Radiology Case Reports 18 (2023) 3529–3534

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journal homepage: www.elsevier.com/locate/radcr

Case Report

A case of ovarian endometrial stromal sarcoma: Radiological and histopathological findings ☆

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ARTICLE INFO

Article history:
Received 3 May 2023
Revised 9 July 2023
Accepted 14 July 2023

Keywords:
Endometrial stromal sarcoma
Ovary
MRI
CT
Histopathological

ABSTRACT

Endometrial stromal sarcoma (ESS) is an uncommon uterine mesenchymal neoplasm. The primary extra-uterine location of ESS is a very rare occurrence.

We present a case of a 39-year-old woman presented with severe abdominal pain, MRI showed bilateral ovarian tumors with heterogeneous intensity on T2-weighted imaging (T2WI) and T1-weighted imaging (T1WI), with restricted diffusion, including hyperintense areas on T1WI, not erased on T1-weighted fat-suppressed imaging, hypointense on T2WI, and not enhanced after contrast. This mass extended to the Douglas and invaded the uterine and the rectum serosa suggesting an underlying endometriosis. No abnormalities were suspected in the endometrium.

The exploration revealed a friable mass arising from ovaries associated with nodules in the small intestine and sigmoid. The patient underwent bilateral adnexectomy and the anatomopathological study revealed a low-grade endometrial stromal sarcoma.

This is one of the few reports covering the radiological features of low-grade extra-uterine ESS in the ovary which is probably secondary to degeneration of endometriosis with no evidence of primary uterine ESS.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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https://doi.org/10.1016/j.radcr.2023.07.027

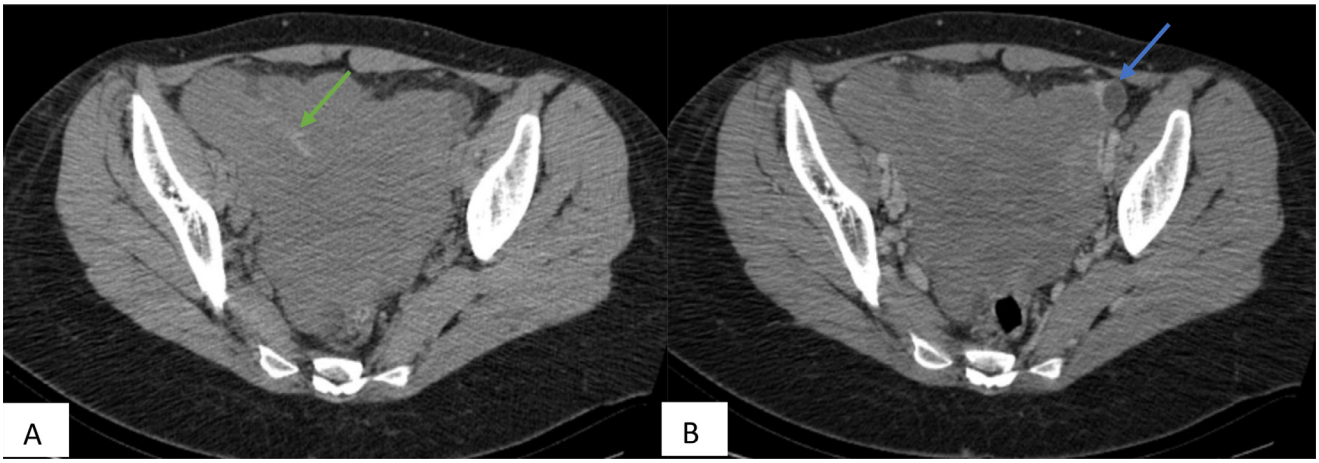

Fig. 1 – An axial computed tomography scan of the abdomen and pelvis before (A) and after contrast (B) showed bilateral ovarian tumors with heterogeneous contrast enhancement, including hemorrhagic (green arrow) and cystic areas (blue arrow).

Introduction

Endometrial stromal sarcomas (ESS) are rare histological entities, accounting for only 1%-2% of endometrial malignancies and ≤ 10% of all uterine sarcomas [1].

