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Case Report
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Vaginal Endometrial Stromal Sarcoma - A Case Report and Literature Review

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Abstract

Vaginal Endometrial Stromal Sarcoma (VESS), an extrauterine endometrial stromal sarcoma in the absence of a primary uterine lesion, is an extremely rare disease. Currently only fourteen cases have been documented. VESS may be correlated with endometriosis and chronic high-dose estrogen levels. It is usually diagnosed by a pathological examination. Specific clinical symptoms are often not present. In most cases, VESS is a low-grade endometrial stromal sarcoma. The malignant transformation of endometrial stroma is marked by uniform stromal cells with minimal cellular pleomorphism, mild nuclear atypia and variable mitoses. We report the case of a 33-year-old woman who presented with increasing pelvic pain and was diagnosed with a low-grade VESS. On examination, a smooth-bordered 2.0 cm × 2.1 cm lesion between the right levator muscle and the right posterior vaginal wall was found. There are no guidelines based on large prospective-randomized studies for the treatment of Endometrial Stromal Sarcomas and existing recommendations are mostly derived from case reports due to the rarity of these tumors. Our patient received a local excision of the tumor and an adjuvant treatment with a GnRH analogue combined with an aromatase inhibitor.

In conclusion, this case report clearly shows that LG-ESS must also be considered in women with rectovaginal fistula, even though it is very rare. Due to the rarity of VESS, the creation of a register for one is also useful.

Keywords: Vaginal endometrial stromal sarcoma; Malignant transformation of endometriosis; Extrauterine sarcoma

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Case Presentation

A 33-year-old female patient complained of increasing, severe pelvic pain for around one year. Initially, the dull pain occurred only during movement, but with time it also manifested at rest. The patient reported no other complaints. Her past medical history was unremarkable and there was no relevant family history and no history of other clinically relevant diseases.

During physical examination a hard, palpable lesion was found on the right pararectal side on her posterior vaginal wall. The pelvic Magnetic Resonance Imaging (MRI) showed a smooth-bordered 2.0 cm × 2.1 cm lesion between the right levator muscle and the right posterior vaginal wall (Figure 1). No other abnormalities were found in the lower abdomen. The patient first presented at a regional hospital and received a fractional, transvaginal resection of the tumor due to the primary suspicion of an endometriosis cyst. Histological work-up including a reference pathological evaluation diagnosed a Low-Grade Vaginal Endometrial Stromal Sarcoma (LG-VESS). Two months after the initial resection the patient noticed a discharge of bowel contents through her vagina and was admitted to our hospital.

Physical examination revealed a scarred vaginal density at 6 o'clock without any sign of dehiscence or stool leakage. No sonographic abnormalities were found in the uterus and the adnexal region, as well as no free fluid in the Douglas space.

Rectoscopy showed a supra-sphincteric rectovaginal fistula in the scarred area of the vagina at 6 o'clock. The histopathological results detected a regular cervix without any infiltrates of the clinically known low-grade stromal sarcoma. The radiologic workup, including a CT scan of the chest/abdomen, did not find evidence of metastatic disease. Since the patients' desire to have children

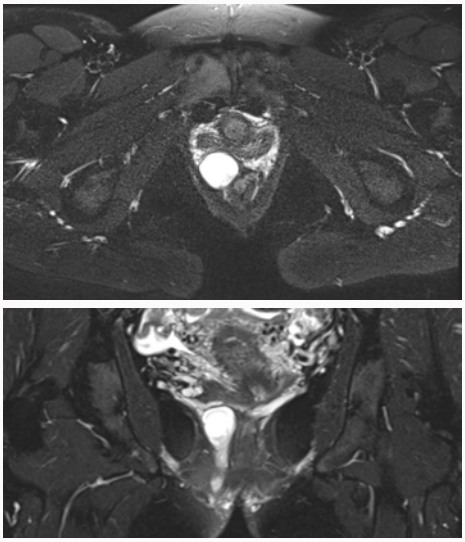

Figure 1: MRI-scan; 2.0 cm × 2.1 cm lesion between the right levator muscle and right the posterior wall of the vagina.

had not been fulfilled yet, a fertility-preserving surgery including an en-bloc resection of the scarred recto-vaginal part as part of shared decision-making therapy was planned.

