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Low-grade primary endometrial stromal sarcoma of the ovary: Histological, immunohistochemical, and molecular genetic features

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Abstract
Primary endometrial stromal sarcomas of the ovary are rare mesenchymal tumors. They account for 0.2% of tumours of the female reproductive system and 10% of all uterine sarcomas. The tumor is usually located in the uterus, but can also be found elsewhere, posing major diagnostic problems. The majority of ectopic ESS occurs in areas with pre-existing endometriosis. The most common site is the ovary. The behavior of primary ovarian tumors is similar to that of their uterine counterparts. Low-grade ESS is an indolent tumor with a propensity for late recurrence. We report this case here, together with the literature.

Keywords: Endometrial stromal sarcoma, Ovary, Immunohistochemistry, FISH, Endometriosis

Introduction
Primary endometrial stromal sarcomas (ESS) of the ovary are rare mesenchymal tumors with limited data on their behavior and optimal treatment, making their clinical presentation misleading and diagnosis challenging. An association between extra-uterine endometrial stromal sarcomas and endometriosis has often been observed, though inconsistently. Awareness of the potential extra-uterine location of this tumor should guide clinicians and pathologists towards the correct diagnosis, as ESS exhibits histopathological features and clinical behavior similar to those of the uterine tumor.

Case Presentation
This is a 39-year-old woman who presented with pelvic pain and abdominal enlargement associated with a sensation of discomfort lasting for one month, without any history of metrorrhagia or leucorrhea, prompting her consultation. The patient also reported digestive symptoms, leading to digestive explorations with staged colonic biopsies.
Investigations revealed a solid, whitish tumor-like mass originating from the left ovary, infiltrating the right ovary, and adherent to the sigmoid colon. Intraoperatively, a tumor arising from the left ovary and adherent to the sigmoid colon was observed. There was no ascites or lymphadenopathy. A bilateral adnexectomy was performed."
The histopathological examination of the surgical specimen (Fig 1A), and colonic biopsy (Fig1B), revealed the presence of a highly vascularized tumor proliferation, arranged in diffuse sheets composed of cells of variable size, with oval nuclei containing fine chromatin and a prominent nucleolus. The cytoplasm was abundantly eosinophilic. Fibrous septa intersected the tumor nodules. The mitotic count was 10 mitoses per 10 high-power fields. The stroma was fibrous. Tumor cells were found on the surface of the ovarian capsule and had infiltrated blood and lymphatic vessels. Adjacent to the tumor tissue, glands resembling endometrium were found, which could be compatible with ovarian endometriosis.