ESS mainly originates from the uterus but can also arise from extra-uterine locations. The major part of ectopic ESS cases originates from endometriosis through a malignant transformation process [2].

Extra-uterine ESS can arise from various sites such as the ovaries, fallopian tubes, vagina, broad ligament, intestine, peritoneum of the pelvic cavity, mesentery, bladder, and liver, with the ovary being the most common site [1].

In this case, we present a low-grade extra-uterine ESS in the ovary with radiological features suggesting underlying endometriosis, with no evidence of primary uterine ESS.

Case presentation

A 39-year-old woman with no prior medical history presented with severe abdominal pain. A computed tomography (CT) scan revealed a bilateral pelvic tumor without identification of both ovaries with ovarian veins joining the tumor, suggesting the ovarian origin of the mass. The tumor showed heterogeneous contrast enhancement, with hemorrhagic and cystic areas (Fig. 1).

On magnetic resonance imaging (MRI), the tumors exhibited heterogeneous intensity on T2WI and T1WI, with restricted diffusion. Some areas were hyperintense on T1WI, not erased on T1-weighted fat-suppressed imaging, hypointense on T2WI, and did not enhance after contrast. The mass extended to the Douglas and invaded the uterine and the rectum serosa. No abnormalities were suspected in the endometrium (Fig. 2).

Based on the combined results, the ovarian mass was considered a malignant scored ORADS 5 (due to the invasion of the uterine and the rectum serosa) arising from underlying endometriosis, and a laparotomy was planned. During surgery, direct visual inspection confirmed that the uterus was intact, and the mass arose from the ovaries. The exploration revealed a friable mass of soft consistency that bled on contact measuring 10 cm, associated with nodules in the small intestine and sigmoid with the same appearance not visualized in imaging (Fig. 3).

The patient underwent bilateral adnexectomy with residual mass in the Douglas and a biopsy of the nodules in the small intestine and sigmoid.

The histological examination of the adnexal lesion showed ovarian parenchyma largely occupied by a diffuse tumoral proliferation composed of round to spindle cells surrounded by small vessels with sometimes hyalinazed walls in a striking whorling pattern (Fig. 4A), mitotic activity was 10/10 High Power Fields (HPF) and no necrosis was seen. The histological examination of the other specimens (nodules in the small intestine and sigmoid) was identical to the adnexal tumor.

At immunohistochemistry, tumor cells were positive for CD10 (Fig. 4B), Vimentin, and estrogen and progesterone receptors (ER, PR). They were negative for smooth muscle actin (SMA), inhibin, calretinin, CD31, and CD34. Ki 67 proliferation index was 30%.

The recombination of the JAZF1 gene was identified by fluorescence in situ hybridization (FISH).

Upon the morphological, immune-histochemical, and molecular findings, it was diagnosed as Low-Grade Endometrial Stromal Sarcoma.

The MRI control showed a lesion of the cul-de-sac attached to the cervix with the invasion of the anterior wall of the rectum, with irregular contours, related to a residual tumor, associated with thickening of the torus on hyposignal T2WI, which suggests endometriosis (Fig. 5).

