CASE REPORT

Fertility-Sparing Surgery for Patients with Endometrioid Stromal Sarcoma of Ovary: A Case Report with a Review of the Literature

Bo Ra Park

Department of Obstetrics and Gynecology, Soonchunhyang University Seoul Hospital, Seoul, Korea

Endometrioid stromal sarcoma (ESS) of ovary is very rare disease. These mesenchymal neoplasms occur most commonly in the uterus and occasionally originate from extra-uterine sites, such as the ovary. It is often associated with endometriosis of the ovary. Here we present this rare case to emphasize on the clinicopathologic features and fertility-sparing surgery outcome of ovarian ESS in patients with endometriosis.

Keywords: Endometrioid stromal tumor; Ovarian neoplasms; Fertility preservation

INTRODUCTION

Primary endometrioid stromal sarcomas (ESS) of the ovary are rare mesenchymal tumors. It occurs in women over wide age range of 11 years to 76 years accounting for only 0.2% of all uterine malignancies and for 10%–15% of primary uterine sarcomas [1]. ESS is originated from cells that resemble those of the endometrial stroma during the proliferative phase of the menstrual cycle [2]. When uterine in location, ESS is easy to diagnose because of their characteristic histology and patterns of invasion. However, when they occur at extrauterine sites, they produce nongynecologic signs and symptoms. The majority of extrauterine ESS arises from endometriosis, suggesting that most of these tumors originate by malignant transformation of the stromal component of endometriosis [3]. ESS arising from endometriosis is commonly well differentiated and considered as an indolent tumor with good prognosis.

We recently encountered a case of ESS with endometriosis in pre-menopause woman. To provide an insight into the sites of occurrence, clinicopathologic features and fertility-sparing surgery outcome of ESS, we report a case of ESS with endometriosis with literature review.

CASE REPORT

A 25-year-old, unmarried woman came to gynecology outpatient department with the complaint of both ovarian mass which was noticed January, 2013, by accidentally. Her menstrual cycles were regular but were associated with dysmenorrhoea. Routine papanicolaou smear showed reactive cellular changes with inflammation. Ultrasound examination and computed tomography (CT) revealed thin-walled slightly high attenuated (about 30 Hounsfield unit) lesion measuring 60 × 31 mm in right adnexa (Fig. 1). There was no abnormal lymphadenopathies or fluid collection in abdomen and pelvis. And left adnexa had 40 × 29 mm monoloculated diffuse hypoechoic mass (Fig. 2). On physical examination, the patient was generally well, weighed 57.6 kg and not pale. Hematological examination was within normal range. The patient underwent laparoscopic right ovarian pseudocystectomy with removal of left ovarian mass with adhesiolysis on March 7, 2013. At operation, right ovary had 6-cm sized pseudocyst include chocolate like material and adhered to uterus posterior wall. Left ovary had 4-cm sized yellow and fragile solid mass adhered to uterus posterior wall (Fig. 3). Tumor arising from left ovary consists of multiple fragments of partly brownish and partly yellowish, friable soft tis-

Correspondence to: Bo Ra Park

Department of Obstetrics and Gynecology, Soonchunhyang University Seoul Hospital, 59 Daesagwan-ro, Yongsan-gu, Seoul 04401, Korea

Tel: +82-2-709-9329, Fax: +82-2-709-9790, E-mail: borapark@schmc.ac.kr

Received: Oct. 22, 2018 / Accepted after revision: Nov. 5, 2018

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Fig. 1. 52×34-mm sized thin-walled slightly high attenuated (about 30 Hounsfield unit) mass in right adnexa at enhanced computed tomography.

Fig. 3. Right ovary had 6-cm sized pseudocyst include chocolate-like material and adhered to uterus posterior wall. Left ovary had 4-cm sized yellow and fragile mass. This mass adhered to uterus posterior wall.

Fig. 2. Transverse ultrasonography image shows 40×29-mm monolocular diffuse hypoechoic mass in left adnexa (A) and 60×31-mm sized hypoechoic multiloculated mass in right adnexa (B).

sue. Specimen retrieval bag was used for contained removal of adnexal masses because of the concern regarding spread of malignant cells. Final pathology was consistent with left ovarian ESS with endometriosis. Grossly, the left ovarian tumor consists of multiple fragments of irregularly shaped brown and yellowish fragile soft tissue. Microscopically, the tumor shows diffuse proliferation of small round cells with a few small arterioles. A transition between the tumor and endometriosis is seen. The tumor is composed of relatively small cells with oval to fusiform nuclei and inconspicuous nucleoli (Fig. 4). The tumor was strongly and diffusely positive for estrogen receptor, progesterone receptor, and desmin. CD10 and Ki-67 (less than 5%) were focally positive.

However, calretinin, D2-40, CD34, and inhibin-alpha were all negative (Fig. 5).

