Low grade endometrial stromal sarcoma in a premenopausal woman

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Abstract

Endometrial stromal sarcoma are rare malignant tumors of the uterus and has been described as the second most common malignant uterine mesenchymal tumor. The diagnosis is confirmed on hysterectomy for a presumed benign disease. In the latest World Health Organization (WHO) classification (2003), the term endometrial stromal tumor is applied to neoplasms typically composed of cells that morphologically resemble endometrial stromal cells of the nonneoplastic proliferative phase endometrium. The WHO recognizes three categories of endometrial stromal tumors: Endometrial stromal nodule, low-grade endometrial stromal sarcomas (LGESS), and undifferentiated endometrial sarcoma. We report here an interesting case of a 39-year-old female who presented with irregular bleeding per vaginum and urinary retention with a clinical impression of a leiomyomatous polyp, which on histological examination showed a LGESS.

Key words: Endometrial, low-grade, stromal sarcomas

INTRODUCTION

Endometrial stromal sarcoma (ESS) has been described as the second most common malignant uterine mesenchymal tumor and is said to account for approximately 10-15% of all uterine sarcomas.[1,2] Generally, these are very rare malignant tumors that make up for approximately 3-7% of all uterine cancers and only 1% of all female genital tract malignancies.[3] ESSs usually affect middle-aged women and commonly manifest as abnormal uterine bleeding, an enlarged uterus or pelvic pain. We report a case of a 39-year-old female who presented with irregular bleeding per vaginum and urinary retention. The clinical impression was that of a leiomyomatous polyp, whereas histological examination showed a low-grade endometrial stromal sarcoma (LGESS).

CASE REPORT

A 39-year-old female, reported with complaints of irregular bleeding per vaginum on and off for the past 2 months and retention of urine since the past 2 days. The patient was pale and ill looking. Per speculum examination revealed a large shaggy mass, 8 cm in diameter protruding out from the cervical os. The cervical rim was felt all around and the pedicle was high up. Uterus was 8 weeks in size, soft, anteverted, and both fornices were clear.

Ultrasound of the pelvis suggested a cervical mass lesion measuring 9 × 8 × 8 cm, well defined outline and heterogenous echogenicity. The uterus was bulky with the endometrial cavity pushed anteriorly and bilateral adnexae were normal. The clinical impression was that of a leiomyomatous polyp with the base in the posterior uterine wall. Polypectomy was done and sent for histopathological examination.

Gross examination of the polyp showed a globular mass measuring 6 × 5.5 × 4 cm. Cut surface was fleshy, grayish pink in color. [Figure 1] Outer surface had a ragged appearance. Microscopic examination of the polyp showed a cellular tumor with a close morphological resemblance to nonneoplastic proliferative-phase endometrial stromal cells. The tumor cells were small uniform looking with dark staining round to oval nuclei, granular chromatin, and inconspicuous nucleoli [Figure 2] Areas of sex cord-like differentiation [Figure 3] were also seen. Mitosis was minimal and less than three mitotic figures per high power field were seen. Stromal myometrial interface was not provided hence myometrial invasion could not be commented upon. These histological features were suggestive of endometrial stromal tumor and the possibilities considered were endometrial stromal nodule (ESN) and LGESS. Subsequently, 9 days after the polypectomy, the patient was operated again and a hysterectomy with bilateral salpingo-oophorectomy was undertaken. On laparotomy, uterus was noted to be enlarged to 8 weeks size, with bilateral cystic ovaries and no enlarged regional lymph nodes were present. A polyp was seen protruding into the endometrial cavity from posterior wall of the uterus which measured 4 × 5 × 3 cm, with a fleshy, gray-white cut surface [Figure 4]. Microscopic examination of the polyp showed a similar histological picture as described. These tumor cells were arranged concentrically around spiral arteriole like vessels with cord-like projections into the myometrium. Lymphovascular invasion was not seen. Both ovaries showed luteal cysts and the tumor was restricted to the uterine wall. Immunohistochemical analysis showed neoplastic endometrial stromal cells, which were immunoreactive for CD10 monoclonal antibody [Figure 5], while staining for Desmin was negative, thereby ruling out a smooth muscle origin. There were areas of sex cord-like differentiation that showed reactivity for Inhibin, which was not detected in the other stromal areas. The final histological diagnosis was LGESS with sex cord-like differentiation, International Federation of Gynecology and Obstetrics (FIGO) Stage IB.[4]

