Short communication

Primary extrauterine cystic low-grade endometrioid stromal sarcoma mimicking stromal endometriosis. A case report emphasizing the differential diagnosis and its potential local aggressive behavior

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Genital and pelvic endometriosis is a frequently encountered lesion and its importance rely on associated symptoms and its propensity for malignant transformation. In the present paper we comment on the importance of correctly diagnose the malignant transformation of an endometriotic lesion into a cystic low-grade endometrial stromal sarcoma, which is a very rare event. Moreover, we discuss the ability of a low-grade endometrial stromal sarcoma to locally recurr and the differential diagnosis with stromal endometriosis, a lesion that is very rare, almost always microscopic and solid.

Key words: stromal endometriosis, cystic endometrial stromal sarcoma, malignant transformation.

A 38-year-old patient, 8 gravida and 4 para, with morbid obesity and no relevant clinical history, presented with acute abdominal symptoms. Exploratory laparotomy identified a cystic lesion adjacent to the right ovary and fallopian tube, attached to the rectum, which was surgically removed. The 40 mm diameter unilocular cyst, with a smooth 5 mm thick wall and hemorrhagic content, was partially lined with unistratified endometrioid type of epithelium and presented multiple nodules of endometrioid type of stromal cells within the cystic wall and involving vascular spaces, positive for CD10, ER, PR and lacking atypia. Based on the cystic appearance and the dominance of stromal type of cells with benign features, the lesion was misinterpreted as cystic stromal endometriosis (Fig. 1). Hormonal therapy (Zoladex) for a period of 3 months was indicated but after one dose the patient declined further therapy.

Ten months later she presented with pelvic pain. Intraoperative examination revealed a 70 mm diameter solid tumor mass attached to the rectum and associated with a second 30 mm diameter solid nodule, involving the right ovary while the uterus was unremarkable. Microscopically, both solid nodules presented a diffuse proliferation of small, uniform tumor cells, resembling endometrial stromal cells, with round to ovoid nuclei, finely granular chromatin, poorly defined cell borders, low mitotic activity, positive for ER, PR and CD10. There was a proliferation of small vessels and arterioles present throughout the tumor stroma, while vascular invasion as well as irregular tongues of tumor cells invading the stroma

