The Journal of Obstetrics and Gynecology of India (July-August 2021) 71(4):448–451
https://doi.org/10.1007/s13224-021-01442-9

CASE REPORT

Endometrial Stromal Sarcoma, an Unusual Recurrence: A Case Report

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Received: 12 October 2020 / Accepted: 12 January 2021 / Published online: 3 March 2021

Introduction

Sarcomas account for < 10% of uterine cancers [1, 2]. ESS belongs to the class of homologues uterine sarcomas as they consists only of elements normally found in uterine tissue. Based on WHO 2014 classification ESS is classified into 5 types—Endometrial stromal nodule (ESN), Low-grade endometrial stromal sarcoma (LG-ESS), High-grade endometrial stromal sarcoma (HG-ESS), Undifferentiated uterine sarcoma (UUS), Uterine tumour resembling ovarian sex cord tumour (UTROSCT) [3]. LG-ESS are low grade sarcomas with metastatic potential. Their ability to invade myometrium or vascular structures differentiates them from ESN. Usual age of incidence is above 40 years (40–50 Years). Pre-operative differential diagnosis includes fibroid uterus, endometrial carcinoma, and endometrial sarcoma as all these entities present with similar clinical picture of abnormal uterine bleeding and pelvic pain. Usually low grade ESS has an indolent course with late recurrences as high as about 50% [4].

Case Report

A 39-year-old multiparous lady underwent total abdominal hysterectomy and bilateral salpingo oophorectomy in 2009 for fibroid uterus at local hospital. Her complaint was abnormal uterine bleeding. Histopathological report was high grade endometrial sarcoma. Patient received adjuvant radiotherapy (EBRT 46 Gy 23# and brachytherapy 7 Gy 3 #) and chemotherapy with Ifosfamide and Adriamycin. Patient was kept on routine follow up with symptom review and clinical examination. Following a disease free interval of 8 years, the patient presented with vague abdominal pain. Evaluated with ultrasonography which showed an elongated mass over the psoas muscle. Hence referred to our institute.

On clinical examination patient's performance status was good and there was no palpable mass or nodes. CT scan showed a well-defined heterogeneously enhancing tubular lesion with peripheral enhancement extending from paracaval region to pelvis, in the location of right ovarian vein. It measures 30 × 32 mm in axial dimension and 12 cm craniocaudally Figure 1.

Trucut biopsy from the mass showed ESS low grade. Multidisciplinary tumour board discussed and decided for secondary cytoreduction. Intraoperatively there was no ascites, 12 × 3 cm tumour mass extending along the right ovarian vein from the level of external iliac vein up to inferior venacava, sparing distal 1 cm. Tumour mass was abutting right psoas major. Right common iliac node was enlarged. Tumour mass dissected along with debulking of enlarged right common iliac node Figs. 2, 3.

Post-operative period was uneventful and patient discharged on post-operative day 4.

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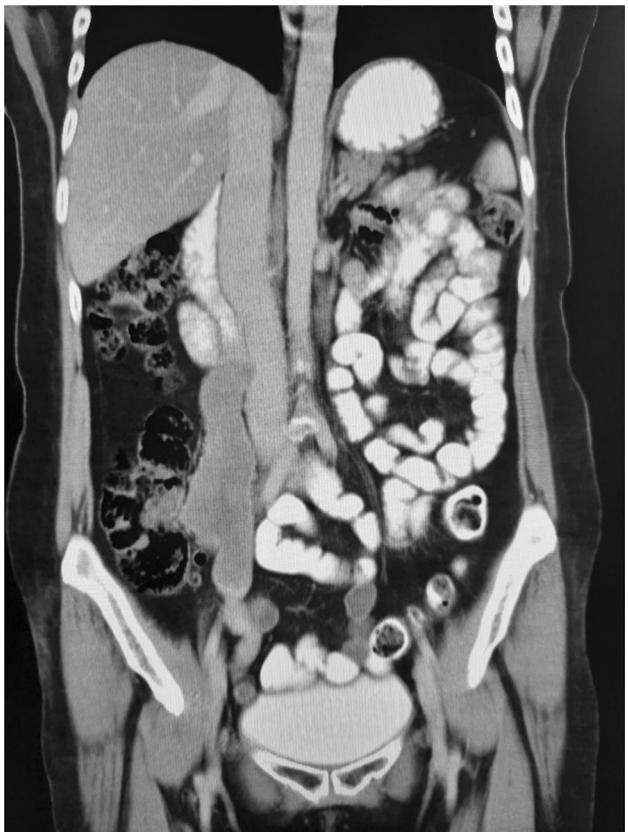

