Original Research Paper

Volume - 11 | Issue - 06 | June - 2021 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Pathology

EXTRAUTERINE LOW GRADE ENDOMETRIAL STROMAL SARCOMA – A RARE CASE REPORT

Dr. J. Suresh Durai MD(Pathology), Consultant Pathologist, Histolab Tirunelveli, Tamilnadu, India - 627109.

Dr. R. Chandhru Mari* MD(Pathology), Consultant Pathologist, Histolab Tirunelveli, Tamilnadu, India - 627109. *Corresponding Author

Dr. Jasmine Rajesh MBBS., DGO., Consultant Obstetrician & Gynaecologist, Rajesh Tilak Hospital, Thoothukudi – 628008.

ABSTRACT Endometrial stromal sarcoma (ESS) is a rare neoplasm accounting for only 0.2% of female genital tract tumors. The extra-uterine ESS is an extremely uncommon in younger age groups. Most reported cases of extra-uterine ESS (EESS) were found to be associated with foci of endometriosis. ESS may involve various sites such as peritoneum, ovary, vagina, colon, small bowel and stomach. We report a case of 24-year-old married nulliparous woman presenting with a left paraovarian mass. Laprotomy was done and histopathological examination of the resected specimen showed feature of primary extra-uterine Low grade ESS . Immunohistochemistry was done to confirm the diagnosis. Low grade ESS has histopathological features and clinical behavior similar to uterine ESS . Awareness about this neoplasm and a careful histopathological and immunohistochemical evaluation are essential for diagnosis .

KEYWORDS : Endometrial stromal sarcoma, Low grade ESS, Immunohistochemistry, Paraovarian mass

INTRODUCTION

Endometrial stromal sarcoma (ESS) is a rare neoplasm found in the uterus, however it can arise elsewhere posing significant diagnostic challenges(1) . Most reported cases of extra-uterine ESS (EESS) were found to be associated with foci of endometriosis [2]. Many organs could be involved, such as peritoneum, ovary, vagina, colon, small bowel and stomach. Often the diagnosis of a primary ESS is made when the uterus is free of tumor as it is the primary site of ESS [3]. Depending on mitotic activity, vascular invasion or prognosis, there are three categories of endometrial stromal tumors: endometrial stromal nodule, low-grade endometrial stromal sarcoma, high-grade endometrial stromal sarcoma We report a case of a 24-year-old married nulliparous woman presenting with left paraovarian mass diagnosed histologically as low grade ESS .

Case History

A 24-year-old woman presented to OPD with abdominal pain and abnormal vaginal bleeding. Ultrasonography was done and reported as uterus with 8 mm thick endometrium. Both ovaries were normal in size and echogenicity. A paraovarian mass measuring 7 x 6 x6 cm was noted. Magnetic resonance imaging (MRI) was also performed and reported as Left Broad ligament mass. The mass was adherent to left parametrium . Uterus ,endometrial cavity ,both ovaries and fallopian tubes were normal .The patient underwent laparotomy with removal of paraovarian mass . Peroperatively the tumor was friable in focal areas and removed in piecemeal.

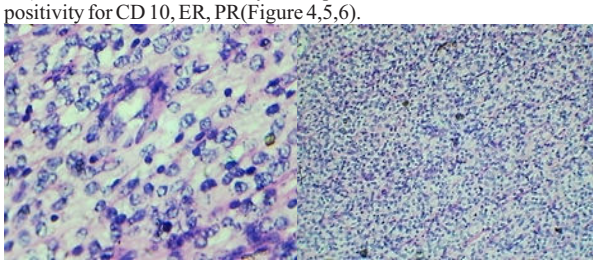
Macroscopic Examination

Grossly, Received multiple fragments of greyish soft tissue fragments totally measuring 7 x 6 x 6 cm (Figure 1).

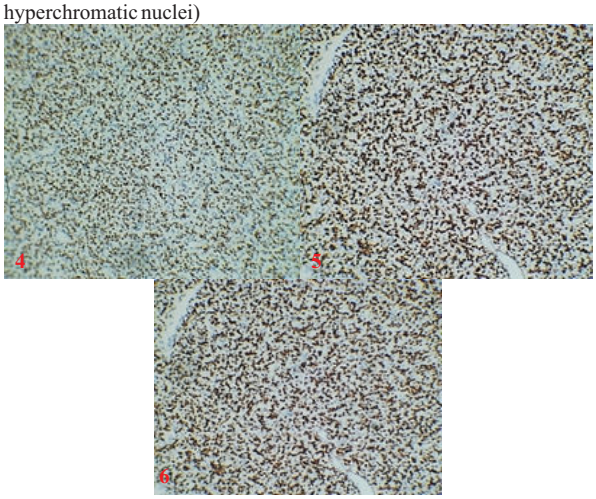
(Fig-1 Macroscopic appearance of the specimen)

Microscopic Examination

The histological examination of the sections studied showed sheets of proliferating oval to spindle cells with hyperchromatic nuclei and moderate cytoplasm , and scanty mitotic figures (2/10 high-power fields). The stroma showed numerous small arterioles with some showing hyalinazed vessel walls. Prominent whorling pattern of tumor cells around these vessels was observed in multiple sections(Figure 2,3) . Immunohistochemistry was performed and shows diffuse positivity for CD 10, ER, PR(Figure 4,5,6).



