

Collision tumour of endometrial stromal sarcoma, uterine tumour resembling ovarian sex-cord tumour, and leiomyoma

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Summary

Purpose of investigation: Endometrial stromal (ES) tumours may show different types of differentiation including smooth muscle and sex-cord like elements. A common problem is the differential diagnosis of ES tumour variants from other tumours that have smooth muscle or sex cord-like elements. We report a case of collision tumour that was composed of a low-grade endometrial stromal sarcoma (ESS) with sex-cord like differentiation, a uterine tumour resembling ovarian sex-cord tumour (UTROSCT), and a leiomyoma. **Result:** The patient was a 63-year-old multiparous woman who was referred to us with the complaint of recurrent abnormal uterine bleeding. Magnetic resonance imaging demonstrated a 3.4-cm-dia. solid tumour in her uterine cavity. We performed an abdominal total hysterectomy and a bilateral salpingo-oophorectomy. On gross and microscopic examination, the tumour consisted of a CD10-positive low-grade ESS, a CD10-negative UTROSCT, and a leiomyoma. **Conclusion:** ESS should be distinguished from UTROSCT and leiomyoma because of its malignant potential.

Key words: Endometrial stromal sarcoma; Leiomyoma; Sex-cord differentiation; Uterine neoplasm; uterine tumour resembling ovarian sex-cord tumour.

Introduction

Endometrial stromal (ES) tumours may show different types of differentiation including smooth muscle, fibrous or myxoid change, sex-cord like elements, and glandular differentiation.[1] Submucosal myomas, which are derived from myometrial cells just below the endometrium, favorably occur in endometrium.[2] Mixed endometrial stromal and smooth muscle (ES-SM) tumours are composed of a prominent component of smooth muscle and endometrial stroma. Although mixed ES-SM tumours are not part of the latest classification by the World Health Organization (WHO), when the smooth muscle component is more than 30% of the total volume, a tumour is generally categorized as a mixed ES-SM tumour.[3] A common problem is distinguishing ES tumours from other tumours that present a smooth muscle or sex-cord like element.

A uterine tumour resembling an ovarian sex-cord tumour (UTROSCT) is defined as a tumour with prominent sex cord-like differentiation in which there is no conspicuous endometrial stromal background.[4] Microscopically, ES sarcoma (ESS) may also reveal sex cord-like differentiation.[5] It is important to differentiate a UTROSCT from a low-grade ESS, because the former behaves in a benign fashion, whereas the latter has malignant potential.

We report here a case of collision tumour that was com-

posed of a low-grade ESS with sex-cord like differentiation, a UTROSCT, and a leiomyoma.

Case report

A 63-year-old postmenopausal Japanese woman (gravida 5, para 3) was referred to us with the complaints of recurrent abnormal uterine bleeding and anaemia. An intrauterine tumour was revealed by hysteroscopy. Her history included an appendectomy, and she had mild diabetes mellitus and hypertension.

On pelvic examination, the uterus was enlarged to over fist-size. Transvaginal echography revealed the intrauterine tumour with heterogeneous echogenicity. Endometrial cytology and tumour markers such as carbohydrate antigen (CA) 125, CA19-9, and CA72-4 were all negative. The results of other serological examinations were within the normal range. Magnetic resonance imaging demonstrated a 3.4-cm-dia. multiple nodulous tumour in the patient's uterine cavity (Figure 1a). Contrast-enhanced computed tomography demonstrated that the tumour was confined to the uterus, with no enlarged lymph nodes or distant metastases.