In cooperation with the department of general- and visceral surgery, a whole resection of the vagino-rectal fistula with preservation of the sphincter muscle was performed. A Gracilis muscle flap from the right thigh was used to cover the anastomosis resulting from the resection. A protective ileostomy was placed laparoscopically to protect the anastomosis. The histopathological report described a low-grade endometrioid stromal sarcoma infiltrate which was locally resected in Sano. The microscopical analysis showed bland oval cells surrounding arterioles resembling endometrial spiral arterioles with no nuclear atypia or pleomorphism. The immunohistochemical evaluation was as follows: Positive for CD10 and ER, isolated cells were Desmin positive, negative for CK, PR and S100. The Ki-67 labelling index was 1% (Figure 2). Due to positive hormone receptor status adjuvant treatment with a GnRH analogue (11.25 mg Leuprorelin acetate q3m) and aromatase inhibitor (Letrozol® 2.5 mg q1d) was started after surgery for at least two years. Five months after the operation, the ileostomy was moved back without complications (Figure 3).

Discussion and Conclusion

Uterine mesenchymal neoplasms also include Endometrial Stromal Sarcomas (ESS), which account for less than 10 percent of uterine sarcomas and only about 1 percent of all malignant neoplasms of the uterus [1]. There have been numerous reports of primary ESS cases in extrauterine locations such as the pelvic cavity, ovary, abdominal cavity, fallopian tube, retroperitoneum, vulva, and vagina [2]. However, the vagina (VESS) is with fourteen documented cases an extremely rare site of these extrauterine locations [3].

The origin of extrauterine ESS tumor cells is not yet fully understood. In most cases of extrauterine ESS, foci of endometriosis have been detected in the vicinity of the neoplasm, and the presence of endometriosis may explain the estrogen-depending occurrence of these tumors in extrauterine sites such as the ovaries, fallopian tube, and pelvic peritoneum [4].

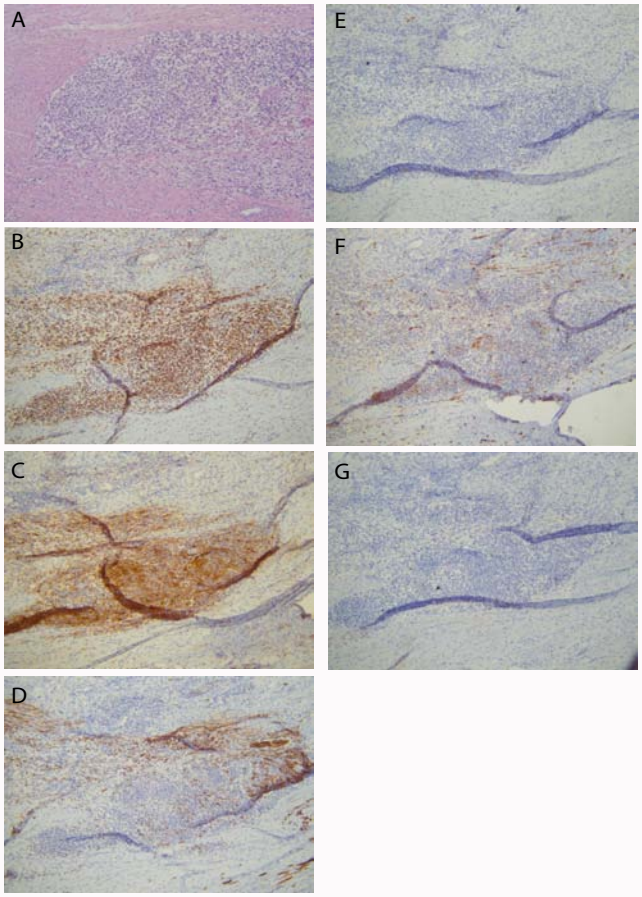

Figure 2: Vaginal endometrial stromal sarcoma (© A. Schad). Light microscopic examination showed proliferates of spindle cells with slightly enlarged nuclei (A). Immunohistochemically, the tumor cells were positive for ER (B) und CD10 (C), isolated cells were Desmin positive (D), negative for CK, PR (E) and S100 (F). The Ki-67 labelling index was 1 % (G).

