Vaginal Endometrial Stroma Sarcoma: A Case Report of A Rare Disease

Abstract

Vaginal endometrial stromal sarcoma (VESS) arising in the vagina is an extremely rare extrauterine endometrial stromal sarcoma, Stroma Sarcoma. To the best of our knowledge, there have been only a few reported cases in the literature. Therefore, we herein report a case of VESS originating in the vagina. A 27-year-old woman complained of heavy and intermittent vaginal bleeding and abdominal swelling all of six months duration. A mass that was firm to hard in consistency was found in the middle and upper segments of the anterior, right lateral, and posterior fornices and wall of the vagina. An excisional biopsy was taken and the immunohistochemistry result revealed VESS. A cystoscopy was done which revealed polypoid metastatic nodules in the bladder (this put the stage of the disease at IV). She also had laparotomy following features of mechanical obstruction from previous pelvic and intraperitoneal adhesions which were formed from previous laparotomy.

Keywords: *Endometrial stromal, immunohistochemistry, vaginal sarcoma*

Benjamin Howe Nongo[1], Dennis Anthony Isah[1,2], Abimiku Bawa Ahmed[2,3], Oluwasesan Adelowo Abdul[3]

[1]Department of Obstetrics and Gynaecology, University of Abuja Teaching Hospital, Abuja, [2]College of Health Sciences, University of Abuja, Abuja, [3]Department of Histopathology, University of Abuja Teaching Hospital, Abuja, Nigeri

Introduction

Endometrial stromal sarcoma (ESS) is a rare neoplasm, it accounts for only 0.2% of all uterine malignancies.[1] It is even less likely to be found in extrauterine sites such as the vaginal. In a few cases, ESS arise primarily in extrauterine sites such as pelvic cavity, ovary, abdominal cavity, fallopian tube, retroperitoneum, vagina, and vulva,[2] of the above extrauterine sites, the vagina is an extremely rare site. To the best of our knowledge, current literature has shown that only 13 cases of extrauterine ESS have been reported.[1]

The initial planned total abdominal hysterectomy (TAH) and partial vaginectomy were shelved. An excision of the suspicious areas of the myometrium, the endometrium and the right ovary did not reveal any ESS lesion. She was eventually referred to another teaching hospital that has the facilities for radiotherapy to have combined chemoradiation treatment. The diagnosis of vaginal endometrial stromal sarcoma (VESS) largely depends on pathological examination of specimens taken from the mass because it has no typical symptoms. The mainstay of treatment for VESS is surgery, however, the following may also be incorporated: chemotherapy, radiotherapy, and hormonal therapy. The prognostic factors for VESS may include the tumour stage and the expression of hormonal receptors.

Case Report

A 27-year-old P_0^{+1} was admitted to our hospital following presenting complaints of intermittent heavy and prolonged vaginal bleeding and a pelvic mass of six months duration. She also had symptoms suggestive of a partial mechanical intestinal obstruction and cannot have sexual intercourse due to pain and per vaginal bleeding. She had received about 20 pints of blood in two different hospitals prior to presentation in our hospital. Her past medical history was not contributory. She had two doses of goserelin injection (3.6 mg), and laparotomy prior to presentation following a provisional diagnosis of uterine fibroids and cervical fibroid polyp.

Clinical examination revealed a young lady who was in mild intermittent painful distress; she was not pale and not dehydrated. The abdomen was mildly distended. A 14-week-sized abdomino-pelvic mass was palpated. Bowel sounds were present but hypoactive, vaginal examination revealed a lobulated hard, broad-based mass with nodular surface, it measured about 12 cm × 12 cm × 12 cm. The mass involved the anterior, right lateral and posterior fornix of the vagina. It

How to cite this article: Nongo BH, Isah DA, Ahmed AB, Abdul OA. Vaginal endometrial stroma sarcoma: A case report of a rare disease. J West Afr Coll Surg 2024;14:233-7.

Received: 28-Jun-2023
Accepted: 06-Nov-2023
Published: 22-Feb-2024

Address for correspondence:
Dr. Dennis Anthony Isah, Department of Obstetrics and Gynaecology, University of Abuja Teaching Hospital, Abuja 902101, Nigeria.
E-mail: dennisanthonyisah@yahoo.com

Access this article online

Website: www.jwacs-jcoac.com

DOI: 10.4103/jwas.jwas_120_23

Quick Response Code:

displaced the external cervical OS to the left lateral fornix. It infiltrated deep into the pelvis. It did not bleed easily with contact. A digital rectal examination revealed that there were no thickened or irregular rectal mucosa and the gloved finger was not stained with blood.