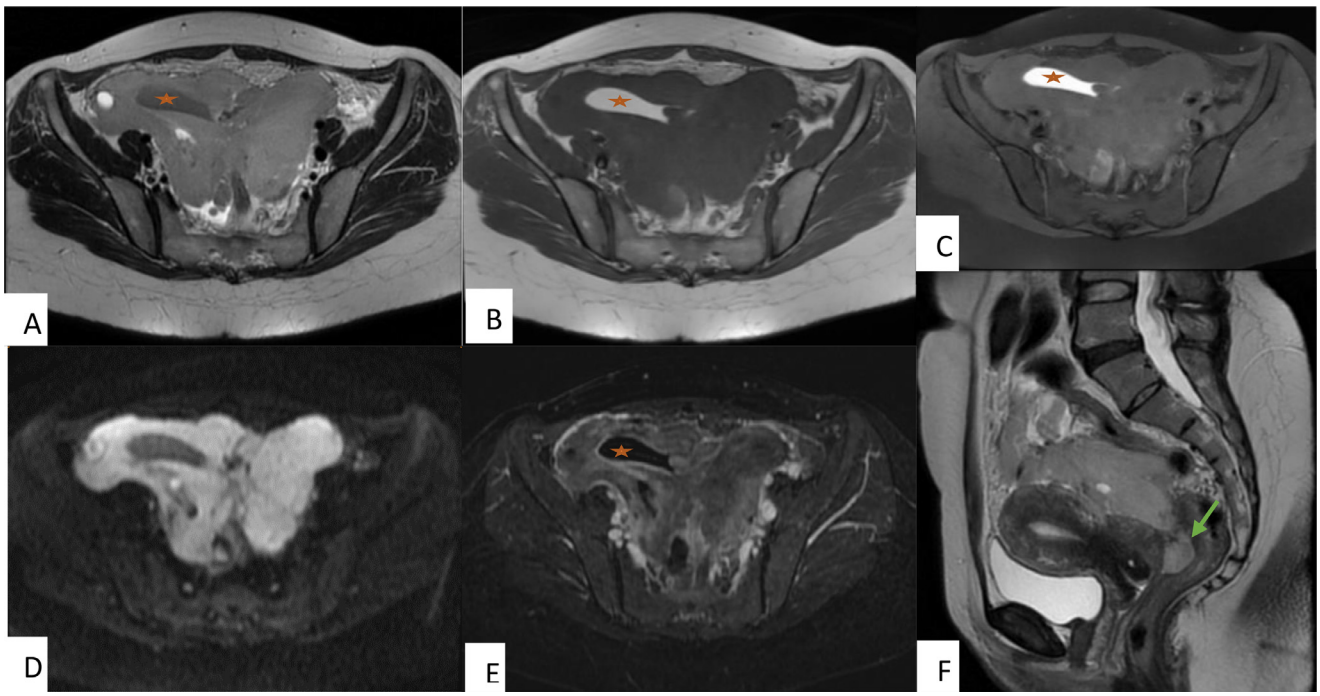

Fig. 2 – Pelvic magnetic resonance imaging (MRI), including axial T2WI (A), T1WI (B), T1-weighted fat-suppressed images (C), diffusion (D), dynamic contrast-enhanced images (E) and sagittal T2WI (F): the tumor presented heterogeneous intensity on T2WI and T1WI with restricted diffusion, including areas "that were hyperintense on T1WI, not erased on T1-weighted fat-suppressed images, hypointense on T2WI, and did not enhance after contrast (orange star)." This mass extended to the Douglas and invaded the uterine and the rectum serosa (green arrow).
No abnormality was suspected in the endometrium.