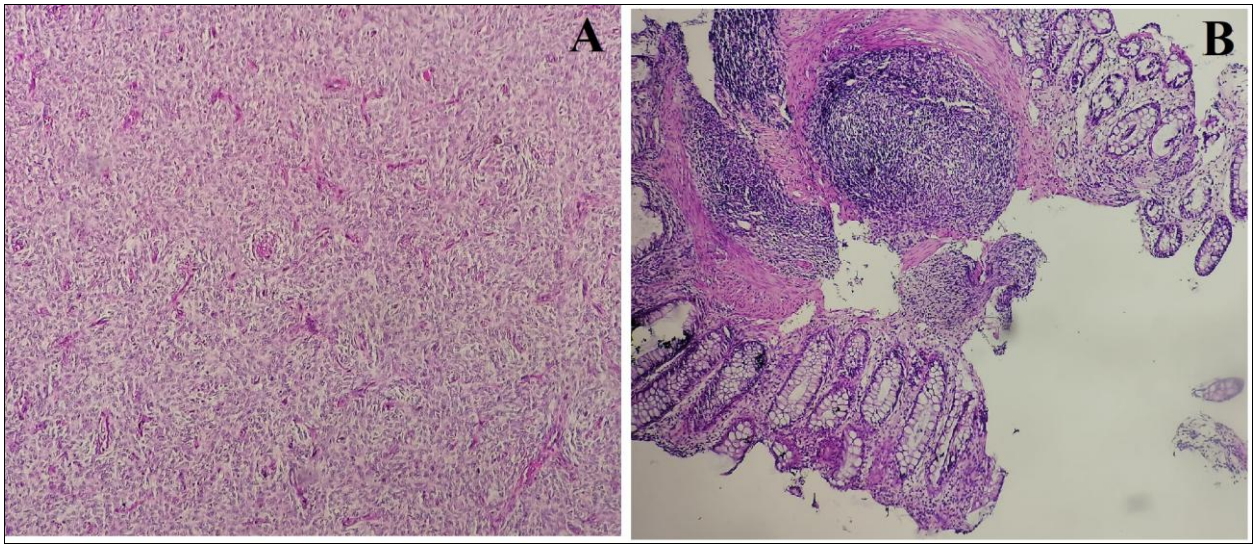
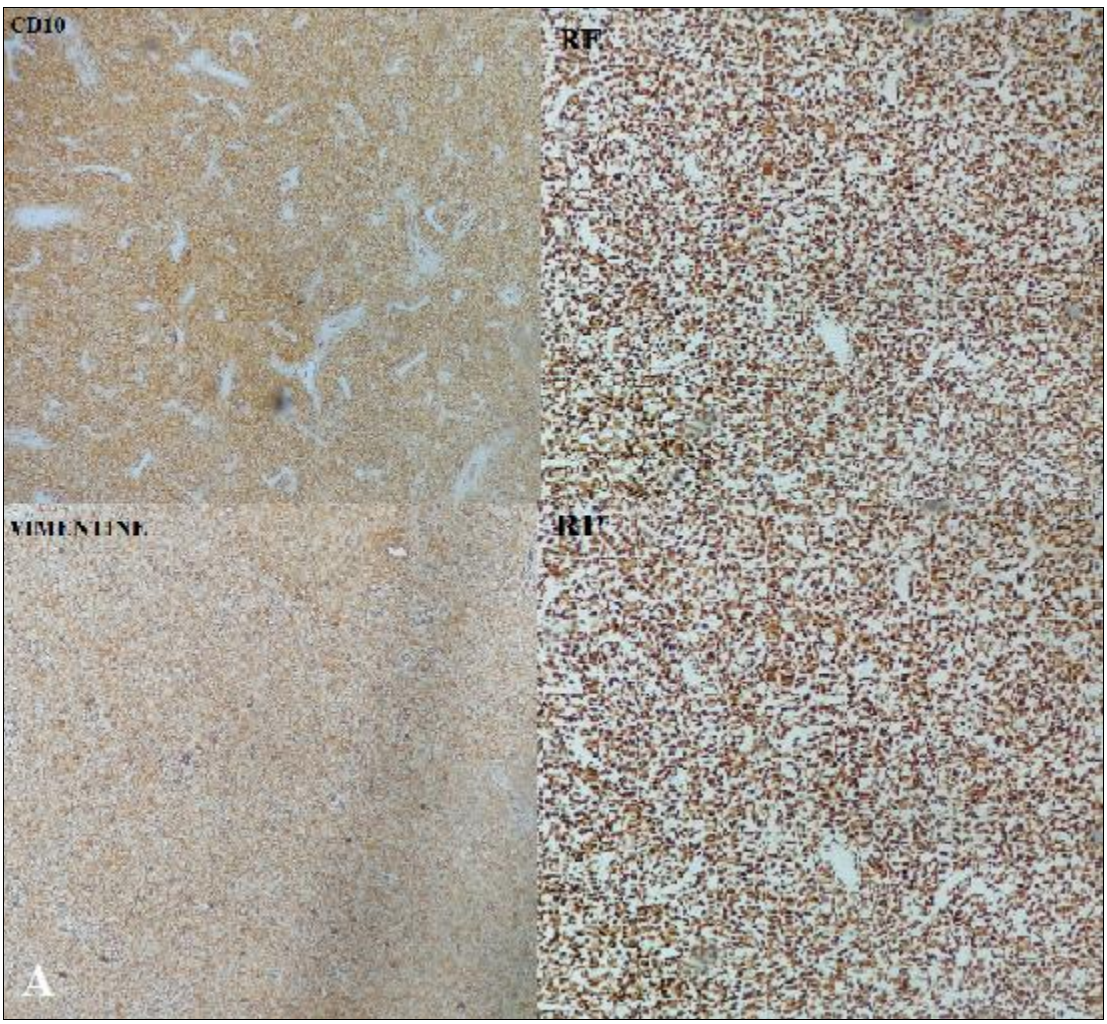


Fig 1: Histologic features of endometrial stromal sarcoma (A) and colonic localization (B) (HES× 200).