The following investigations were done with their outcomes also noted as stated here. Full blood count, normal parameters, serum, electrolyte, urea and creatinine, and chloride were all mildly elevated, the other parameters were normal. Abdominal X-ray, erect and supine revealed features of partial intestinal obstruction from adhesive bands. Intravenous Urography revealed an abdominal-pelvic mass with left hydronephrosis (grade IV) with a non-functional right kidney (grade V) hydronephrosis. A computed tomography scan (CT scan) of the abdomen and pelvis revealed uterine fibroid with bilateral hydronephrosis and hydroureters (worst on the right). An abdominopelvic ultrasound scan (USS) revealed a bulky uterus with multiple fibroid seedlings, severe bilateral grade 4 hydronephrosis, and severe hydroureters, which was worst on the right side. Adult renal scintigraphy revealed evidence of global renal dysfunction with essentially a non-functional right kidney, the hydronephrotic left kidney showed relatively good function but very poor clearance. A chest X-ray, posterior–anterior view, showed normal findings.

The following provisional diagnoses were made-vaginal mass? Cause to rule out cervical fibroid polyp, multiple uterine fibroids, pressure effects of the vaginal and uterine masses on both the ureters and kidneys, and partial mechanical intestinal obstruction. She was counselled on the need to have a cystoscopy, biopsy of the vaginal mass and laparotomy. Two pints of whole blood were grouped and cross-matched and made available for the surgery.

Findings at cystoscopy revealed multiple polypoid masses in the bladder wall, that made it impossible to visualise the ureteric orifices. Thus the stenting of both ureters could not be done. The vaginal mass earlier described was seen and an excisional biopsy was taken at four different sites. At laparotomy, a well-healed Pfannenstiel incisional scar was seen. There were moderate pelvic and intraperitoneal adhesions. The bowel loops were mildly dilated because of a constrictive adhesion band that involved about 12 cm of the ileum, with the involved segments looking pale. The uterus was about 14 weeks in size and was firm to hard in consistency with diffuse infiltrative lesions. There were no uterine fibroids nor obvious uterine masses seen. Both fallopian tubes were buried in adhesions but both ovaries looked healthy. An excisional biopsy of the suspicious parts of the myometrium and the endometrium was taken but histology revealed no features of endometriosis, adenomyosis nor any other pathology in the uterus. Intestinal resection and anastomosis were done, post-operatively she did well. The result of immunohistochemistry on a sample from the vaginal mass confirmed ESS.

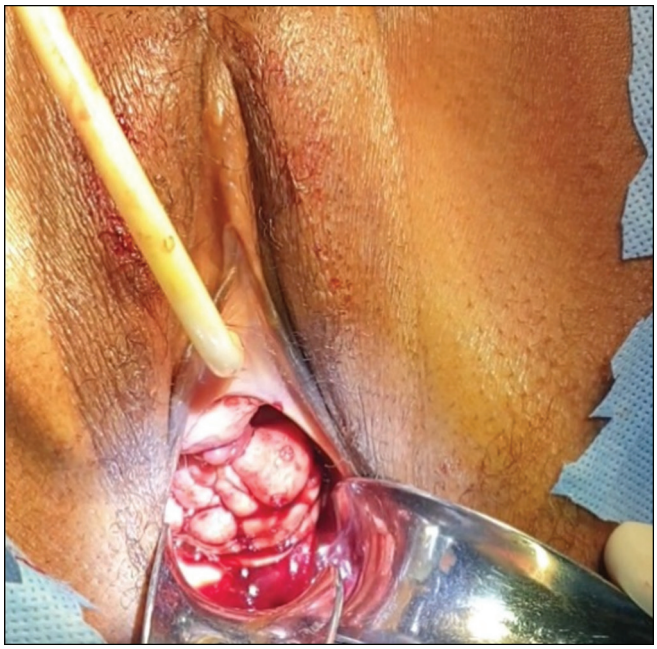

Figure 1: Vaginal mass as seen on speculum examination

She was subsequently counselled and was discharged and referred to another hospital with facilities to offer radiotherapy, chemotherapy, and hormonal therapy.

