Myxoid Endometrial Stromal Sarcoma: A Case Report and Literature Review

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Abstract
Myxoid type of endometrial stromal sarcoma (ESS) is a very rare tumor of uterine malignancies. Although several cases about pathologic features for the diagnosis of ESS have been reported, it remains still difficult to find a study of the clinical course, management, and outcome of ESS. Here we report a postmenopausal woman with myxoid ESS of low grade type who had aggressive clinical features. Although adequate progestin therapy was provided followed by total abdominal hysterectomy and bilateral salpingo-oophorectomy, they failed to prevent early recurrence of ESS. Following cytoreductive surgery and chemotherapy were also proved to be refractory to control the rapid progression of ESS.

Keywords: Endometrial stromal sarcoma; Uterine cancer; Recurrence

Introduction
Endometrial stromal sarcoma (ESS) is a type of uterine sarcomas that constitute 8% of uterine malignancies [1]. According to World Health Organization (WHO) classification, ESS is divided into low grade ESS and undifferentiated endometrial sarcoma according to features such as nuclear pleomorphism and necrosis [2]. Low grade ESS generally occurring in perimenopausal women is indolent. However, its recurrence rate is 36% to 56% in patients with early stage [3,4]. We report a 52-year-old menopausal woman with myxoid ESS of the low grade type. Several case reports about this disease have focused on its unusual pathologic features more than its treatment and prognosis [5,6]. In the present case report, distinct characters of ESS are presented, unlike previous cases. Our report may provide informative data about the clinical course and management for myxoid ESS of low-grade type.

Case Presentation
A 52-year-old woman (Gravida 2, Para 2) visited a local hospital with a complaint of low abdominal pain for two months. She was a patient with an alleged leiomyoma that was measured at 5.3 cm on 2013 by transvaginal ultrasonography (TVS) after menopause. At this time, the mass was increased. It seemed to be a secondary degeneration. She was referred to the Department of Obstetrics and Gynecology at Ewha Womans University Medical Center. Pelvic exam revealed a 2-3 months gestational sized pelvic mass. Ultrasonic examination showed 7.47 cm² × 6.59 cm² sized complex mass in uterus. The border was not clear to demarcate between endometrium and myometrium. In magnetic resonance imaging (MRI), the mass in the posterior wall of the uterus suggested a leiomyoma with cystic degeneration, a leiomyosarcoma with necrosis, or an endometrial stromal sarcoma. Enlargement of lymph node and other abnormality in pelvis were not found. Tumor markers were as follows: carcinoembryonic antigen (CEA), 1.4 ng/mL (normal reference, less than 0.7 ng/mL); CA 19 9/15 to 9, 25.3 U/mL (normal reference, less than 3.6 U/mL); CA-125, 21.6 U/mL (normal reference, less than 35 U/mL).

Total abdominal hysterectomy with bilateral salpingo-oophorectomy was done on June 1st, 2015. Severe pelvic adhesion was observed between peritoneum, uterosacral ligament, and uterus. Result for frozen pathology was spindle cell tumor with marked myxoid change, inflammatory myofibroblastic tumor with myxoid change, or myxoid leiomyosarcoma.

Grossly opened uterus showed that a whitish gray to yellow solid mass with central cystic change. (Figure 1A). On microscopic examination, 80% of the mass contained abundant myxoid stroma with low to moderate cellularity (H&E, x100). (Figure 1B). The rest was composed of hypercellular area without myxoid stroma. Beyond myometrium, uterine serosa was consistent with the tumor. Tumor necrosis and lymphovascular tumor emboli were not present. Results of immunohistochemical study were as follows: CD10, positive (Figures 1C and 1D); SM-Actin, negative; CK, negative; S-100, negative; Desmin, negative; C-kit, focal positive. Pathologic diagnosis was myxoid ESS of low grade. Both ovaries were intact.

She was discharged on postoperative day 5 in good condition. Evaluations about distant metastasis after recovery showed non-specific.

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Low grade ESS is composed of cells resembling those of endometrial stroma. It invades myometrium and plexiform arterioles linked to spiral arterioles of the endometrium [7]. It mainly occurs in women between age of 45 and 50 years, causing symptoms such as abnormal vaginal bleeding and low abdominal pain. However, 25% of these patients are asymptomatic. At diagnosis of low grade ESS, metastasis occurs up to one-third. The most common organ is ovary, although metastases of lung, bone, and bladder have been reported [8]. A study on 831 patients with all stages of ESS has shown that the 5-year disease-specific survival rate is more than 90% [9].

Discussion

ESS has been traditionally classified as low or high grade type. Considering that high grade ESS lacks nature of endometrial stroma, WHO adopted ‘undifferentiated endometrial sarcoma (UES)’ instead of ‘high grade ESS’ in 2003. Therefore, low grade ESS and UES are divided according to features such as nuclear pleomorphism and necrosis instead of mitotic count. [2]

At 3 months post chemotherapy, she had consistent constipation and dysuria for 3 weeks. PET-CT revealed hypermetabolic masses in pelvic cavity and hydronephrosis in both kidneys. On July 21st, 2016, we visited our clinic with complaints of tenesmus and constipation. Vaginal vault was clear. Rectal exam revealed left sided fixed tumor. CA 19-9 level was mildly elevated to 7.4 U/mL. On PET-CT, cystic mass on vaginal stump pressed recto-sigmoid junction. Bilateral common iliac nodes were free of tumor. Between December 18th, 2015 and April 3rd, 2016, the patient received 6 cycles of adjuvant chemotherapy consisting of Ifosfamide and Cisplatin every 3 weeks. After the 6th chemotherapy, the inhibition rate for each chemotherapeutic agent as following: Docetaxel, 82%; Taxol, 81%; Topotecan, 80%; Cisplatin, 79%; Carboplatin, 78%. As a result, she received 6-cycle of Paclitaxel-Carboplatin chemotherapy between August 13th and November 11th, 2016. Sequential radiographic studies and symptoms showed aggravation. She had consultation with the radiation oncology department. However, they did not recommend radiotherapy due to bowel complication. Although the patient was started on 800 mg pazopanib daily for 22 days, she expired on January 23rd, 2017, 20 months after the initial diagnosis.

Our case provides insights that myxoid ESS of low grade type could have different clinical course compared to usual low grade ESS. First, the interval to relapse is much shorter than that of typical low grade ESS at stage I. It is well-known that low grade ESS grows slowly. In our case, 6 months after the diagnosis, recurrence on pelvis occurred. Previous two case reports of myxoid ESS noted that there was no evidence of recurrence after surgery within 21 months. One was treated adjuvant chemotherapy with Adriamycin and dacarbazine [6], while the other was a 16-year old woman who underwent mass excision of uterus and adjuvant progestin therapy [14]. Second, 13 months after the diagnosis, the myxoid ESS of low grade type in the present case was transitioned into high grade sarcoma. This might be related to the loss of hormonal sensitivity of tumor. [15] Therefore, it is important to understand tumor biology to establish therapy strategy. As adjuvant therapy after operation was done based on reported cases, our report may provide informative data to provide treatment option for this rare disease.

Conclusion

In conclusion, we report a rare case of myxoid ESS of low grade.
There are a few studies of myxoid ESS about its clinical course. They have suggested that myxoid ESS is similar to typical low grade ESS in recurrence and survival time [6,14]. However, our case provides a different view about this disease.

References