When histological results confirmed the diagnosis of ESS, we performed positron emission tomography (PET)-CT and rescheduled for laparoscopic left oophorectomy for preserving fertility. Torso PET-CT showed 11×11×10-mm sized hypermetabolic nodule in left ovary, cystic mass in right adnexa without FDG uptakes. No evidence of lymphadenopathy in entire pelvis, abdomen, and mediastinum. In March 26, 2013, laparoscopic left oophorectomy, endometrial biopsy, adhesiolysis, and washing cytology was done. No evidence of tumor was found in the endometrium. The result of peritoneal cytology was negative for malignant cells, but a

Fig. 4. (A) Grossly, the tumor consists of multiple fragments of irregularly shaped brown and yellowish fragile soft tissue. (B) Microscopically, the tumor shows diffuse proliferation of small round cells with a few small arterioles (H&E, $\times 40$). (C) A transition between the tumor and endometriosis is seen (H&E, $\times 100$). (D) The tumor is composed of relatively small cells with oval to fusiform nuclei and inconspicuous nucleoli (H&E, $\times 400$).

few clusters of reactive mesothelial cells are seen. We have not encountered any recurrence in the study at this time following conservative treatment.

DISCUSSION

ESSs are characterized by infiltrative growth of cells resembling normal proliferative-phase endometrial stroma. Endometriosis is commonly identified adjacent to the neoplastic tissue, so it suggests that there would be correlation between ESS and endometriosis. Young et al. [4] found that endometriosis presented in 11 cases among 23 ESS cases. According to Baiocchi et al. [5], 29 of 45 cases of extrauterine ESSs qualified as arising from endometriosis

by Sampson's criteria [6]. Above Sampson's three criteria was (1) clear examples of endometriosis in close proximity to tumor, (2) no other primary site of malignancy, and (3) histological appearance compatible with origin from endometriosis. Baiocchi et al. [5] found approximately half of the patients were nulliparous. It would be explained by that 30% to 40% of patients with endometriosis are infertile. However, Ober and Black [7] suggest another possibility for the origin of this tumor is metaplasia from the ovarian surface mesothelium or from the subcoelomic mesenchyme.

Tumors are usually solid, but others are solid and cystic and sometimes cystic. The surfaces of solid areas are yellow-white and some cases have foci of hemorrhage or necrosis. Our case also has yellow and fragile mass that identified ESS. Microscopic features

Fig. 5. Immunostaining of the tumor. The tumor cells are positive for estrogen receptor (A), progesterone receptor (B), and CD10 (C) and low level of MIB-1 expression (D).

of ESSs are typically composed of sheets of uniform cells, which are usually round or oval, but occasionally have a spindle shape and resemble the stromal cells of normal proliferative endometrium. Numerous small blood vessels resembling the spiral arterioles of late secretory endometrium are present. The neoplastic cells usually cluster around these vessels. The nuclei are oval to round and nucleoli are present but not prominent. The cytoplasm is pale and scanty and the cell borders are not clearly recognizable. There is sometimes fibromatous areas. Foam cells and vascular invasion often present in the tumors [4].

Immunohistochemically, tumors express vimentin, muscle markers like desmin, muscle-specific actin and α -smooth muscle actin, and progesterone receptors [8]. Recent studies have described the unique expression of CD10 in endometrial stromal cells as well as in ESS, which was proven diagnostically useful in

the distinction of ESS from smooth muscle neoplasms like low-grade leiomyosarcoma [9]. Other study suggests CD 34 might be helpful in diagnosing extrauterine extraovarian ESS [10].

Tumors with less than 10 mitosis per 10 high power fields (HPF) are classified as low grade and those with 10 or more as high grade. Chang et al. [11] reported that neither mitotic index nor cytologic atypia in primary extrauterine endometrial stromal neoplasm were predictive of tumor recurrence or death from tumor. In their study, 77% of patients whose tumors had a mitotic index of less than 10 per HPF had one or more recurrences and 30% died from their neoplasm.

Low-grade ESS has an indolent clinical course with a tendency for late recurrence. In stage I low-grade ESS, 5- and 10-year survival rates have been estimated to be 98% and 89%, respectively.

Primary therapy is complete resection of neoplasm. Thus, if the

patient is perimenopausal or postmenopausal, hysterectomy with bilateral salpingoophorectomy is recommended. The effect of chemotherapy in both low grade and high grade endometrioid stromal sarcomas is unclear. Cyclophosphamide, adriamycin, vincristine, methotrexate is used and some case have complete remission, but follow-up is short [4]. Also there is no clear evidence about effectiveness of radiotherapy and progesterone therapy. The risk of recurrence is as high as 50% along with interval for recurrence and a median interval of about three years from hysterectomy to relapse [12]. Because performing fertility-sparing surgery for low-grade ESS is rare, and because the long-term follow-up results are still lacking, it is not possible to draw any definitive conclusion about its safety.

In conclusion, ovarian ESS has an indolent clinical course, and is associated with a favorable prognosis; ovary-sparing procedures might be considered in younger patient. However, no standard treatment strategy has yet been established. Future investigation should include more study participants and focus on the response to treatment and prognosis.

ACKNOWLEDGMENTS

This work was supported in part by the Soonchunhyang University Research Fund.

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