The histological sections showed a tumour consisting of small primitive hyperchromatic cells which are scattered with some fibres showing a cartwheel pattern. The tumour cells are hyperchromatic with round to oval nuclei, eosinophilic granular cytoplasms and resembling tennis rackets. These cells are disposed of within a myxoid stroma with oedema and inflammatory cells in other areas beneath the overlying epithelium. Mitotic figures are few <3/10 hpf. There are focal areas containing atypical cells. Histological sections from the vaginal tissue show similar malignant mesenchymal tumours composed of spindle-shaped cells. They are arranged in basket weave appearances. The cells have fusiform nuclei and elongated cytoplasm.

The result from the immunohistochemistry was as follows: SMA—Strong diffuse reactivity. Myogenin—Mild diffuse reactivity. Desmin—Nonreactive. EMA—Faint, non-specific reactivity. S100—nonreactive. Synaptophysin—nonreactive. Ki 67—Reactivity in <10% of the tumour and is perivascular. CD34—Strong positive vascular background, negative for the spindle cell component. Thus, the diagnosis from the immunohistochemistry was that of an extrauterine ESS [Figures 1–3].

Discussion

ESS is said to be a rare and malignant mesenchymal tumour of the uterus, it accounts for about 0.50% of all uterine malignancies and 10% of all uterine sarcomas.[3] The most recent (2014) World Health Organization Classification of endometrial stromal tumour is in four

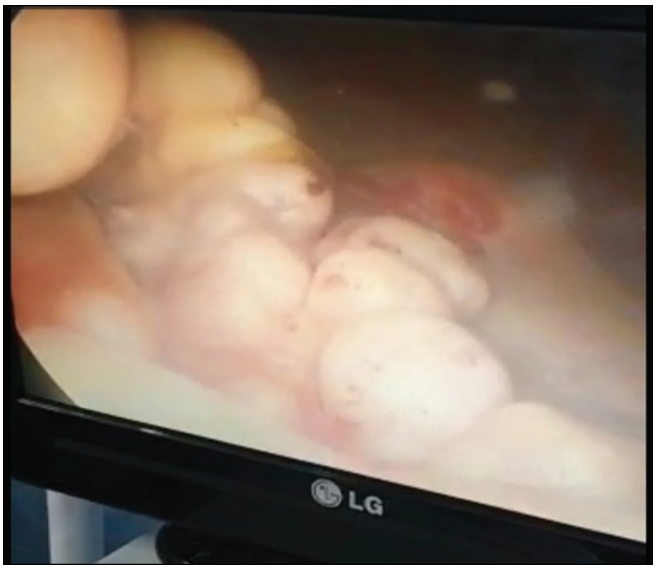

Figure 2: Polypoid metastatic masses into the bladder from VESS as seen during Cystoscopy

categories: Endometrial Stromal Node (ESN), low-grade ESS (LG-ESS), high-grade ESS (HG-ESS) and undifferentiated uterine sarcoma.[4] Our patient falls under HG-ESS. VESS are classified using the same classification of endometrial stromal tumour above. The presence of distinct translocations as well as tumour morphology and prognosis defines these categories.[4] Specifically, the JAZFI-SUZ 12 fusion identifies a large proportion of ESN and LG-ESSs, whereas the YWHAE—FAM 22 translocation identifies HG-ESSs. The latter tumour appears to have a prognosis intermediate between LG-ESS and UUS, which exhibits no specific translocation pattern.[4] The histological morphology of VESS is very similar to some other tumours. There is therefore always a need for comprehensive analysis of multiple immunohistochemical markers, such as desmin, SMA, CD34, CD117, Ki67, VIM, EMA, BCL2, CK, S100, Caldesmon, Calponin, CD10, ER, PR, STAT 6, CD99, and Cyclin D is needed to different it from other disease. In LE-ESS ER, PR, CD10, and vimentin are usually positive.[2,5] In our patient the following immunohistochemical markers were used and their outcomes are as follows: SMA—Strong diffuse reactivity, myogenin—Mild diffuse reactivity, Desmin—Nonreactive, EMA—Faint, non-specific reactivity, S100—nonreactive, synaptophysin—nonreactive, Ki67—reactivity <10% of the tumour and is perivascular CD34—Strong positive vascular background negative for the spindle cell component. Extrauterine Stromal Sarcoma can occur at the following sites, pelvic cavity, ovary, abdominal cavity fallopian tube, retroperitoneum, vulva, and vagina.[2] Of these extrauterine sites, the vagina is an extremely rare site. To the best of our knowledge, only 13 cases of VESS have so far been reported.[1] Our case is therefore the fourteenth.