We performed a staging surgery for a presumed uterine leiomyosarcoma, including a total extra-fascial hysterectomy with bilateral salpingo-oophorectomy, and the intraoperative diagnosis was made using frozen sections. Be-

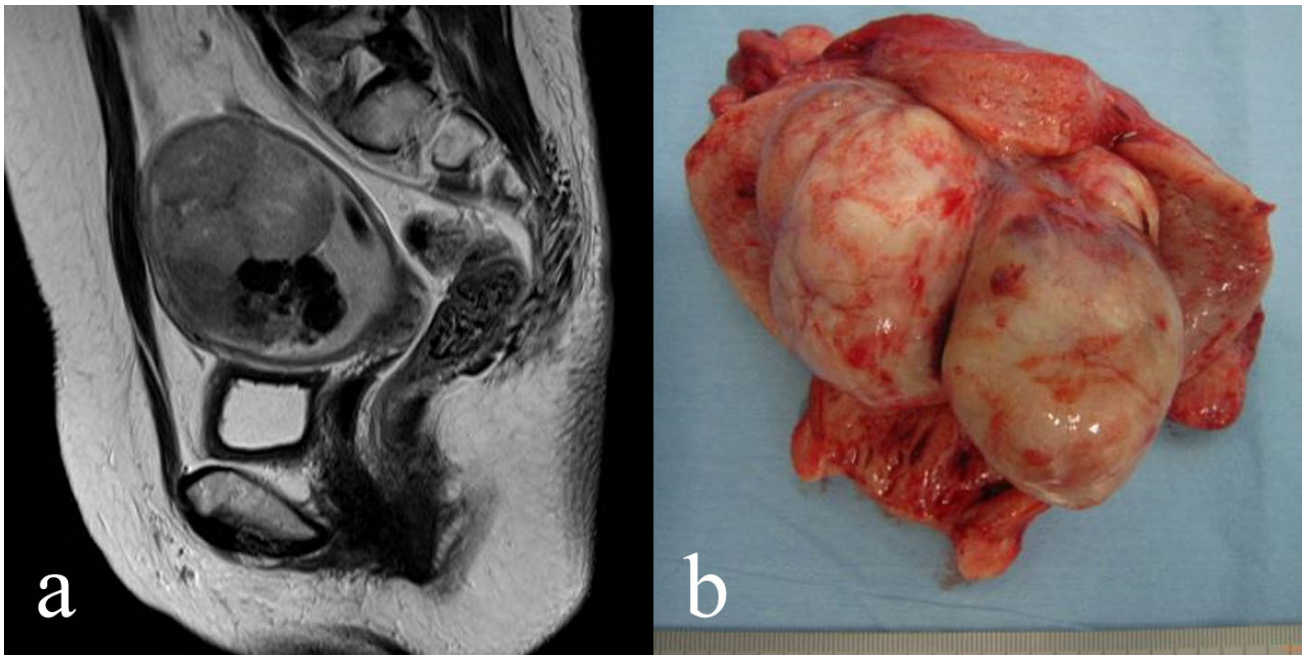


Figure 1. — (a) T2-weighted MRI showing the tumour's appearance as large masses without evidence of myometrial invasion. (b) The tumour was composed of three clearly distinguishable nodules.

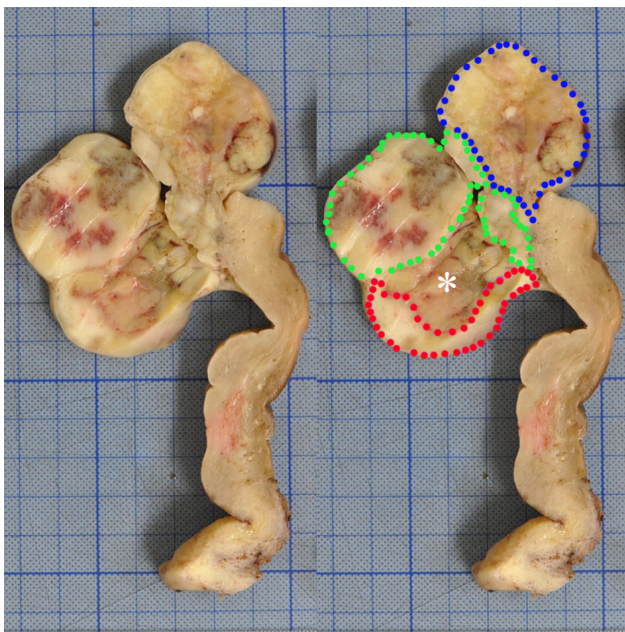


Figure 2. — On gross examination, all three pedunculated tumours arose from the endometrium; leiomyoma (blue dashed line), ESS (green dashed line), and UTROSCT (red dashed line). The insular lesion surrounded by the ESS and UTROSCT showed necrotic degeneration (asterisk).

cause the diagnosis based on frozen sections was a smooth muscle tumour of uncertain malignant potential and there were no findings of lymphadenopathy, we did not perform a pelvic lymphadenectomy.

