Myxoid Endometrial Stromal Sarcoma: A Case Report and Literature Review

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Abstract

Myxoid type of endometriall 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 cm 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 was an ill-defined bulging out whitish gray (Figure 1A). On microscopic examination, 80% of the mass contained abundant myxoid stroma with low to moderate cellularity and partially dense inflammatory cell infiltration (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. Grossly opened uterus showed that a yellowish mass with extensive gelatinous appearance bulged out to posterior myometrium, measuring 8.3 cm³ × 5.6 cm³ × 2.7 cm³ (Figure 1A). On microscopic examination, 80% of the mass contained abundant myxoid stroma with low to moderate cellularity and partially dense inflammatory cell infiltration (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].

We reported a low-grade type of myxoid ESS. Although its microscopic features are the same as usual low grade ESS, it is not easy to diagnosis owing to an abundant myxoid matrix and hypocellularity. Immunohistochemistry is essential to aid differential diagnosis among other uterine tumors. Low grade ESS is positive for CD10 which might also be immunoreactive in uterine mesenchymal tumor and smooth muscle tumor. As ESS represents estrogen and progesterone receptor, immunostaining for these is positive. In addition, ESS is negative for SM-Actin while smooth muscle cell tumor is positive. S-100 is negative for its differential diagnosis from neurogenic tumor [10].

Hysterectomy with bilateral salpingo-oophorectomy is the treatment of choice for low grade ESS at early stage in peri- or post-menopausal women. [8] The issue of lymphadenectomy is unsettled except for cytoreductive surgery or enlarged lymphnode on preoperative imaging.

In recent research studies, the incidence of lymphnode metastases in ESS is reported to be 9.9% (28/282) or 7% (7/100). [9,11] Progestin or aromatase inhibitors can be considered for adjuvant therapy. However, currently there is no standard for appropriate dose, regimen, or duration of treatment because there are insufficient prospective and retrospective data [12].

Although low grade ESS is indolent, relapse can occur in 36% to 56% of patients at early-stage. Organs of recurrence include pelvis, lungs, and abdomen. The median time to recur is 5.4 years for stage I and 9 months for stage III-IV [3,4]. Cytoreductive surgery should be considered preferentially as the treatment for recurrence. Repeated surgery with resection of metastatic organ is acceptable. It has been reported that hormone therapy using progestins, gonadotrophin-releasing hormone agonists, and aromatase inhibitors is effective compared to radiotherapy and chemotherapy. Chemotherapy could be prescribed on ineffective hormone therapy and transition into high grade sarcoma. Ifosfamide and doxorubicin are the most frequently used [13].

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 with 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.

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]

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.

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