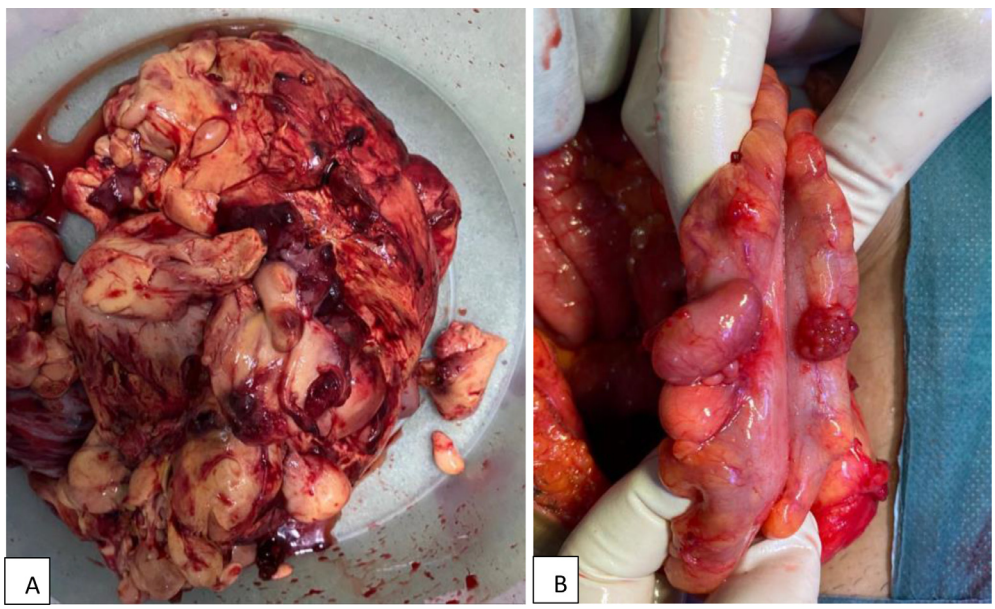

Fig. 3 – (A) A friable mass of soft consistency that bled on contact measuring 10 cm arising from the ovaries. (B) nodules in the small intestine with the same appearance.

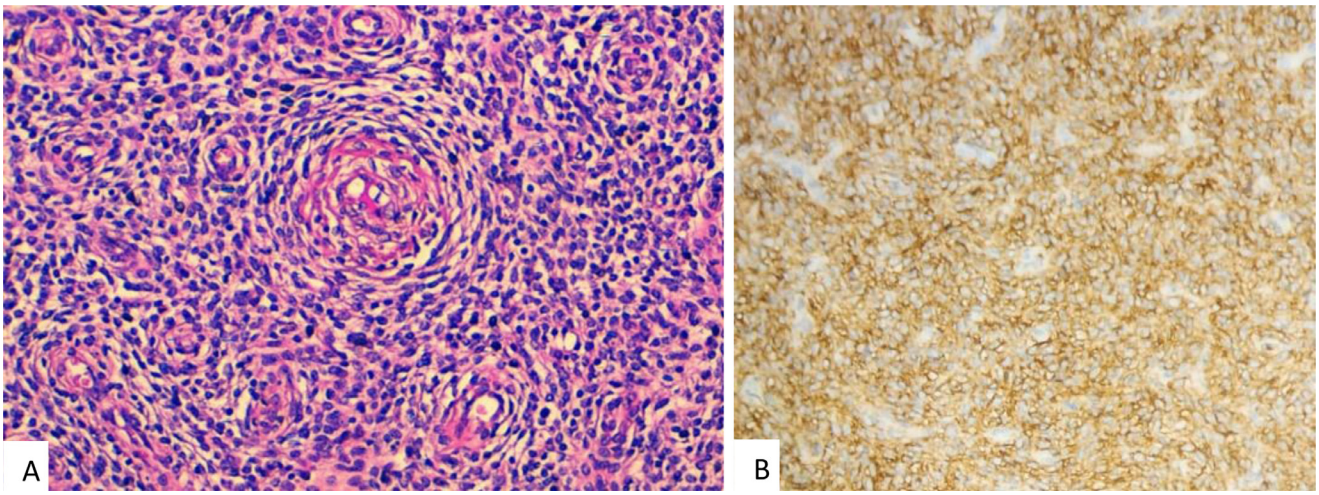

Fig. 4 – (A) SSE-diffuse tumor composed of round to spindle cells surrounded by small vessels with sometimes hyalinized walls in a striking whorling pattern. (B) CD10 immunostaining positive.