Fig. 1 CT scan showing tumour extending from pelvis to para caval region

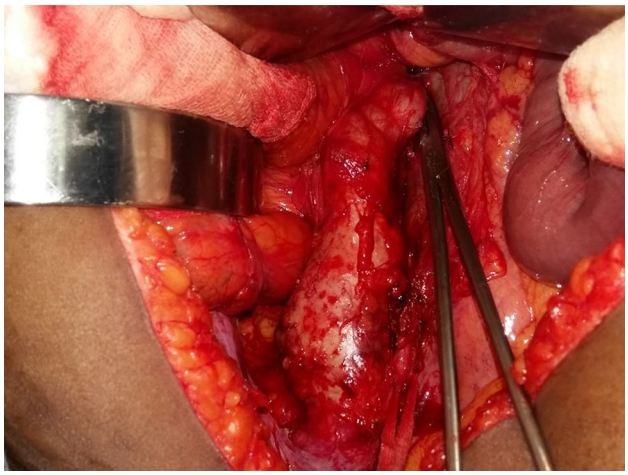

Fig. 2 Intraoperative picture showing tumour recurrence

Histopathology Report

Cut section showed grey white and grey brown tissues with central cystic change and glistening areas. Microscopy showed sheets of oval to spindly cells having oval hyper chromatic mildly pleomorphic nucleus with intervening plenty of tiny vascular channels (Fig. 4). Stroma in focal area showing hyalinisation and necrosis. Occasional mitosis present. MIB-1 labelling index 10–15%; Cyclin D1—weak nuclear positivity (< 20%). ER—positive. MIB-1 labelling index was 12–15% (Fig. 5). Lymph node showed reactive changes only. DIAGNOSIS**:** Endometrial stromal sarcoma—low grade.

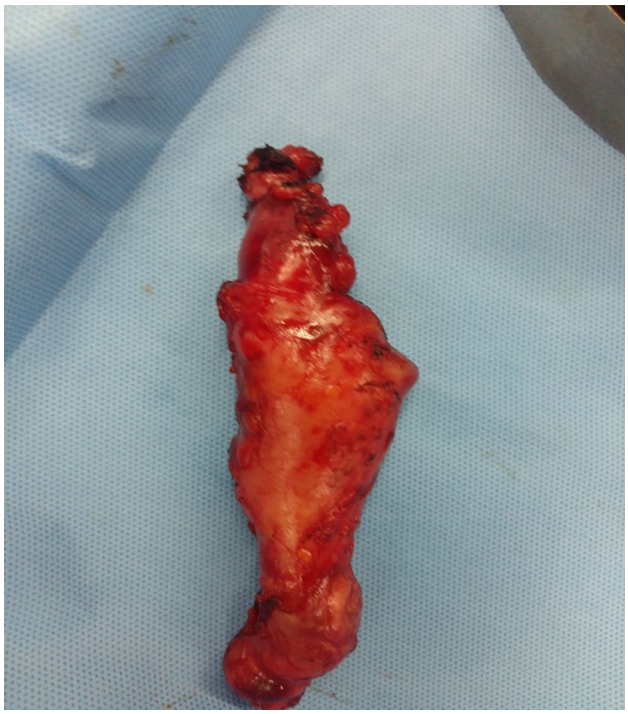

Fig. 3 Specimen picture

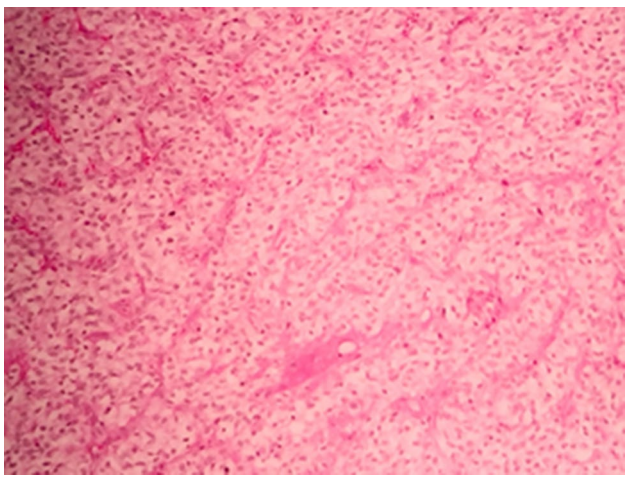

Fig. 4 Ess-H&E(100x) Section shows uniform round or oval tumour cells with abundant vessels

