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Research Letter

Rupture of a high-grade endometrial stromal sarcoma with intraperitoneal carcinomatosis and sepsis

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We present here the case of a woman whose high-grade endometrial stromal sarcoma ruptured and was accompanied by intraperitoneal carcinomatosis superimposed with sepsis caused by a urinary tract infection. The woman died 3 months after the debulking surgery, hysterectomy, salpingo-oophorectomy, and colostomy.

A 69-year-old woman suffered with post-stroke aphasia following a cerebral vascular accident 10 years ago. Her caregiver noted that she had abdominal distension, purulent vaginal discharge, and oliguria, so she was sent to our emergency room. The laboratory data showed leukocytosis, anemia, electrolyte imbalance, and urinary tract infection, and she was admitted to our infections ward for further care. During admission, we noted a large, palpable low abdominal mass and a significant amount of purulent vaginal discharge. The gynecological ultrasound showed a complex tumor about 15 cm in size located between the uterus and the rectum. Similar findings were noted in the computed tomography and included bilateral severe hydronephrosis with delayed renal excretory function due to pelvic mass compression or invasion. Between the uterus and sigmoid colon, we located an extremely large pelvic hypervascular mass about 15 cm in size with intratumoral bleeding (Fig. 1). Owing to the poor medical condition of the patient, the intra-abdominal local invasion into the sigmoid colon, and the massive tumor bleeding during the operation, we performed a maximal debulking of the tumor (no obvious gross tumor could be seen), a total hysterectomy with bilateral salpingo-oophorectomy, and a colostomy. The total blood loss during the surgery was about 4000 mL.

The tumor was highly necrotic, arose from the posterior wall of the uterus, and invaded directly into the adjacent sigmoid colon. The uterus was atrophied and filled with purulent fluid, and the vagina had been extremely elongated by the tumor (Fig. 2A–D). From the microscopic findings, the histologic type of the endometrial stromal sarcoma (high-grade), the tumor cells were a mixture of irregular short spindle and ovoid nuclear shapes, with vesicular nuclei, frequent mitotic activity, central necrosis and hyaline change. The immunohistochemical stains showed the tumor cells to be focal positive for CD10 (20%) and ki-67 (30%) (Fig. 3), but negative for desmin and smooth muscle actin stains (Fig. 4). Based on these pathological findings, high-grade endometrial stromal sarcoma (undifferentiated stromal sarcoma) was considered. The final histopathological findings were high-grade endometrial stromal sarcoma with colon invasion, no assessable lymph node, FIGO IVA (pT4aNx), and malignant washing cytology. After discussing her poor prognosis with her family, the decision was made not to administer adjuvant therapy.

Unfortunately, the patient suffered from abdominal distension, poor appetite, and fever for 2 months after the operation. Her urinary analysis showed pyuria, and her white blood cell count was 18,000/μL with a left side shift. The tumor marker CA-125 was elevated (75–194 U/mL). The small bowel series showed a bilateral double-J *in situ*, smooth contrast passage from the stomach to the colostomy, and no significant intestinal dilatation or evident filling defect could be seen. The empirical antibiotic cefuroxime (Zinacef) was given for urosepsis control. The sepsis was in progress and the patient became irritable with her abdomen distended and hard. An abdominal computed tomography showed severe intraperitoneal seeding of the tumor, carcinomatosis, retroperitoneal and inguinal lymph node metastasis, and bilateral severe hydronephrosis. After further discussion with her family, they decided she should receive palliative care. She passed away a few days later (3 months after the operation).

Uterine sarcomas represent between 2% and 5% of all uterine malignancies. There are three types of uterine sarcomas: carcinosarcoma (also known as a mixed Müllerian tumor) is the most common, leiomyosarcoma is the next, and