DISCUSSION

ESSs are rare tumors of mesenchymal origin. According to the latest World Health Organization (WHO) classification (2003), the term endometrial stromal tumor is applied to neoplasms typically composed of cells that morphologically resemble

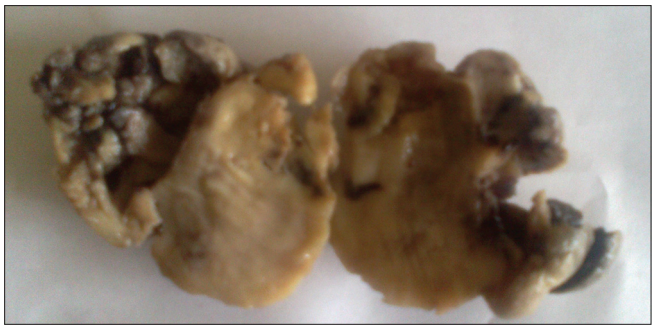

Figure 1: Gross photograph of the globular fleshy polyp

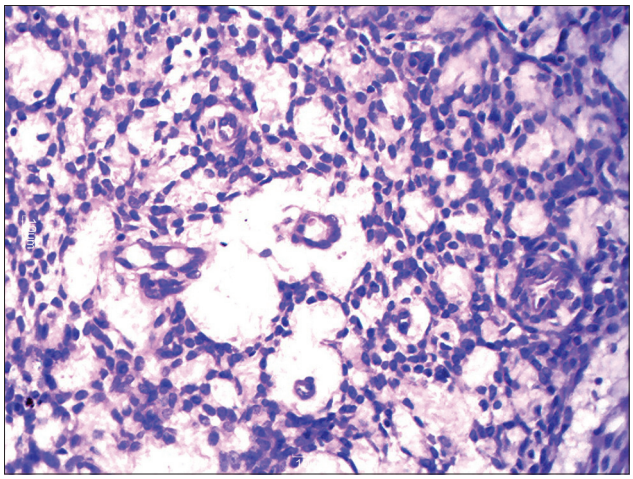

Figure 3: Photomicrograph showing areas of sex cord-like differentiation (H and E, ×40)

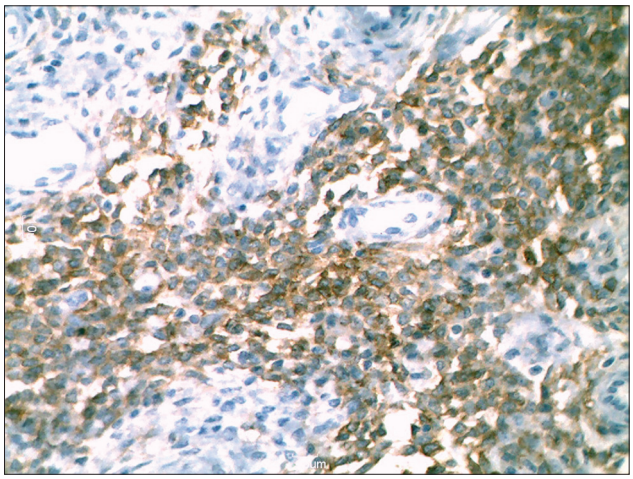

Figure 5: Immunohistochemical analysis showed neoplastic endometrial stromal cells immunoreactive for CD10 (IHC, ×40)

endometrial stromal cells of the nonneoplastic proliferative phase endometrium. The WHO recognizes three categories of endometrial stromal tumors: ESN, LGESSs, and undifferentiated endometrial sarcoma (UES).[5] Majority of these patients are premenopausal with an average age of 45 years and usually present with abnormal uterine bleeding.

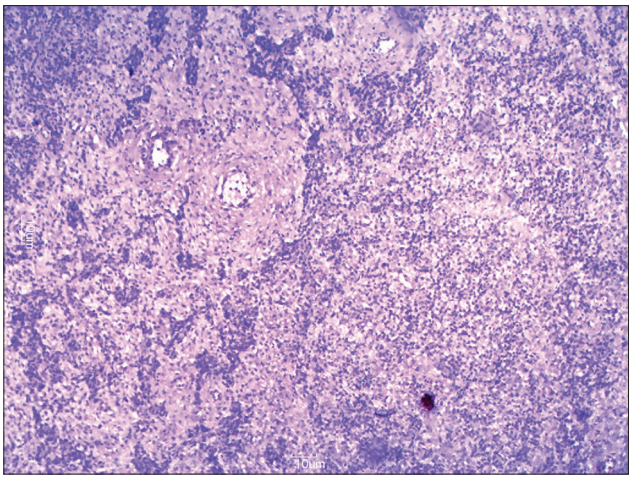

Figure 2: Microscopic examination showed a cellular tumor composed of small uniform looking cells with dark staining round to oval nuclei (H and E, ×10)