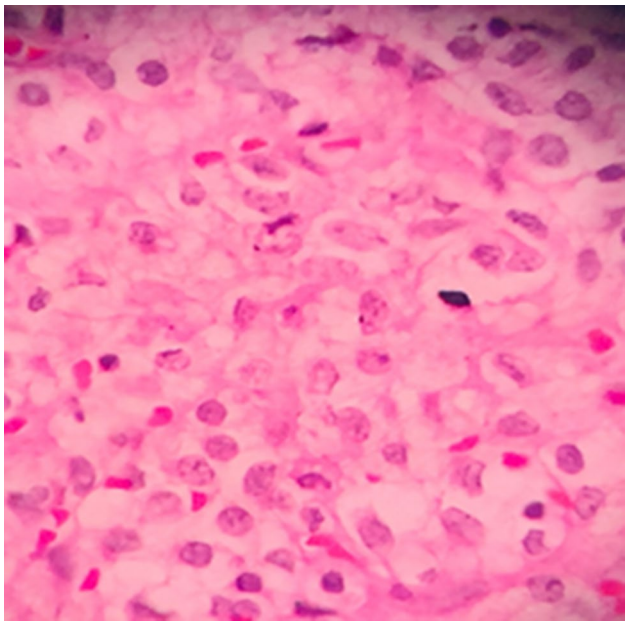

Fig. 5 Ess-H&E(400x)–Nucleus are pleomorphic vesicular coarse chromatin and nucleoli

As the clinical features (indolent course of disease) and intraoperative findings are typical of low grade ESS recurrence, request given for slide review of original TAH + BSO. Review of slides in our institute confirmed it as low grade ESS. Multidisciplinary tumour board discussed and decided for adjuvant hormone treatment Medroxy progesterone acetate) in view of final histopathological diagnosis of ESS low grade. Patient took adjuvant treatment and now disease free for last 3 years.

Discussion

Endometrial stromal sarcoma is a rare uterine sarcoma with characteristic molecular markers like immune reactivity for oestrogen, progesterone and androgen receptors. Jeoj youl park demonstrated better survival outcome with triple receptor positive ESS and worst outcome with triple negative ESS [5]. They are typically positive for CD 10 and smooth muscle actin and are negative for h- caldesmon and histone deacetylase 8 (HDAC8).

Cytogenetic analysis of ESS showed chromosomal translocations involving 6p, 7p and 17q. Most common translocation is between short arm of chromosome 7 and long arm of chromosome 17.This translocation results in fusion of two zinc finger genes JAZF1 and JJAZ1. This gene fusion explains the increased cellular proliferation in LG- ESS [6–8].

Surgical management of LG—ESS is hysterectomy along with bilateral salpingo oopherectomy as these tumours are hormone dependent. For patients with extrauterine disease cytoreduction is recommended but a study by Leath et al. [9] showed no survival advantage with extensive cytoreduction. Preservation of ovaries in premenopausal patients is associated with increased recurrence rate in certain studies [10–12] but a Surveillance Epidemiology and End Results (SEER) analysis did not show any adverse effects on overall survival when ovaries were preserved in premenopausal patients [13]. Role of systematic lymphadenectomy is controversial [14, 15]. Reported incidence of lymph node metastasis is around 10%.Eventhough lymph node metastasis is associated with poor survival outcome Chan JK et al. [1] demonstrated no survival advantage with routine lymphadenectomy.

For stage I patients only surveillance is recommended. For stage II to IV NCCN [16] recommends adjuvant hormone treatment on the basis of receptor status. Adjuvant radiotherapy can be given to reduce locoregional recurrence.

LG—ESS recurs most commonly in abdomen or pelvis, followed by lung [16]. Surgical resection is feasible for solitary metastasis. Many cases reported long term survival following surgical resection of ESS recurrences [17, 18]. Patients with increased disease free interval were reported to have improved survival outcome.

Conclusion

This is an unique case report of Endometrial stromal sarcoma recurrence—recurrence via right ovarian vein, 8 years post hysterectomy and bilateral salpingo oophorectomy. Here we intent to review the clinical, molecular, cytological features of low grade ESS. Here by we declare no conflict of interest among authors.

Compliance with Ethical Standards

Conflict of interest The authors have no conflicts of interest relevant to this article to disclose.

Informed Consent Informed consent was obtained from patient party.

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