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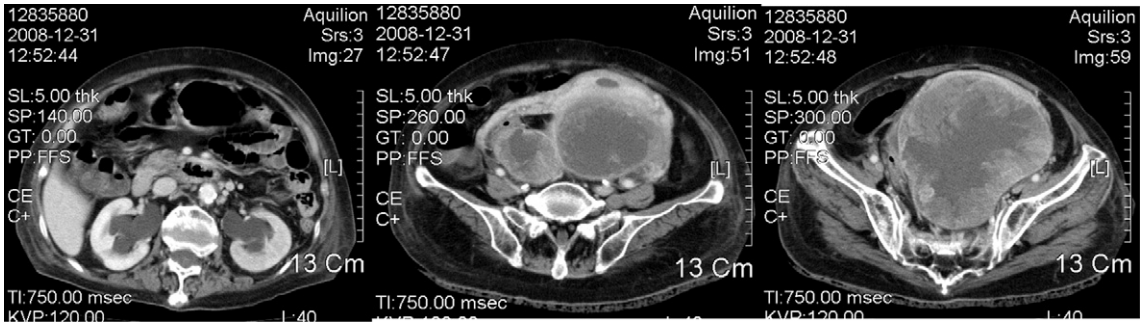

Fig. 1. Computed tomography of the abdominal region shows bilateral hydronephrosis, fluid accumulation in the uterus, a right adnexa tumor mass, and a large necrotic tumor (about 15 cm in size) between the uterus and the rectum. No obvious retroperitoneal lymph nodes could be identified.

endometrial stromal sarcoma (ESS) is the least common (< 10% of uterine sarcomas). Endometrial stromal sarcomas are composed of cells that are identical to or closely resemble normal proliferative-phase endometrial stromal cells. These tumors may arise from the endometrium, but can also be seen in the setting of adenomyosis or endometriosis. Endometrial stromal sarcoma typically occurs in women between the ages of 42 and 59 years. Symptoms include abnormal uterine bleeding and pain. Risk factors are fairly nonspecific and include prior pelvic radiation therapy, age, and race (a slightly higher incidence among black people). There may also be hormonal factors, though this is not well understood.

The differential diagnosis for ESS includes leiomyoma with cystic degeneration, leiomyosarcoma, and endometrial carcinoma. The subtle differences between these options are best demonstrated with magnetic resonance imaging (MRI). On the MRI, endometrial stromal sarcomas appear as large tumors in the endometrial cavity and/or myometrium.