On immunohistochemistry (Fig 2), the tumor cells showed intense and diffuse expression of vimentin, CD10, and hormone receptors (RE, RP) (Fig2A). A few isolated and scattered cells expressed CK8/18, WT1, and CD99. There was no expression of EMA, CD34, Melan-A, HMB45, inhibin, calretinin, chromogranin, synaptophysin, S100, STAT6, CD117, AML, desmin, h-caldesmon, PLAP, or AFP (Fig2B). The Ki-67 index was estimated at 30%. An endometrial stromal sarcoma was first considered.

A molecular biology (Fig 3) analysis was performed, revealing a rearrangement of the JAZF1 gene, JAZF1 gene rearrangements are distinct genetic alterations found in endometrial stromal sarcomas (ESS) that help in diagnosis, JAZF1 fusions are associated with low-grade (LG) ESS. The diagnosis of a low-grade ovarian endometrial stromal sarcoma was confirmed.



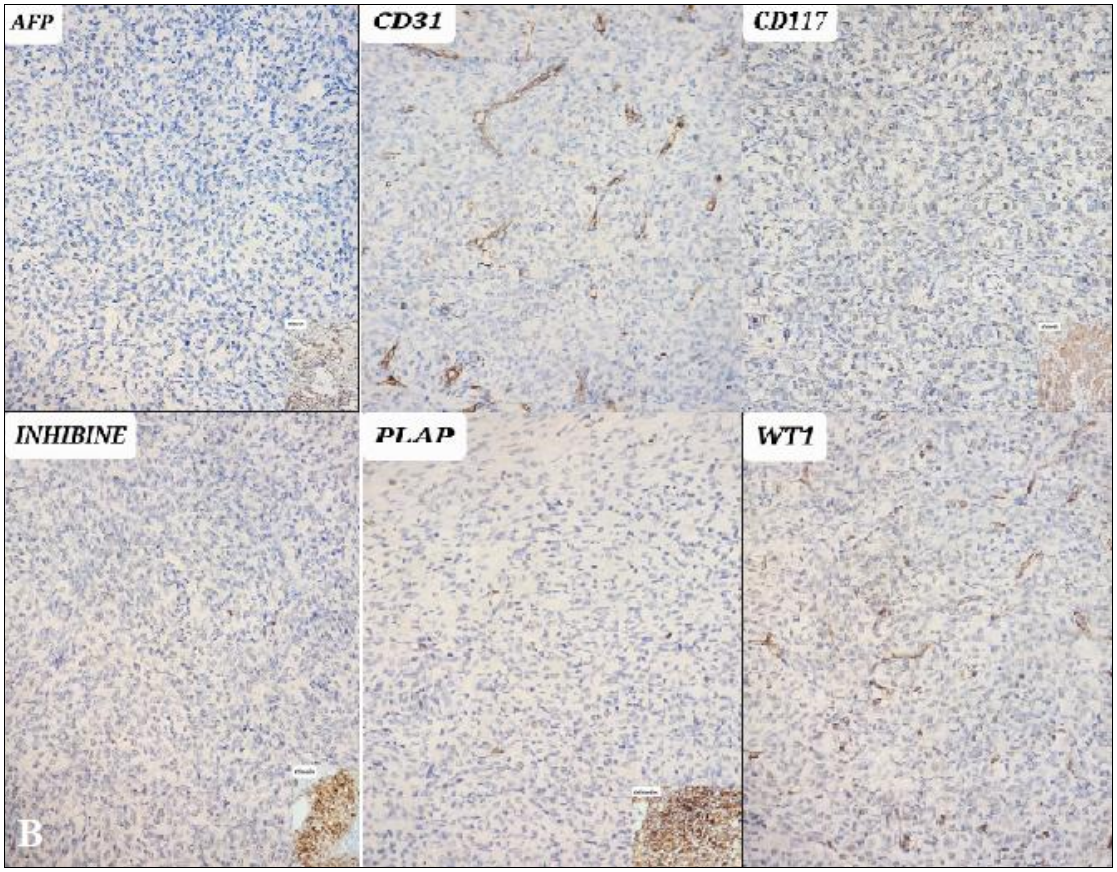


Fig 2: Immunohistochemical characteristics of the tumor (× 200): On immunohistochemistry, the tumor cells showed intense and diffuse expression of vimentin, CD10, and hormone receptors (fig2A). A few isola ted and scattered cells expressed CK8/18, WT1, and CD99. There was no expression of EMA, CD34, Melan-A, HMB45, inhibin, calretinin, chromogranin, synaptophysin, S100, STAT6, CD117, AML, desmin, h-caldesmon, PLAP, or AFP (fig2B).