Where the extrauterine ESS tumour cells originate from is yet unknown. The finding of endometriotic foci in the vicinity of the neoplasm in the majority of extrauterine ESS cases and the presence of endometriosis could explain the occurrence of this tumour in extrauterine sites such as fallopian tubes, ovaries, and pelvic peritoneum, according to the hypothesis of secondary mullerian system.[6] The above hypothesis suggests that the mesenchymal cells present in tissues derived from the coelomic epithelium have the potential to differentiate into mullerian-type epithelium and stromal such as endometriosis tissue. An alternative possible origin for the tumour in the absence of endometriosis is the primitive cells of the pelvis and retroperitoneum.[7] Above two possible theories cannot be applied to extrauterine Stromal Sarcoma occurring in sites such as vulva and vagina without foci of endometriosis as was found in our case. About 4 out of 5 cases of VESS are not associated with endometriosis. Herein lies the need for further interrogation as to the pathogenesis of VESS.[2]

VESS has no specific symptoms, however, the symptoms include vaginal mass, vaginal bleeding, vaginal discharge, pelvic mass, increasing abdominal girth, abdominal discomfort, and pelvic pain. Our patient presented with symptoms of vaginal bleeding, vaginal mass, pelvic mass, pelvic pain, and the symptoms of partial intestinal obstruction. The diagnosis of VESS depends on pathological examination after ruling out metastatic ESS.[1] The samples that were taken from our patient's endometrium and myometrium did not reveal ESS. Immunohistochemistry is most often what is used to differentiate it from other extrauterine tumours.[2] Vaginal endometrial Sarcoma must be diagnosed after excluding metastatic ESS.[8] This was ruled out in our case. VESS is mostly LG-ESS.[1] None of the immunohistochemical markers is specific for the diagnosis of ESS, CD10 staining is consistently positive in most ESS cases, as demonstrated in a recent study, which included 17 ESS cases, 94% of them had a positive staining.[5] In our patient it was CD34 that had a strong positivity with SMA also having a strong diffuse reactivity, myogenin had a mild diffuse reactivity while Ki 67 was reactive in <10% of the tumour. EMA had a faint non-specific reactivity. Desmin, S100 and synaptophysin were all nonreactive.

VESS is a rare tumour, as such there is as of now no consensus on its treatment. The same guidelines for treating ESS are followed. For patients with early-stage disease or desire for future fertility, excision of local lesions is usually performed. There has been a report of one patient who achieved a successful pregnancy after hysteroscopic resection of the tumour.[9,10] TAH and Bilateral Sulpingoophorectomy are usually performed for patients with advanced clinical-stage disease. Some experts favour ovarian sparing surgery for young patients with early disease, bilateral salpingophorectomy did not appear