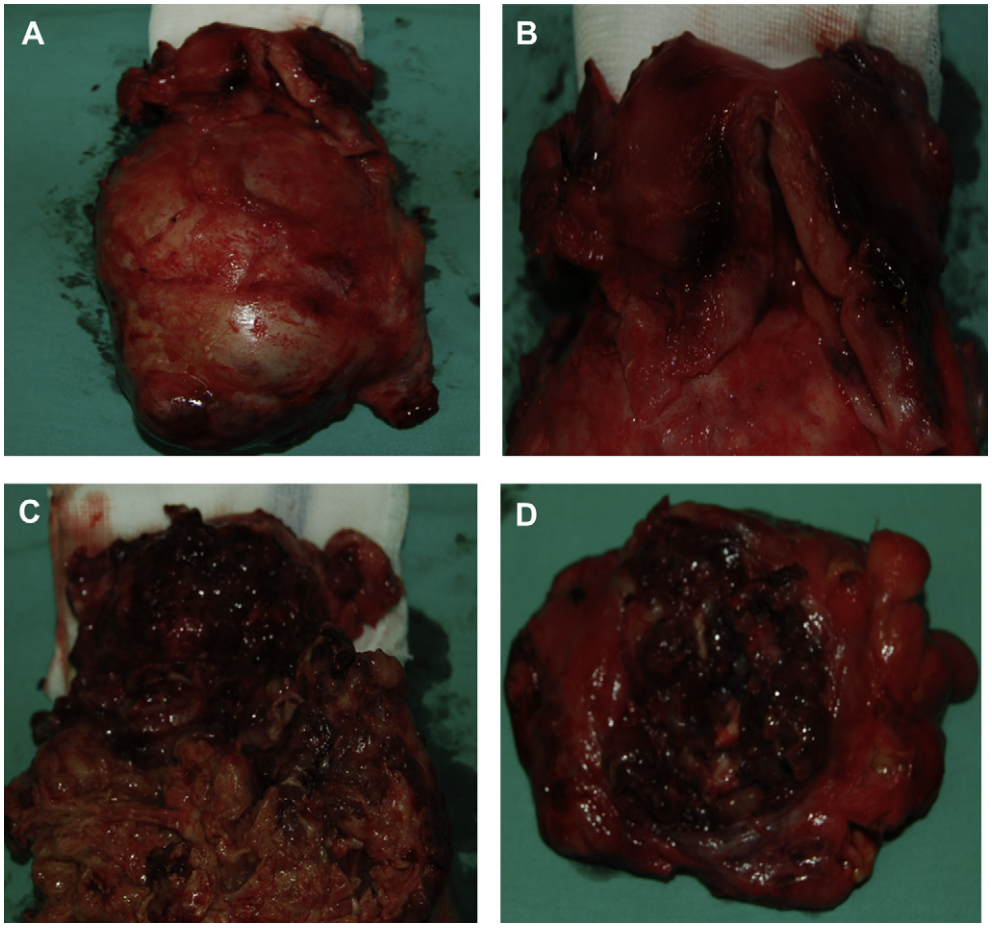

Fig. 2. (A) Large necrotic tumor mass protruding from the posterior wall of the uterus toward the cul-de-sac and rectum, massive bleeding occurred during manipulation, the anterior surface of the tumor was relatively smooth and intact; (B) close-up view of the cervix and the atrophic uterine body and adnexa; (C) close-up view of the posterior surface of the tumor, marked necrotic change, and invasion into the sigmoid colon; (D) segmental resection of the invaded sigmoid colon.

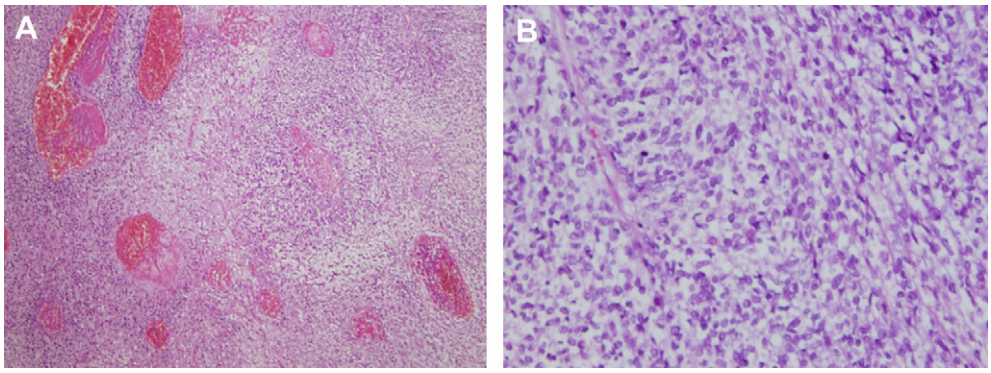

Fig. 3. (A) Tumor cells were a mixture of irregular short spindles and ovoid nuclear shapes, with vesicular nuclei, frequent mitotic activity, and central necrosis and hyaline change; (B) close-up view.

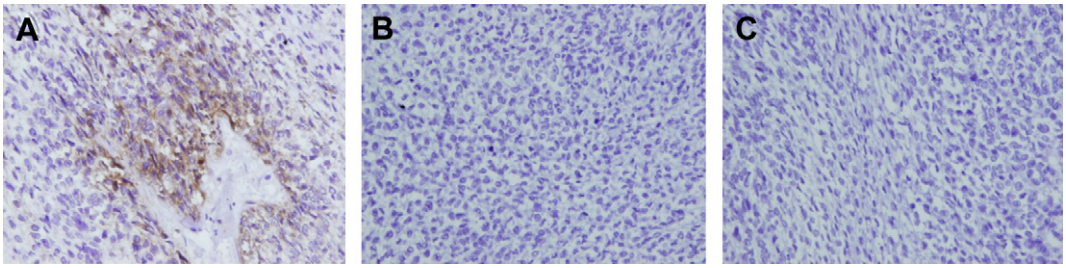

Fig. 4. The immunohistochemical stains showed the tumor cells to be focal positive for CD10 (20%) and ki-67 (30%), but negative for desmin and smooth muscle actin stains. (A) CD10; (B) desmin; (C) smooth muscle actin.

Intramyometrial worm-like nodular extensions may be visible. Hemorrhage and necrosis are common. Most ESS show greater than normal enhancement of the myometrium. Overall, ESS is difficult to differentiate from leiomyomas with cystic degeneration, leiomyosarcomas, and endometrial carcinomas [1–3].

We have presented a case of a rare uterine malignant tumor and its rapid progression, and shared our limited experience in managing this complicated condition.

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