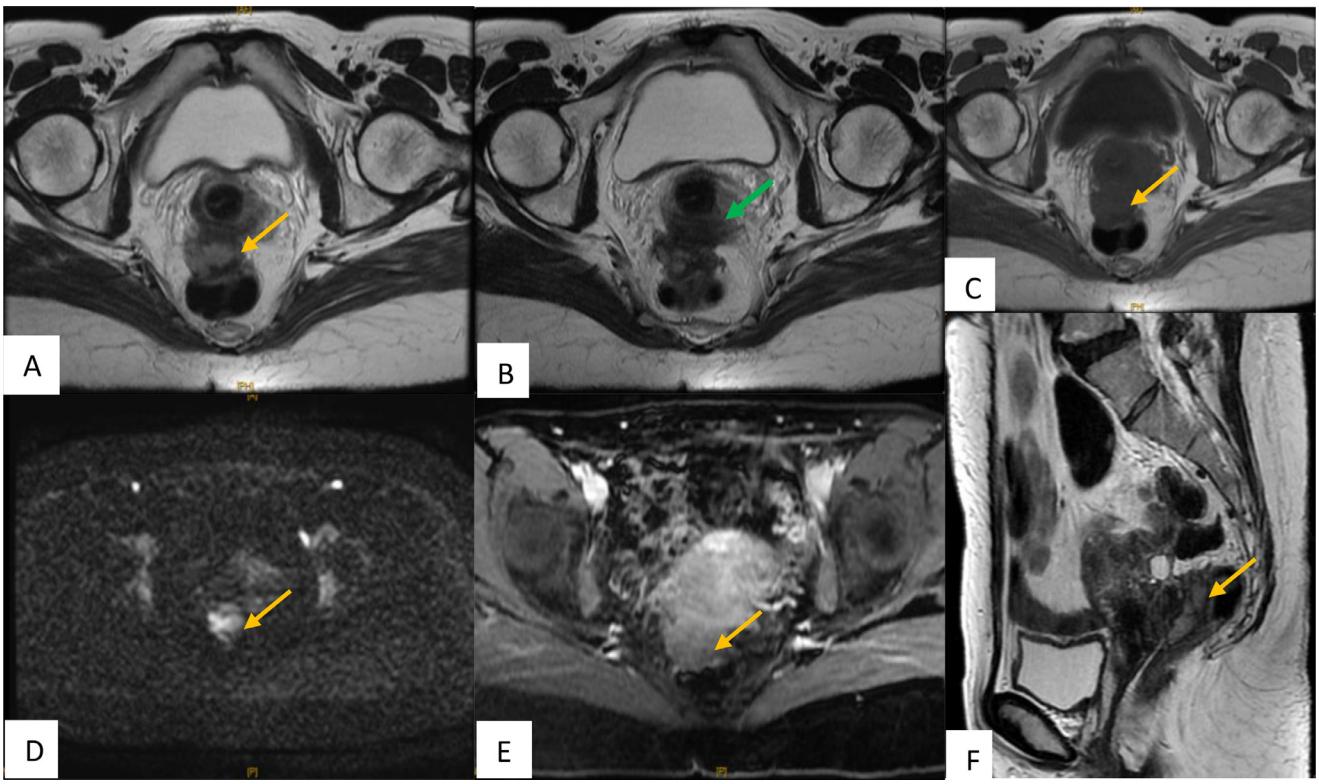

Fig. 5 – Pelvic MRI control on axial T2WI (A, B), T1WI (C), Diffusion (D), dynamic contrast-enhanced images (E) and sagittal T2WI (F) sequences revealed a lesion in the cul-de-sac (yellow arrow) that was attached to the cervix and invading the anterior wall of the rectum. The mass had irregular contours and showed intermediate signal intensity on T2WI and low signal intensity on T1WI, with restricted diffusion. The dynamic contrast-enhanced images indicated subtle and delayed enhancement compared to the external myometrium, which was attributed to a residual tumor. Additionally, the MRI showed thickening of the torus with hyposignal T2WI (B: green arrow), which suggests endometriosis.

Adjuvant hormonal therapy with an aromatase inhibitor drug was performed, and the patient is still on active surveillance.

Discussion

ESS is a rare malignant tumor originating from endometrial stromal cells, characterized by local invasion, vascular invasion, and easy recurrence. However, preoperative diagnosis is extremely difficult because the imaging properties of this rare tumor type have not been established [3].

Sometimes ESS originate primarily from extra-uterine sites such as the pelvis, ovary, abdominal cavity, retro-peritoneum, and vagina. In such cases, endometriosis lesions are often associated [2].

Primary ESS of the ovary is an extremely rare entity. The age of cases ranges from 34 to 76 years, with an average of about 50 years old [4].

