High grade endometrial stromal sarcoma is a rare endometrial malignancy. This case report is aimed to present a case of high grade endometrial stromal sarcoma because of its rarity. This report is peculiar due to the metastatic involvement of ovary and retroperitoneal nodes in high grade endometrial stromal sarcoma. Surgical staging may be necessary in uterine sarcomas similar to that done in endometrial carcinoma.

**Keyword:** Endometrial stromal sarcoma, ovary, lymph node

**INTRODUCTION**

Uterine sarcoma accounts for 3 to 9 percent of all uterine malignant neoplasms [1]. Endometrial stromal tumors are a subset of uterine mesenchymal tumors that account for less than 10 percent of uterine sarcomas and approximately 1 percent of all uterine malignancies [2]. Endometrial stromal sarcomas affect younger women between the ages of 42-58 years who typically present with abnormal uterine bleeding and uterine enlargement, with associated dysmenorrhea and pelvic pain [3].

**CASE REPORT**

A 36 year female (para 2, live 2) evaluated for abnormal uterine bleeding and pelvic pain 5 months with ultrasound and MRI abdomen and pelvis which showed endometrial thickness 6 mm and endometrial polyp and fractional curettage showed poorly differentiated carcinoma and diagnosed as uterine malignancy. Patient underwent surgical staging – type 1 hysterectomy, bilateral salpingoophorectomy, bilateral pelvic lymph node dissection and paraaortic node sampling. Definitive histopathology showed myometrial invasion, cellular and nuclear pleomorphism and mitotic count >10/ HPF which confirmed high grade endometrial stromal sarcoma (undifferentiated endometrial sarcoma, according to WHO classification), one of the ovaries showed metastasis, 10 out of 13 pelvic nodes and 3 para-aortic nodes were positive for malignancy and staged as III C. Considering the age of the patient and no level 1 evidence for adjuvant therapy, patient was planned for observation as per tumor board policy. Patient is on follow up for past 3 months without clinical evidence of disease.
HISTOPATHOLOGY - HIGH GRADE ENDOMETRIAL SARCOMA

HISTOPATHOLOGY - SPINDLE CELLS WITH HIGH MITOSIS

HISTOPATHOLOGY- OVARIAN METASTASES

DISCUSSION

Uterine sarcomas fall into two broad categories histologically: mesenchymal tumors and mixed mesenchymal and epithelial tumors. Mesenchymal tumors include leiomyosarcomas (LMS), endometrial stromal sarcomas (ESS), and smooth muscle tumors of uncertain potential (STUMP), as well as mixed endometrial stromal and smooth muscle tumors. According to the WHO classification [4], the term endometrial stromal tumor is applied to neoplasms typically composed of cells that resemble endometrial stromal cells of the proliferative endometrium. The World Health Organization classifies endometrial stromal tumors into three categories: endometrial stromal nodule (ESN), endometrial stromal sarcoma (ESS), and undifferentiated endometrial sarcoma (UES).

ESN is a benign entity treated with simple hysterectomy. The main distinguishing feature of endometrial stromal nodules is their expansile, non-infiltrating, smooth margin that contrasts with the infiltrating irregular margin of stromal sarcomas [5]. Endometrial stromal sarcoma (ESS) refers to low grade stromal sarcomas composed of cells resembling the endometrial stroma in its proliferative phase, with infiltrating borders invading the myometrium and myometrial vessels. Clinically, low-grade ESS displays indolent behavior that mirrors its low-grade histologic appearance [5]. Undifferentiated endometrial sarcoma (UES) refers to high-grade ESS having marked cellular pleomorphism and brisk mitotic activity, in addition to destructive myometrial invasion [5]. Preoperative imaging is needed for surgical planning, as ESS can metastasize impressively throughout the vascular and lymphatic system, as well as along the peritoneum. The preoperative diagnosis of ESS has long been challenging, with pathological features similar to that of normal myometrium and findings on ultrasound and CT similar that may resemble uterine leiomyomata. On ultrasonography, ESS may present with irregular margins and vascularization, which may prompt further radiological evaluation in the form of MRI. ESS may present on MRI as an invasive endometrial or myometrial mass with tumor extension into the myometrium or into vascular or lymphatic structures.

The standard approach to surgical treatment for ESS is hysterectomy and bilateral salpingo-oophorectomy. The role of nodal dissection is unresolved, because of scarcity of data. Lymphatic involvement of ESS is well established, ranging from 7% to 8.9% [6]. While removal of enlarged lymph nodes may be done as part of a cytoreductive procedure, a survival benefit has not been proven [7].

Postoperative hormonal therapy is recommended for ESS stage II – IVA and palliative radiotherapy for stage IV B [8]. Although hormonal therapy is recommended for ESS, it is not recommended in high grade endometrial stromal sarcoma. Patients diagnosed as high grade endometrial stromal sarcoma were encouraged to enroll in clinical trials.

CONCLUSION

In the present case the disease had occurred at earlier age, 36 years which was younger than reported in literature (42-58 years) [3], ovarian metastases, pelvic and para-aortic lymph nodal metastases were also present which are rare occurrence in endometrial stromal sarcoma. Even though diagnosis was high grade endometrial sarcoma stage III C, patient was put on observation. Surgical staging was the only definitive method to stage uterine malignancy either carcinoma or sarcoma and to decide on adjuvant therapy, if any. Adjuvant cytotoxic chemotherapy may be tried after surgery for high grade endometrial sarcoma. Prospective trials are needed to address the issue of lymphadenectomy and adjuvant therapy in uterine sarcomas.

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