The resected specimen revealed a fist-sized, soft in-

trauterine tumour occupying the entire uterine cavity that arose from uterine endometrium (Figure 1b). On gross examination, although each component of the tumour was side by side, the boundary line of each tumour was clear without invading the other components (Figure 2). Microscopically, the tumour was consisted of three histological types: (1) a leiomyoma with infarct-type necrosis (Figure 2a); (2) a tumour with a mixed pattern of cords, tubules, and nests related to sex cord-like differentiation (Figure 2b,c, right side) that was positive for CD10 (Figure 3d, right side), suggesting a low-grade ESS with sex-cord like differentiation; and (3) a tumour with cords of epithelioid or spindle cells and a hyalinised stroma (Figure 3b,c, left side) that was negative for CD10 (Figure 3d, left side), suggesting a UTROSCT.

The ESS component had extensive necrosis and the mitotic index 20/10 (MI = number of mitoses/10 high-power fields), but low reactivity for cyclin D1. The histological examination of the tumour tissue revealed an orthotopic and simultaneous tumour consisting of low-grade ESS, UTROSCT and leiomyoma. We diagnosed ESS Stage IB (pT1bN0M0) based on the 2010 International Federation of Gynecology and Obstetrics (FIGO) staging system. She did not receive adjuvant therapy but did undergo routine post-treatment surveillance. We followed-up the patient with monthly physical exams and echography, and she remains free from disease 6 months after the surgery.

Discussion

We report here a case of low-grade ESS with sex-cord differentiation that had extensive necrosis, a high MI, and