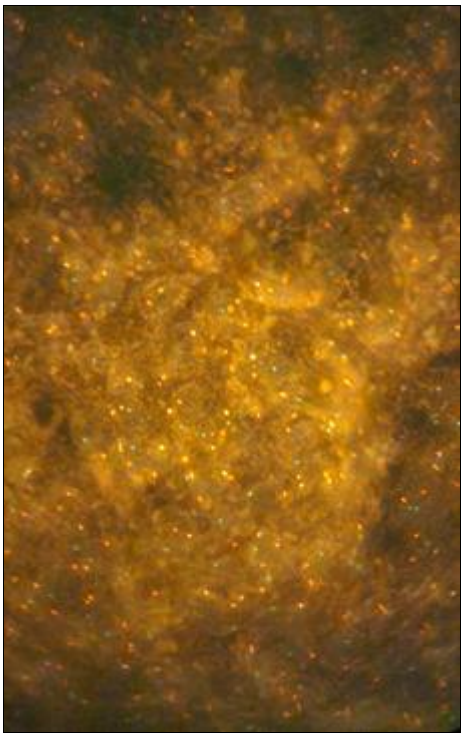


Fig 3: Molecular characteristics of the tumor: A rearrangement of the JAZF1 gene.

Discussion

Extra-uterine endometrial stromal sarcoma (EESS) is an exceptionally rare condition, with the current literature providing only a few case reports and brief series [1]. Ovaries are the most common site of EESS [2] and its occurrence in the colonic wall is exceedingly rare.

The clinical presentation of EESS is non-specific and often depends on the tumor's location and size. Reported cases of ovarian ESS have shown a wide range of clinical symptoms, varying from asymptomatic to significant abdominal

distension [2].

The 2014 WHO classification system categorizes primary ovarian ESS into low-grade ESS and high-grade ESS [3].

Diagnosing extra-uterine ESS is challenging, particularly when uterine ESS is not present, as in this case. The rarity of the condition, combined with its similarities to other tumors, makes the diagnosis even more difficult [4].

Associated foci of endometriosis have been documented in several case series. In our case, despite extensive sampling, no histological evidence of endometriosis was found in the resected colon or the ovaries. Given that endometriosis foci are typically found near EESS, it was initially suggested that the primary EESS might have originated from ectopic endometrial stroma associated with endometriosis. However, our findings indicate that the absence of endometriosis does not rule out the possibility of a primary EESS at that site. The pathogenesis of primary EESS without any detectable endometriosis remains unclear [5].

The tumor cells are typically positive for CD10, estrogen receptors (ER), and progesterone receptors (PR). Additionally, the characteristic JAZF1 gene rearrangements found in uterine LGESSs are also present in ovarian LGESSs.

Approximately two-thirds of low-grade endometrial stromal sarcomas exhibit genetic fusions involving polycomb family genes, with JAZF1-SUZ12 being the most frequent.

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Conclusion

Extra-uterine low-grade endometrioid stromal sarcoma (EESS) is an exceedingly rare tumor that can present with misleading clinical signs, making diagnosis challenging. Awareness of the possibility of an extra-uterine origin for this low-grade tumor is crucial for clinicians and pathologists. Recognizing that EESS shares similar histopathological features and clinical behavior with its uterine counterpart can aid in achieving an accurate diagnosis [6].

Conflicts of interest.

No potential conflicts of interest.

References

1. Oliva E, Egger JF, 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(3):305-15.
2. Efared B, Sidibé IS, Erregad F, Hammas N, Chbani L, El Fatemi H. Extra-uterine low grade endometrioid stromal sarcoma arising from ovarian endometriosis: a case report and review of the literature. Gynecol Oncol Res Pract. 2019;6:1-7.
3. 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.
4. Ayuso A, Fadare O, Khabele D. A case of extrauterine endometrial stromal sarcoma in the colon diagnosed three decades after hysterectomy for benign disease. Case Rep Obstet Gynecol. 2013;2013:202458.
5. Chen CW, Ou JJ, Wu CC, Hsiao CW, Cheng MF, Jao SW. High-grade endometrial stromal sarcoma arising from colon endometriosis. Int J Colorectal Dis. 2007;22:1551-3.
6. Höhn AK, Brambs CE, Hiller GGR, May D, Schmoeckel E, Horn LC. 2020 WHO classification of female genital tumors. Geburtsh Frauenheilkd. 2021;81(10):1145-53.

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