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CASE REPORT

Successful Pregnancy with Endometrial Stromal Sarcoma (ESS)

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Introduction

Endometrial stromal sarcoma (ESS) is very rare malignant tumor that constitutes approximately 10 % of all uterine sarcomas but only 0.2 % of all uterine malignancies [1]. ESS can be mistaken for leiomyoma because clinical presentation mimics each other and often needs HPE to establish correct diagnosis [2, 3]. In this article, we report a case of ESS in a young unmarried female of 23 years, which was misdiagnosed as leiomyoma, but the patient was able to conceive and deliver a healthy baby after debulking surgery to remove myoma was mistakenly done.

Case Report

A 23-year-old girl presented with excessive bleeding and pain during menses in Aug 2005. On PA examination, a mass was palpable up to pubic symphysis. The ultrasound examination on her revealed a 5 × 4 cm hypoechoic lesion with multiple cystic areas in posterior wall of uterus, suggestive of fibroid with cystic degeneration. Both ovaries were normal. Although was advised for myomectomy, she refused.

Patient appeared in May 2007 with the same complaints with severe anemia (Hb 6 g%). On examination, mass was palpable up to the umbilicus. She was planned for myomectomy on 25/5/2007. At laparotomy, 10–15-cm-sized soft yellowish degenerated mass was found in myometrium which was excised. There was no clear cut capsule surrounding the mass; on the other hand, there were small fingerlike extensions entering the myometrial wall. On HPE, the mass was diagnosed as low grade ESS of body uterus. Although the patient was advised chemotherapy and Wertheim's hysterectomy, she refused.

The patient again presented on May 13, 2008 with marital life of 4 months and amenorrhea (1 month 5 days), nausea, and vomiting. Her pregnancy test was positive and transvaginal USG showed 14 × 11 cm mass in uterus, but no Gestational Sac was visible. Both adenexa were normal. Hence, she was advised MRI pelvis. On MRI, there was 6 week's live intrauterine pregnancy with a mass of around 15-cm size suggestive of relapsed ESS. She decided to continue the pregnancy with mass in situ. All possible complications were explained to her. Her antenatal course was uneventful, and on January 1, 2009, her full term lower segment cesarean section with obstetric hysterectomy and left salpingoophorectomy was done; samples were also collected from omentum and lateral pelvic walls. She gave birth to a 2.8-kg healthy male child. HPE of tissue revealed leiomyoma with hyalinisation without any metastasis. Her postoperative course was uneventful. The patient refused any kind of postoperative treatment for her malignancy.

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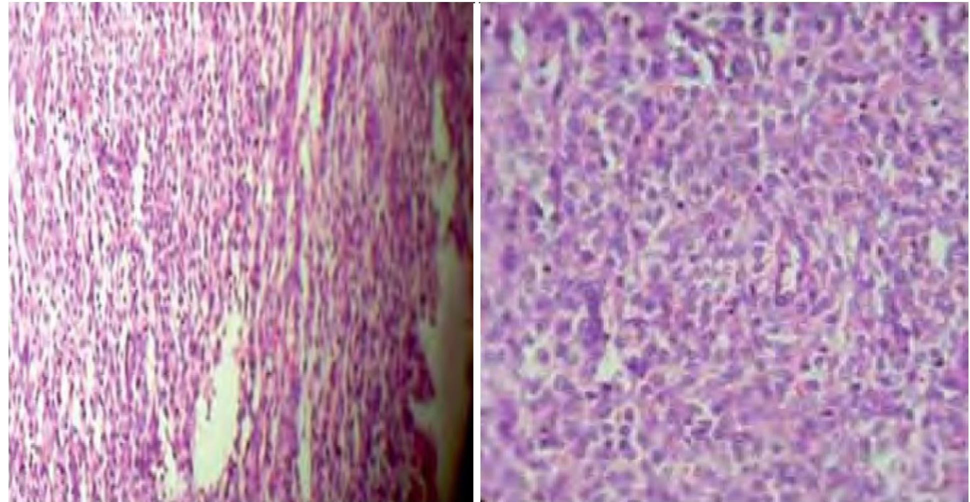

Fig. 1 Low grade ESS

This patient again relapsed in September 2010 with a complaint of nodule in lower anterior abdominal wall. MDCT Scan showed heterogeneously enhancing nodular regions at the right-sided vaginal wall, right ovary and involving left rectus sheath in hypogastrium. Biopsy from nodule showed low grade ESS (Fig. 1), and so she was planned by Oncologist for three cycles of Doxorubicin and Endoxan. After completion of neoadjuvant chemotherapy, laparotomy was done, and portion of vaginal wall, right ovary, and rectus sheath with mass were removed. The patient is presently on depo provera and letrozol.

Discussion

ESS occurs primarily in the perimenopausal age group between 45 and 50 years of age. The age of presentation for this case was rather young. To date only a few cases of pregnancy in patients with ESS have been diagnosed. Adibah et al. [1] reported a case of ESS in a 35-year-old, para 5 lady who became pregnant while having the disease and delivered a healthy baby. Lei Yan et al. reported a case of successful pregnancy after conservative treatment of ESS in a 25-year-old woman. TAH is the treatment of choice in these patients, but conservative management may be attempted in selected patients desiring pregnancy [3]. Considering the indolent growth of low grade ESS, definitive surgery can be postponed until after completion of reproductive function.

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