Although patients with uterine ESS often complain of pelvic pain or metrorrhagia, ovarian ESS often presents with non-specific symptoms, such as abdominal tenderness and/or pain, and is sometimes asymptomatic. Most ovarian ESS is diagnosed at an advanced stage, with tumor extension outside the ovary [5].

In our case, the patient presented with severe abdominal pain, and the ovarian mass was diagnosed at an advanced stage.

In the past, uterine ESS was divided into low-grade and high-grade tumors based on their mitotic count. However, the new version of WHO classification in 2014 divides "endometrial stromal and related neoplasms" into 5 tumor types: endometrial stromal nodules, low-grade ESS, high-grade ESS, undifferentiated uterine sarcoma, and uterine tumor resembling ovarian germ cord tumor [6]. According to the 2014 WHO classification system, primary ovarian ESS can be divided into low-grade ESS and high-grade ESS [5].

Preoperative diagnosis of extra-uterine ESS is difficult, mainly in the absence of uterine ESS, as in this case, due to the rarity of the disease and the similarity of radiological findings with other tumors such as leiomyomas, leiomyosarcomas, and gastrointestinal stromal Tumor (GIST) [7].

Low-grade uterine ESS takes the form of a polypoid endometrial mass with well-defined borders or infiltrates. MRI usually shows T1 inhomogeneous isointense and T2 inhomogeneous hyperintensity, but T2 signal intensity may vary with degenerative changes [8,9]. Some MRI features of low-grade uterine ESS have been reported: enlarged vermiform nodules indicating the invasion in the vessels and lymphatics; and T2 hypointense bands indicating the myometrium bundles separated by the infiltrating tumoral cells; and extension along the ligaments. It is usually moderately enhanced postcontrast with restricted diffusion, with a mean ADC of 1.09 [10–12].

Most articles in our literature review only discuss low-grade extra-uterine ESS from clinical and pathological aspects, without emphasizing radiological findings. However, ill-defined borders, nodular structures, especially intermediate T2 signal intensity, restricted diffusion, and enhancement are in favor of a malignant etiology [13].

Our patient's MRI showed a bilateral ovarian solid mass with these radiological features. In addition, the hemorrhagic areas hyperintense on T1WI, the kissing ovaries sign, the thickening of the torus, the invasion of the uterine, and the rectum serosa suggest underlying endometriosis.

Definitive diagnosis of ESS is based on pathology because imaging studies do not supply specific signs. In fact, when the tumor is located in the uterus, a histological diagnosis of low-grade ESS is easily made. Problems occur when tumors originate in extrauterine sites.

The histological aspects of ectopic ESS are identical to those of the uterine site. They are multinodular tumor proliferations, monomorphic in appearance, which infiltrates in the form of a glove finger or tongue pattern. The vascularization is abundant, made of small blood vessels, surrounded by sleeves of tumor cells giving an aspect of spiral arterioles. Lymphatic emboli are frequent [4]. In terms of immunohistochemistry, ESS is usually positive for CD10, estrogen receptor, progesterone receptor, vimentin, (Wilm's tumor 1) WT-1 and negative for inhibin, calretinin, SMA, desmin, CD31 and CD34, which is consistent with our case. While, areas of sex cord differentiation stained positively for calretinin and inhibin, and areas of smooth muscle differentiation stained positively for SMA and desmin [14]. Low-grade ESS usually involves the fusion of 2 zinc finger genes JAZF1/SUZ12 and/or JAZF1/PHF1 between chromosomes 7 and 17, which helps distinguish low-grade ESS from high-grade ESS [7].

Definitive treatment for uterine or extrauterine ESS (whether high-grade or low-grade) is hysterectomy and bilateral salpingo-oophorectomy. Whether intrauterine or extrauterine, patients with low-grade ESS benefit from adjuvant hormone therapy, whereas patients with high-grade ESS require systemic chemoradiotherapy [13]. The benefits of adjuvant hormonal therapy for extra-uterine ESS may be limited, especially in cases of advanced or metastatic disease [7].