Typically, VESS presents with nonspecific symptoms such as irregular vaginal bleeding, particularly after sexual intercourse, increasing abdominal discomfort, as well as abdominal distension. Therefore, a definitive diagnosis of ESS can only be achieved through pathological examination. Immunohistochemistry is the preferred method for differentiating ESS from other extrauterine tumors, although none of the immunohistochemical markers are specific for the diagnosis of ESS. In our case, the tumor cells were immunoreactive for ER and CD10, but negative for CK, PR and S100. In most cases, VESS is a low-grade endometrial stromal sarcoma with lower than 10 mitosis/10 HPF and sparse necrosis. Dense, uniform stromal cells with minimal cellular pleomorphism, mild nuclear atypia and variable mitoses are typical characteristics of ESS [5,6].

There are currently no established guidelines available based on large prospective-randomized studies for the treatment of ESS and recommendations are mostly derived from case reports due to the rarity of these tumors. Surgery remains the primary treatment option for ESS, especially for early-stage cases confined to the uterus (stage I-II) [7]. Complete resection may be a good choice for patients in early disease stages without any remote metastases. A total hysterectomy combined with Bilateral Salpingo-Oophorectomy (BSO) and careful abdominal exploration is commonly accepted as the treatment of choice in higher stages (stage ≥ 3) [8]. Depending on hormone receptor status of the tumor, ovarian-sparing procedures should only

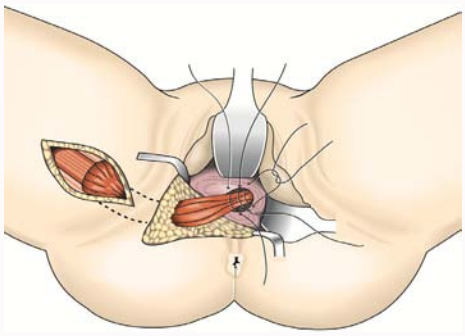

Figure 3: Gracilis plastic (© M. v. Solodkoff)

be considered in young women, as reported in our case. Although local recurrences are common after fertility-sparing treatment, they can be treated by surgery without compromising survival [9]. After surgery, treatment options may include local radiation therapy, adjuvant hormone therapy or a cytostatic chemotherapy [10]. Although radiotherapy may reduce local recurrence, its effect on long-term survival is uncertain. Hormonal therapy, particularly progestin therapy, is an option because low-grade ESS are typically positive for progesterone receptor. Evidence regarding cytostatic chemotherapy (six courses platinum-containing combination chemotherapy (PAC regimen) as adjuvant therapies after surgery) for advanced or recurrent ESS is scarce due to the rarity of the disease and only a few clinical trials have been reported [11].

In general, treatment of primary VESS should be individualized based on stage and grade of the tumor as well as the patient's age and overall health. Further prospective trials are needed to identify the most effective treatments for vaginal ESS and to establish evidence-based guidelines. Close follow-up with imaging and pelvic exams is also important to timely detect any recurrence or metastasis. The current recommendation for high-grade sarcomas is a physical examination and CT imaging of the chest, abdomen and pelvis every three to four months for the first two to three years, every six months for the next five years and annually thereafter. LGESS usually presents with a slow-growing, malignant course with a rather restrained development. Several studies suggest that up to 80% of recurrences can be detected by regular radiological surveillance [12].

Prognosis of uterine sarcomas is usually bad, with a median time to recurrence of 14 months. For stromal sarcomas the situation is different. The prognosis of LG-ESS generally tends to be favorable. The rate for disease-specific five-year survival is 80 to 90 percent and for ten-year survival about 70 percent [13,14].

In conclusion, this case report clearly shows that LG-ESS must also be considered in women with rectovaginal fistula, even though it is very rare. Due to the rarity of VESS, the creation of a register for one is also useful.

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