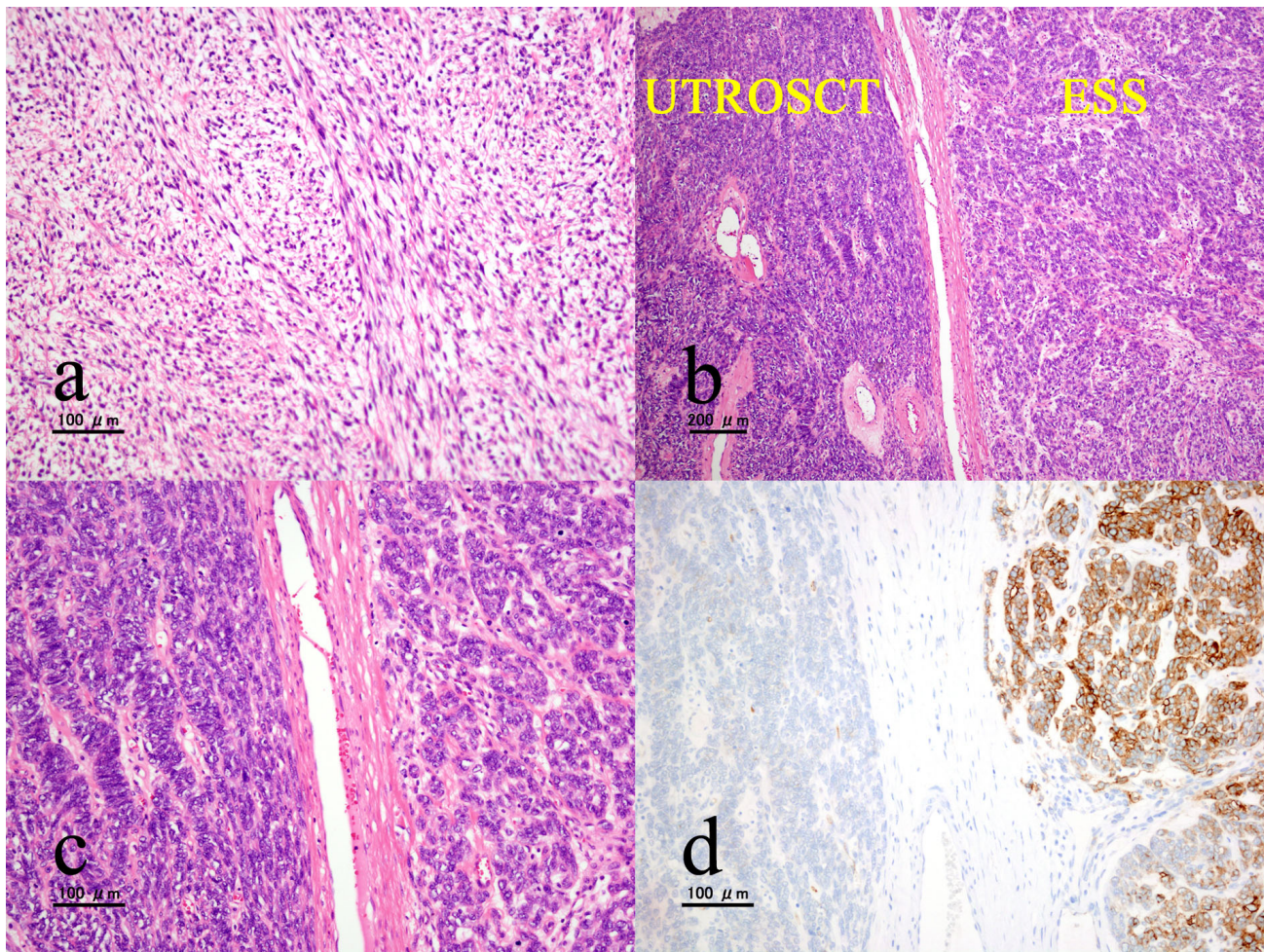


Figure 3. — (a) The leiomyoma was composed of spindle cells arranged in intersecting fascicles. (b) The UTROSCT was more eosinophilic than the ESS. There was a boundary between the two. (c) The UTROSCT showed higher cellular density and a more obvious cord-like structure compared to the ESS. (d) Unlike the ESS, the UTROSCT had no immunoreactivity for CD10.

positive immunoreactivity for CD10, but no destructive infiltrative growth into the myometrium and no immunoreactivity for cyclin D1. In their review of 68 cases of low-grade ESS with the median follow-up of 79 months, Feng et al. demonstrated that the presence/absence of ovarian preservation and the MI (0–3 vs. > 3) had independent prognostic value.[6] Additionally, in analysis of only Stage I low-grade ESS cases, neither the MI nor cytological atypia predicted a favorable prognosis [7] For this reason, our patient should be followed-up for an extended period of time from the prognostic point of view.

To our knowledge, this case is the first report in the English literature of a collision tumour containing the three components of ESS, UTROSCT and leiomyoma. Leiomyomas are benign monoclonal tumours arising from the smooth muscle cells of the myometrium. Low-grade ESS is a malignant subtype of endometrial stromal tumour arising from the stroma of the endometrium rather than the glands. The current WHO classification defines UTROSCT as a miscellaneous tumour, but its origin remains unclear.[8]

UTROSCT has been thought to behave in benign fashion, but four cases of disease recurrence or metastasis were reported [9] In the light of the small number of reported cases, the potential risk of UTROSCT remains uncertain.

The present case was collision tumour with a low-grade ESS, UTROSCT, and leiomyoma. From the prognostic point of view, it is important to distinguish between these three tumours, as leiomyoma and UTROSCT typically do not relapse and low-grade ESSs have a low malignant potential characterized by late recurrences. Because the entity of UTROSCT remains unclear, we should pay attention to the long-term prognosis of this disease. Further evaluation in more patients is necessary.

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Conflict of interest

Written informed consent for this report was obtained from the patient and her husband before the initial therapy. All authors have declared no conflict of interest.

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