In our case, because of the intestinal localizations, a bilateral adnexectomy was performed with a residual tumor invading the anterior wall of the rectum, associated to an adjuvant hormonal therapy.

Conclusion

Extra-uterine low-grade ESS is an extremely rare neoplasm. Knowledge of the possible extra-uterine location of this low-grade tumor can guide clinicians, radiologists, and pathologists in making the correct diagnosis. Therefore, we suggest that the search for signs of endometriosis in MRI such as hemorrhagic areas, kissing ovaries sign, thickening of the torus and distant locations can guides the diagnosis of extra-uterine ESS arising from endometriosis.

Patient consent

I, the author of the article: A case of ovarian endometrial stromal sarcoma: radiological and histopathological findings approve that the patient gives her consent for information to be published in Radiology Case Reports.

REFERENCES

[1] Lee JS, Lee D, Lee J, Han MH, Hong DG, Lee HJ. Primary ovarian high-grade endometrial stromal sarcoma: a case report. J Med Case Rep 2021;15(1):1–5.

[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(4):323–7.

[3] Chen C, Hu Y-Q, Zhang X-M. Magnetic resonance imaging features of endometrial stromal sarcoma: a case description. Quantitat Imag Med Surg 2017;7(1):159.

[4] Efared B, Sidibé IS, Erregad F, Hammas N, Chbani L, El Fatemi H. Extrauterine low grade endometrioid stromal sarcoma arising from ovarian endometriosis: a case report and review of the literature. Gynecol Oncol Res Pract 2019;6(1):2.

[5] Masand RP, Euscher ED, Deavers MT, Malpica A. Endometrioid stromal sarcoma: a clinicopathologic study of 63 cases. Am J Surg Pathol 2013;37(11):1635–47.

[6] Conklin CM, Longacre TA. Endometrial stromal tumors: the new WHO classification. Adv Anat Pathol 2014;21(6):383–93.

[7] Suzuki S, Kurokawa R, Tsuruga T, Mori–Uchino M, Nishida H, Kato T, et al. MRI, and FDG-PET imaging findings of low-grade extrauterine endometrial stromal sarcoma arising from the mesentery: A case report. Radiol Case Rep. 2021;6(9):2774–9.

[8] Huang YL, Ueng SH, Chen K, Huang YT, Lu HY, Ng KK, et al. Utility of diffusion-weighted and contrast-enhanced magnetic resonance imaging in diagnosing and differentiating between high- and low-grade uterine endometrial stromal sarcoma. Cancer Imag 2019;19:63.

[9] Chaitra PA, Gyanchandani M, Goolahally LN, Itagi RM, Kalenahalli KV. Endometrial stromal sarcoma: an aggressive uterine malignancy. J Radiol Case Rep 2016;10(9):35–43. doi:10.3941/jrcr.v10i9.2770.

[10] Li HM, Liu J, Qiang JW, Gu WY, Zhang GF, Ma FH. Endometrial stromal sarcoma of the uterus magnetic resonance imaging findings including apparent diffusion coefficient value and its correlation with Ki-67 expression. Intl J Gynecol Cancer 2017;27(9):1877–87.

[11] Santos P, Cunha TM. Uterine sarcomas: clinical presentation and MRI features. Diagn Interv Radiol 2015;21:4–9.

[12] Huang YT, Huang YL, Ng KK, Lin G. Current status of magnetic resonance imaging in patients with malignant uterine neoplasms: a review. Korean J Radiol 2019;20(1):18–33.

[13] Lipsich F, Causa Andrieu PI, Wernicke A, Patrono MG, Napoli MN, Chacon CRB, et al. Extra-uterine endometrial stromal sarcoma arising from deep infiltrating endometriosis. Clin Imaging 2020;67:250–4. doi:10.1016/j.clinimag.2020.08.015.

[14] D'Angelo E, Prat J. Diagnostic use of immunohistochemistry in uterine mesenchymal tumors. Semin Diagn Pathol 2014;31:216–22.