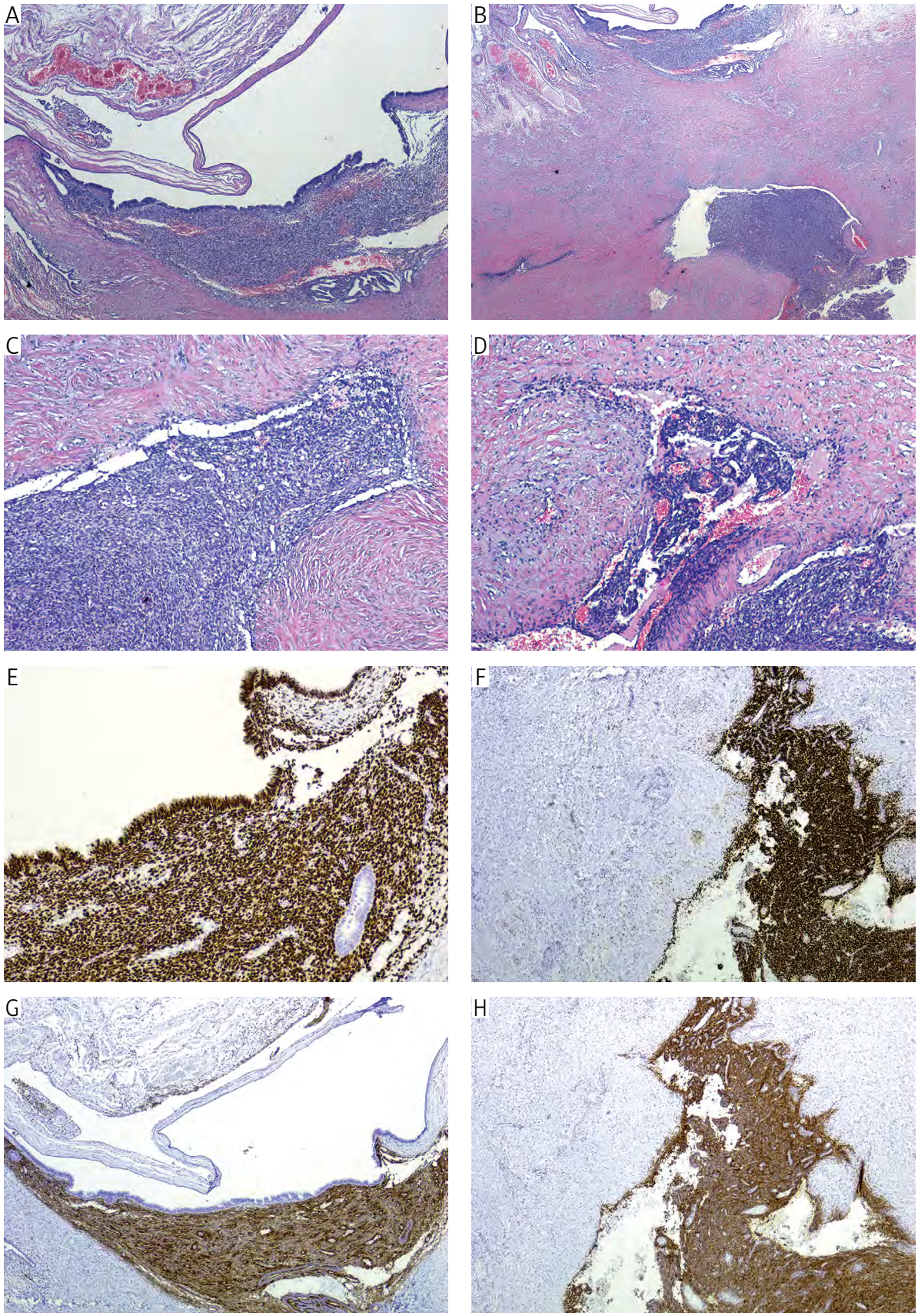

Fig. 1. Low-grade endometrioid stromal sarcoma: the first lesion was misinterpreted as cystic stromal endometriosis due to the cystic appearance, partially lined by unistratified endometrioid type of epithelium (A) although presenting multiple nodules of endometrioid type of stromal cells within the cystic wall (B, C) and involving vascular spaces where it presented an angiomatous pattern (D); the tumor cells were positive for PR (E, F) and CD10 (G, H)

