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Case report

Low-grade endometrial stromal sarcoma with extensive sex cord differentiation, heterologous elements, and complex atypical hyperplasia: Case report and review of literature

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1. Introduction

Uterine mesenchymal neoplasia is a broad category and includes smooth muscle tumors, endometrial stromal tumors (EST's), homologous sarcomas, heterologous sarcomas, sarcomas of uncertain histogenesis, and mixed epithelial and mesenchymal tumors, including uterine tumor resembling ovarian sex cord tumor (UTROSCT). EST's represent the second most common of these, but are still relatively rare, with malignant subtypes accounting for <10% of uterine sarcomas and <1% of primary uterine malignancies (Conklin and Longacre, 2014). The recently revised World Health Organization (2014) classification scheme of this entity defines four categories based on morphologic resemblance to proliferative-phase endometrial stroma, clinical behavior, and specific genetic translocations. Endometrial stromal nodule (ESN) and low-grade endometrial stromal sarcoma (LG-ESS) demonstrate sheets of cells with uniform oval nuclei, scant cytoplasm, minimal cytologic atypia, and variable mitotic activity. The neoplastic cells are only slightly larger than benign endometrial stromal cells. Permeative tongue-like growth into the myometrium and lymphovascular invasion set LG-ESS apart from ESN. High-grade ESS (HG-ESS) demonstrates cytologic atypia beyond that of LG-ESS but not so extreme as to abrogate its resemblance to benign proliferative endometrial stroma. HG-ESS is further characterized by the unique translocation t(10;17) not seen in other uterine mesenchymal tumors. Undifferentiated uterine sarcoma (UUS) demonstrates marked atypia without recognizable stromal differentiation. Destructive, rather than permeative, myometrial invasion is characteristic.

ESN and LG-ESS may focally demonstrate variant morphology including smooth muscle differentiation, fibromyxoid change, sex cord-like differentiation, and endometrioid-type glands. These understandably create diagnostic challenges, especially when the variant is extensive, as in the present case.

2. Case

A 56-year-old woman presented to her gynecologist with postmenopausal bleeding. Past medical history is insignificant. Family history included one sister with "uterine cancer" in her twenties without a history of obesity. The same sister received a diagnosis of breast cancer only several years later. The patient's BMI was 22.4 kg/m^2 upon presentation. A transvaginal ultrasound revealed a thickened endometrial stripe (23 mm). Endometrial dilation and curettage revealed a fragmented mesenchymal lesion with mixed spindled and epithelioid cells suggestive of UTROSCT. Additional areas demonstrated focal complex atypical hyperplasia of endometrial glands.

The patient underwent exploratory laparotomy with total abdominal hysterectomy, bilateral salpingo-oophorectomy, pelvic and paraaortic lymphadenectomy, omentectomy, and abdominopelvic washings. Operative findings included an enlarged uterus (15-weeks) with a retroverted fundus and a normal-appearing cervix. The bilateral adnexae were unremarkable.

Her post-operative course was uncomplicated, and she was followed at our institution for six months. No adjuvant treatment was given, and she remained disease-free during her follow-up period.

3. Pathologic findings

The hysterectomy specimen consisted of a 280 g uterus with a smooth serosal surface distorted anteriorly by a mass. Sectioning revealed a partially cystic tan to yellow lesion within the myometrium that was 5.1 × 4.6 × 4.5 cm (Fig. 1). While the mass appeared circumscribed, areas of myometrium distant from the main lesion contained infiltrative, worm-like extensions of tumor. The endometrium was uninvolved by tumor.