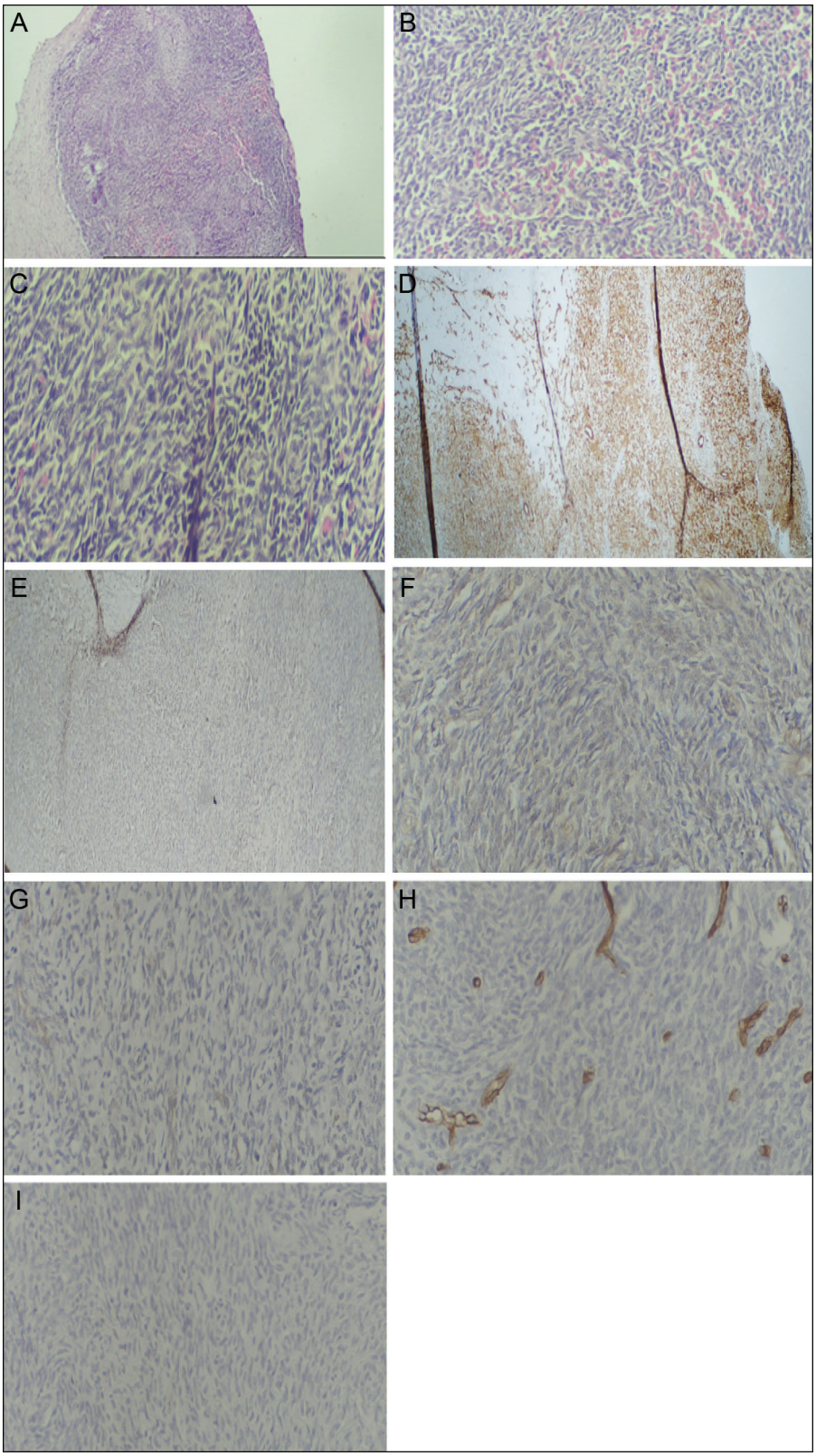

Figure 3: (A) Mesenchymal tumour nodule. H & E ×4 (B) Diffuse array of spindle cells. H &E ×10 tumour (C) Mesenchymal tumour with relatively low-grade nuclear features. H& E ×20 (D) Strong reactivity for SM at ×10 (E) Faint reactivity for EMA at ×10 (F) mild reactivity for MYO at ×20 (G) Faint reactivity for Ki 67 (H) No reactivity for CD34 at ×20 (I) No reactivity for S100 at ×20

to affect the time to recurrence or overall survival in stage I LG-ESS.[11] Our patient had stage IV advanced VESS, that had already metastasized to the bladder.

Therefore, the option of surgery was deemed not to be very feasible for her, and as such she was referred to another hospital that has the facilities to offer her chemoradiation and if needed hormonal therapy.

Adjuvant therapy involves radiotherapy, chemotherapy, and hormonal therapy. Opinions are divided but some claim that surgery and radiotherapy is the most effective treatment for ESS.[12] Our patient was referred to benefit from a possible combination of chemotherapy and radiotherapy, because of the advanced stage of her disease (stage IV).

Chemotherapeutic drugs that are used in the treatment of ESS include cyclophosphamide, cisplatin, Ifosfamide, epirubicin, and others.[13] Hormonal therapy was reported to have a favourable survival outcome in ER/PR-positive ESS.[14] Hormonal therapy is also used as an alternative mode of treatment for patients who desire fertility.[14] Megestrol acetate has been used with spontaneous pregnancy achieved.[15] Our patient being 27 years old Po^{+1} unmarried and with advanced disease may benefit from hormonal therapy. The prognosis of VESS is dependent on the stage of the disease and the expression of hormonal receptors ER and PR.[16,17] Advanced stage of the disease was associated with poor prognosis whereas the presence of hormonal receptors is significantly associated with improved overall survival.[16,17] Extrauterine ESS is a rare disease and LG extrauterine ESS (LG-EESS) has a good prognosis. Surgery remains the available treatment for patients. LG-EESS has a risk of late recurrence, which requires a long-term follow-up. With a limited sample size, our study shows optimal tumour reductive surgery and adjuvant hormone therapy may significantly reduce the risk of recurrence.[18]