(**Fig -2,3 -** Sheets of proliferating oval to spindle cells with hyperchromatic nuclei)



(**Fig- 4,5,6** –ER,PR,CD10 positivity)

DISCUSSION

Endometrial stromal sarcoma usually affects women of postmenopausal age groups (45-57 years)[6]. Usually patients present with abnormal vaginal bleeding, progressive menorrhagia, and abdominal pain. It can be mistaken for leiomyoma clinically making it difficult to diagnose, hence diagnosis is usually made on postoperative histopathological examination. Extra-uterine endometrial stromal

sarcoma (EESS) is an extremely uncommon entity as available in the current literature [2]. The clinical presentation of EESS is highly variable depending upon the site and size . Our current case presented initially with a huge abdominopelvic mass, with a radiological diagnosis of leiomyoma.

Typically, ESS presents as a neoplasm that resembles proliferative phase endometrial stroma, with diffuse architecture and monomorphic cells with oval to spindle nuclei and variable mitotic count .The tumor stroma has a rich vascular network of small vessels sometimes with hyalinized walls, reminiscent of endometrial spiral arterioles, and the tumor cells are frequently arranged in a whorling pattern around these vessels [3]. The tumor borders are usually irregular with vascular invasion and tongue-like projections into vessels wall.

Endometrial stromal sarcoma can be a diagnostic challenge when it arises in abdominal extrauterine locations . ESS should enter the differential diagnosis along with other common spindle cell tumors of the abdominal cavity such as GIST and leiomyosarcoma. Immunohistochemistry plays an important role in distinguishing these tumors. ESS stains positive for CD10, vimentin, WT-1, ER, PR, and negative for SMA, desmin, CD34, CD31, inhibin, calretinin[4].

The extra-uterine locations of ESS make the diagnosis very challenging for both clinicians and pathologists. A part from the extra-uterine locations, these erroneous diagnosis could be in part due to many changes that occurred in histologic classification of ESS during recent years[5,6]. Our case has been misdiagnosed clinically and radiologically as Broad ligament leiomyoma with degenerative changes . However the characteristic microscopic and immunohistochemical findings lead to correct diagnosis. To claim the diagnosis of EESS, the status of the uterus should be determined by imaging techniques or by a thorough macroscopic sampling when the uterus is resected [7].

Our patient had no clinical, radiological or macroscopic evidence of any uterine lesions and histomorphologic features were characteristic of low grade ESS. The immunohistochemical phenotype was also compatible with ESS (positivity of ER, PR, CD10) [8]. The low grade extra-uterine endometrioid stromal sarcoma (EESS) is considered as an indolent neoplasm with late recurrences [9].

CONCLUSION

This case is presented for the rarity of primary extrauterine ESS occurring in abdominopelvic cavity and rarely without an obvious foci of endometriosis. Awareness about extrauterine ESS and careful histopathological and immunohistochemical evaluation should be part of the differential diagnoses of any abdominopelvic spindle cell tumor.

REFERENCES

1. Masand RP, Euscher ED, Deavers MT, Malpica A. Endometrioid stromal sarcoma: a clinicopathologic study of 63 cases Am J Surg Pathol. 2013;37:1635–47.
2. Back JA, Choi MG, Ju UC, Kang WD, Kim SM. A case of advanced-stage endometrial stromal sarcoma of the ovary arising from endometriosis. Obstet Gynecol Sci. 2016;59:323–7.
3. Oliva E, Egger J-F, Young RH. Primary endometrioid stromal sarcoma of the ovary: a clinicopathologic study of 27 cases with morphologic and behavioral features similar to those of uterine low-grade endometrial stromal sarcoma. Am J Surg Pathol. 2014;38:305–15.
4. Nucci MR. Practical issues related to uterine pathology: endometrial stromal tumors. Mod Pathol. 2016;29(Suppl 1):S92–103.
5. Xie W, Bi X, Cao D, Yang J, Shen K, You Y. Primary endometrioid stromal sarcomas of the ovary: a clinicopathological study of 14 cases with a review of the literature. Oncotarget. 2017;8:63345–52
6. Jain R, Batra S, Ahmad A, Elahi AA, Gupta M, Saith P. Low grade endometrial stromal sarcoma: a case report. Iranian journal of medical sciences. 2015 Jan;40(1):81.
7. Leung F, Terzibachian JJ, Aouar Z, Govyadovskiy A, Lassabe C. Uterine Sarcomas: clinical and histopathological aspects. Report on 15 cases. Gynecol Obstet Fertil. 2008;36:628-35. PubMed PMID:18538624
8. Ashraf-Ganjoei T, Behtash N, Shariat M, Mosavi A. Low Grade Endometrial Stromal Sarcoma of uterine corpus, a clinicpathological and survey study in 14 cases. World J Surg Oncol. 2006;4:50. PubMed PMID: 16895611; PubMed Central PMCID:PMC1560376
9. Devi PU, Himaja S, Manasa R, Atla BL. Uterine low grade endometrial stromal sarcoma: a case report with review of literature. International Journal of Research in Medical Sciences. 2017 Jan 12;3(9):2510-3.