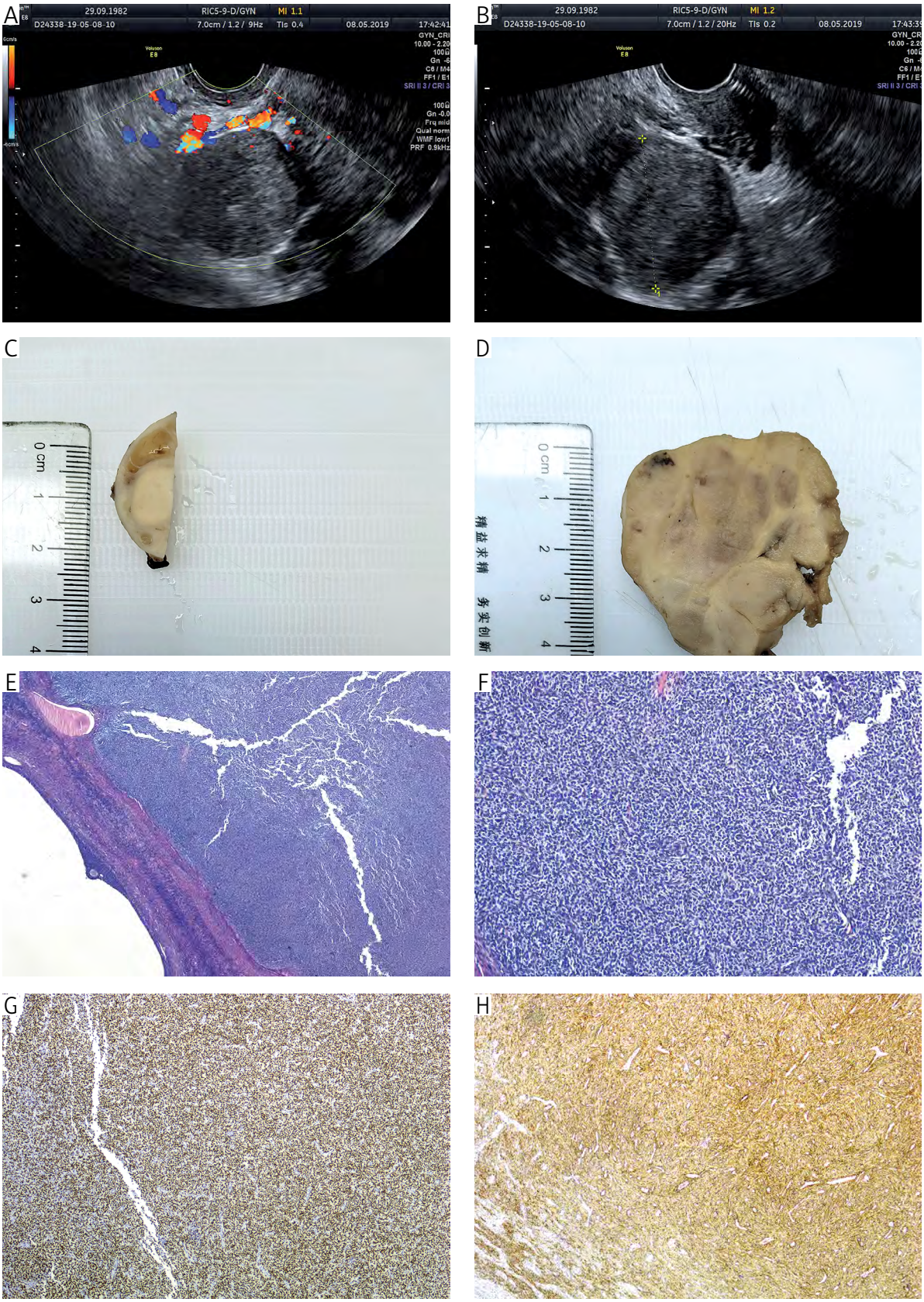

Fig. 2. Recurrence of a low-grade endometrioid stromal sarcoma: ultrasound examination revealed a lesion with unusual peripheral vascularization (A) and solid (B) located adjacent to the rectum; multiple coalescent tan to yellow soft nodules involving the ovary (C) and attached to the rectum (D) were solid in appearance; microscopically, both solid nodules presented a diffuse proliferation of small, uniform tumor cells, resembling endometrial stromal cells (E), with minimal nuclear atypia and mitotic activity (F); the tumor cells were positive for PR (G) and CD10 (H)

were only present in the nodule adjacent to the rectum (Fig. 2). The final diagnosis was of recurrent low-grade endometrioid stromal sarcoma involving the rectum and ovary. The surgical treatment was followed by radiotherapy and hormone therapy.

Endometriosis is a benign lesion characterized by the presence of endometrial tissue outside the endometrium and myometrium. Most frequently, endometriosis affects patients in their reproductive age, and involves organs of the female genital tract and pelvis, but involvement of the intestinal tract or remote organs is also frequently encountered. Depending on their duration and location in relation to the peritoneal surface, endometriotic foci may appear as punctate, spots, or patches, and may form nodules or cysts of various colors. Endometriotic cysts most commonly involve the ovaries or paraovarian tissue, rarely exceed 15 cm in diameter, are commonly covered with dense, fibrous adhesions, which may result in fixation to adjacent structures, and have a semifluid, chocolate-colored content material. Usually both endometrial epithelium and stroma are seen, but cases in which only one component is present can occur. Cases of endometriosis characterized by absence or paucity of glands, so-called stromal (or micronodular) endometriosis, are most commonly encountered in the superficial ovarian cortex, in the form of one or multiple small nodules of endometriotic stroma. Stromal endometriosis does not usually progress into cysts and it is clinically irrelevant.

Malignant transformation of ectopic endometriosis is infrequent, occurring in up to 1% of all women with endometriosis [1]. Most cases of malignant transformation occur in the ovary and are of epithelial type, represented by endometrioid and clear cell carcinoma, while mesenchymal tumors are very uncommon, representing less than 1% of all cases [1, 2, 3, 4]. Among mesenchymal malignant lesions, low-grade endometrioid stromal sarcoma can occur in the form of a solid, soft tan to yellow nodule. Very rare, low-grade endometrioid stromal sarcomas can develop as a cystic lesion or may contain benign-appearing or atypical endometrial glands, to the extent that confusion with endometriosis may occur.

In the present case, the first lesion was a cystic low-grade stromal sarcoma, misinterpreted as benign stromal endometriosis based on the cystic appearance and the presence of endometrioid type of epithelium despite the infiltrative pattern in association with vascular invasion. Pathologists should be aware of the fact that stromal endometriosis is very rare, almost never cystic, and should extensively sample lesions of this type to avoid misinterpretation.

The authors declare no conflict of interest.

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