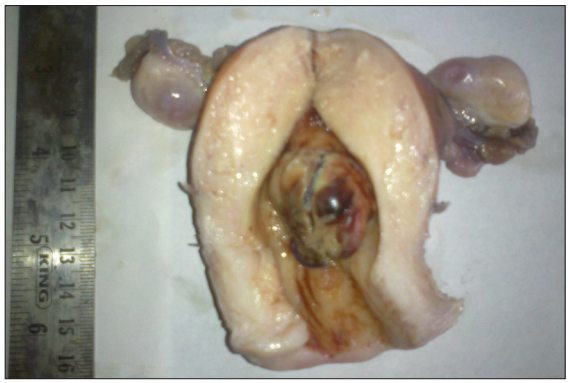

Figure 4: Gross photograph of the polyp protruding into the endometrial cavity from posterior wall of the uterus

[6] Our patient was a 39-year-old female with an interesting clinical presentation of acute onset urinary retention with metrorrhagia, for which she came to medical attention and a polypoid lesion distending the cervical os was detected. The clinical impression was a leiomyomatous polyp and polypectomy was done.

In a review of 10 cases of LGESS, Landreat *et al.* noted that the common presentations were abnormal uterine bleeding and myoma-related complications like pelvic pain, pressure symptoms, and urinary abnormalities, the initial clinical impression was usually a leiomyoma.[7] The diagnosis in these cases of endometrial stromal tumors is often made by chance, from either a myomectomy or a curettage specimen. Similar findings have been found in other large studies of endometrial stromal tumors.[1,6] In the present case, the base of the polyp remained high up in the uterine cavity and was not removed in the polypectomy, hence no comment on myometrial invasion was possible. In most cases, it is impossible to differentiate between an ESN and a LGESS on the basis of curettage specimens and, thus, distinction can

only be confidently established in a hysterectomy specimen. The smooth, expansile, noninfiltrating margins of the ESN contrasts with the infiltrating irregular margin of ESS and is the most important histological criterion in distinguishing between these two categories of endometrial stromal tumors. Focal irregularities in the form of lobulated or finger-like projections into the adjacent myometrium that are not >3 mm and are not exceeding three in number, however, may be seen in ESN.[8] Vascular invasion, if present leads to a diagnosis of LGESS. Dionigi *et al.* in a study of 50 cases of ESN descriptively referred to a subgroup of tumors as "endometrial stromal tumors with limited infiltration".[9] These tumors had a greater irregularity of their margins as compared with the laid down guidelines, but without the typical and commonly extensive infiltration seen in ESS. Also, the biological behavior of the tumors was similar to ESN. Apart from the classical histological picture of small cells resembling endometrial stromal cells, low grade sarcomas may show features of differentiation, including smooth muscle and sex cord differentiation. In the latter, the tumor contains epithelial or sex cord-like elements arranged in nests, cords, trabeculae, solid, or tubular structures. Similar areas were seen in this case, with epithelial appearing cells forming cords and trabeculae but they were small and the majority of the tumor showed sheets of small blue stromal cells with a concentric arrangement at places. Clement and Scully first described Uterine Tumor Resembling an Ovarian Sex Cord Stromal Tumor (UTROSCT) in 1976.[10] UTROSCT is defined as a tumor with prominent sex cord-like differentiation in which there is no conspicuous endometrial stromal background. The clinical presentation and the gross appearance of both LGESS with sex cord-like differentiation and UTROSCT is similar. But it is important for the pathologist to differentiate UTROSCT from LGESS with sex cord-like differentiation as the biological behavior of these lesions differ markedly. Frequent recurrences and metastases to the pelvic organs have been reported in LGESS, while UTROSCT behaves in a benign fashion.[6,11]

In conclusion, LGESS is a rare malignant tumor in which differentiation from ESN and also from UTROSCT, when evidence of sex cord differentiation is present is mandatory. Prognosis and outcomes greatly differ and a correct histological diagnosis plays an important role in the subsequent follow up and management of such cases.

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