Conclusion

VESS is a very rare disease. It has no documented classic symptoms for accurate diagnosis, immunohistochemistry is needed. There is as of now no known optimal treatment however, surgery is highly recommended. Other treatment options, such as chemotherapy, radiotherapy, and hormonal therapy may be cautiously used. The prognostic factors for VESS are the tumour stage and expression of the hormonal receptors. In view of the few numbers of the reported cases of VESS, further research into this disease is needed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Hongwen W, Yi L, Nanlin Y, Wenwu G, Yi C. Vaginal endometrial sarcoma: A literature review. Eur J Obstet Gynecol Reprod Biol X 2019;4:100094.
2. Liu Z, Ding J, Li X, Yu K. Endometrial stromal sarcoma arising in vaginal. Int J Clin Exp Pathol 2013;6:1997-3002.
3. Pautier P, Genestie C, Rey A, Morice P, Roche B, Lhomme C, *et al*. Analysis of clinicopathologic prognostic factors for 1st uterine sarcoma and evaluation of a grading score validated for soft tissue sarcoma. Cancer 2000;88:1425-31.
4. Conklin CM, Longacre TA. Endometrial tumours: The new WHO classification. Adv Anat Pathol 2014;21:383-93.
5. Rauh-Hain JA, del Carmen MG. Endometrial stromal sarcoma: A systematic review. Obstet Gynecol 2013;122:676-83.
6. Lauchlan SC. The secondary mullerian system revisited. Int J Gynecol Pathol 1994;13:73-9.
7. Chang YC, Wang TY, Tzen CY. Endometrial stromal sarcoma of the vagina. Zhonghua Yi Xue Za Zgi (Taipei) 2000;63:714-9.
8. Rivard C, Carter JS, Downs LS. Low-grade endometrial stromal sarcoma primarily arising in the vagina: A case report. J Reprod Med 2015;60:433-5.
9. Koskas M, Morice P, Yazbeck C, Duvillard P, Walker F, Madelenat P. Conservative management of low-grade endometrial stromal Sarcoma followed by pregnancy and several recurrence. Anticancer Res 2009;10:4147-50.
10. Chang B, Lu LX, Tu XY, Cheng YF, Bi R, Yang WT. Endometrial Stromal Sarcoma: Morphologic features and detection of JAZFI-SUZ12 and YINHAE—FAM 22 fusion genes. Zhonghua Bing Li Xue Za Zhi 2016;45:308-13.
11. Kim WY, Lee JW, Choi CH, Kang H, Kim TJ, Bim BG. Low grade endometrial stromal sarcoma: A single centre's experience with 22 cases. Int J Gynecol Cancer 2008;18:1084-9.
12. Weitmann HD, Knocke TH, Kucera H, Potter R. Radiation therapy in the treatment of endometrial stromal sarcoma. Int J Radiat Oncol Biol Phys 2001;49:739-48.
13. Yamazaki H, Todo Y, Mitsube K, HareyAma H, Shimada C, Kato H, *et al*. Long term survival of patients with recurrent endometrial stromal sarcoma: A multicenter, observational study. J Gynecol Oncol 2015;26:214-21.
14. Park JY, Baek MH, Park Y, Kim YT, Nam JH. Investigation of hormonal receptor expression and its prognostic value in endometrial stromal sarcoma. Virchows Arch 2018;473:61-9.
15. Delaney AA, Gubbels AL, Remmenga S, Tomich P, Molpus K. Successful pregnancy after fertility—Sparing local resection and uterine reconstruction for low-grade endometrial stromal sarcoma. Obstet Gynecol 2012;120:486-9.
16. Ngan HY, Fisher C, Blake P, Shepherd JH. Vaginal sarcoma: The Royal Marsden experience. Int J Gynecol Cancer 1994;4:337-41.
17. Mizuno M, Yatahe Y, Nawa A, Nakanishi T. Long term medroxyprogestrone acetate therapy for low-grade endometrial stromal sarcoma. Int J Clin Oncol 2012;17:348-54.
18. Wu Y, Li N, Zhang R, Bai P. Primary low-grade extrauterine endometrial stromal sarcoma: Analysis of 10 cases with a review of the literature. World J Surg Oncol 2022;20:17.