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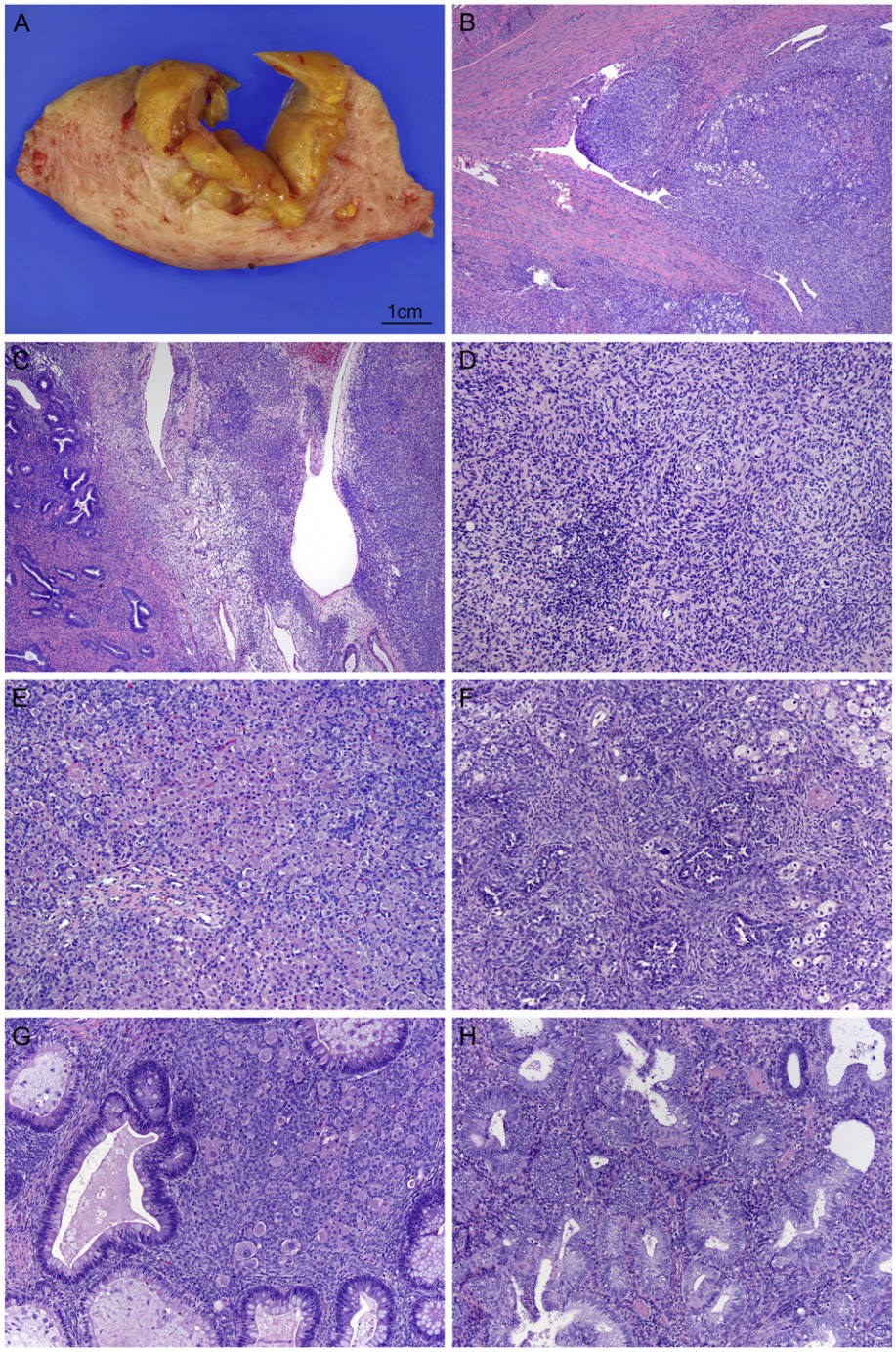

Fig. 1. A–B – Gross and microscopic sections through tumor demonstrate permeative growth into surrounding myometrium. C – Tumor (right) adjacent to uninvolved endometrium (left). Note the resemblance between benign and malignant endometrial stroma. D – Tumor cells whorl around a delicate capillary network. E – Sheets and nodules of Leydig-like cells occupied the majority of the tumor. F – Sertoli-like elements were also present. G – Heterologous glands resemble colonic epithelium with columnar absorptive cells and goblet cells. Leydig-like cells are also seen (center). H – Complex atypical hyperplasia is present in the overlying endometrium. Note the differing cytology of neoplastic glands compared with a single entrapped benign gland. (b–h. hematoxylin and eosin; b–c. 40×, d. 100×, e. 20×, f–h. 100×).

Microscopic examination revealed heterogeneous morphology (Fig. 1). The most striking feature was overwhelming sex cord-like differentiation, namely sheets and nodules of large polygonal cells with abundant eosinophilic foamy cytoplasm, central round nuclei, and prominent nucleoli, suggestive of Leydig cell differentiation and interspersed cords and tubules resembling Sertoli cell tumor. Intimately associated with the sex cord-like areas were glands lined by colonic-type epithelium with columnar absorptive cells interspersed with goblet cells. A minor component of uniform round cells with scant cytoplasm and minimal cytologic atypia, mimicking proliferative endometrial stroma, was present in the background. Irregular permeative growth into surrounding myometrium was identified, and there was focal lymphovascular invasion that included both endometrial stromal and sex cord-like elements. Altogether, the sex cord-like areas and heterologous elements comprised >60% of the tumor volume. The endometrium, which was grossly uninvolved by tumor, demonstrated complex atypical hyperplasia. The bilateral adnexa, all eighteen lymph nodes, and omentum were free of metastatic disease as were concurrent abdominopelvic washings.

Immunohistochemical studies were performed on multiple sections of tumor and adjacent benign tissue. The LG-ESS was positive for ER, PR,