cut surface of uterus showing infiltrative grayish white fleshy tumor involving uterus and cervix

The distinction between ESN and ESS is based on the findings of irregular tongue-like projections of myometrial invasion, >3 mm, and presence of endolymphatic invasion in ESS. ESS formerly termed as low-grade ESS is distinguished from high-grade ESS microscopically by a mitotic rate of <10 mitotic figures/10 high power field as well as clinically by a more protracted course. IHC helps in the detection of tumor markers specific for ESS. Strong and/or diffuse positivity for CD10 is found in ESS, which are helpful in distinguishing these tumors from histological mimics such as cellular leiomyoma and leiomyosarcoma that are generally negative.[4] They express h-caldesmon, desmin, and oxytocin receptors while CD10 and inhibin expression is a feature of ESS.[5]

Recurrences typically occur late, and local recurrence is more common than distant metastases.[6] Distant metastases occur in lungs.[7] Recurrence occurs in almost one-half of cases at an average interval of about 5 years after initial therapy. A beneficial effect of radiation therapy has been reported, and pelvic irradiation is recommended only for inadequately excised or locally recurrent pelvic disease.[8] The National Comprehensive Cancer Network Guidelines for ESS Stage I and II disease recommends TAH and BSO and thereafter only observation and follow-up of the patient. Follow-up includes patient education regarding symptoms of any recurrence, physical examination along with chest, abdomen, and pelvic imaging done 3 monthly for 2 years, thereafter repeated at 6–12 monthly interval.[9]

The ESS is a rare mesenchymal sarcoma of the uterus occurring in peri- and post-menopausal age group. The mean age is between 42 and 58 years. Review of literature has revealed only very few cases of ESS in the younger age group. Hence, a high index of suspicion is required

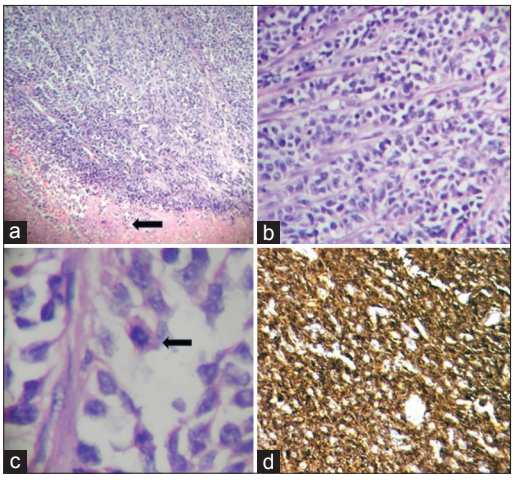

Figure 2: Microscopic photographs, (a) diffuse sheets of tumor cells with areas of necrosis (arrow) (H and E, ×40), (b) diffuse sheets of tumor cells with areas of necrosis (arrow) (H and E, ×100), (c) pleomorphic cells with abnormal mitosis (H and E, ×400), (d) tumor cells showing strong and diffuse CD10 positivity (immunohistochemistry × 100)

in young patients especially in the adolescent age group. The pathogenesis of ESS is unknown, but exposure to tamoxifen, unopposed estrogens, and polycystic ovaries are implicated.[10]

The tumor may be mistaken for leiomyoma uterus, and a conservative approach may be hazardous. Our case was a 28-year-old with a high-grade tumor with IHC suggesting atypical Ewings or PNET, which on special mutation studies was proven to be ESS.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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