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A Case Report of Low Grade Endometrial Stromal Sarcoma in a 14-Year-old Teenager

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Abstract

This article mainly reported a rare case: a 14-year-old teenager with irregular vaginal bleeding for 2 months was performed an operation and diagnosed as LGESS stage IIIA by pathology. The subsequent treatments were radiotherapy and letrozole treatment with closely followed up. Therefore, it is very important to pay attention to abnormal symptoms, not only in adults, but also in adolescents, for the early diagnosis of malignant tumors. The purpose of this case report is drawing attention to the diagnosis of this rare tumor in young patients.

Keywords: Endometrial Stromal Sarcoma; Surgical Therapy; Hormone Therapy; Case Report; Letrozole

Introduction

ESS is an infrequent and highly malignant endometrial carcinoma. It mainly included LGESS, high grade ESS and undifferentiated endometrial sarcoma. The onset age of ESS is usually between 40 and 55 of women. ESS possesses the characteristic of non-specific clinical symptoms, and the treatment of ESS is mainly surgery. However, the extremely high recurrence rate of ESS causing the poor prognosis of patients [1]. The patient in this case is only 14 years old and the FIGO stage of LGESS is IIIA. Because it is rare for such a young teenager to be diagnosed as advanced LGESS, the case was reported to draw attention to the diagnosis of this rare tumor in young patients.

Case Presentation

General Information

A 14-year-old teenager with no sexual history was hospitalized for irregular vaginal bleeding for 2 months. The patient had no family history of cancer or other medical history. The patient started menstruating at the age of 13 and had regular menstrual cycle. Since December 10, 2020, the patient began to experience persistent vaginal bleeding, which was slightly more intense than menstruation. The gynecological abdominal ultrasound in our hospital showed no obvious normal uterine shape and a mass measuring 130× 117×121 mm (Sarcoma?) in the pelvic cavity. MRI showed myometrium mass with the possibility of local malignant (Figure 1A, B). CT showed yometrium occupied with the possibility of uterine sarcoma, abdominal and pelvic effusion, nodules in both lungs, dilated hydronephrosis of right kidney and right ureter. Hemoglobin was 44g/L. The CA125 was 181.7U/ml. Preliminary diagnosis: abnormal uterine bleeding, pelvic mass? uterine malformation? severe anemia. Hemoglobin level rose to 96.0g/L after the hemostatic treatment and the transfusion of 6U red blood cells after hospitalization.

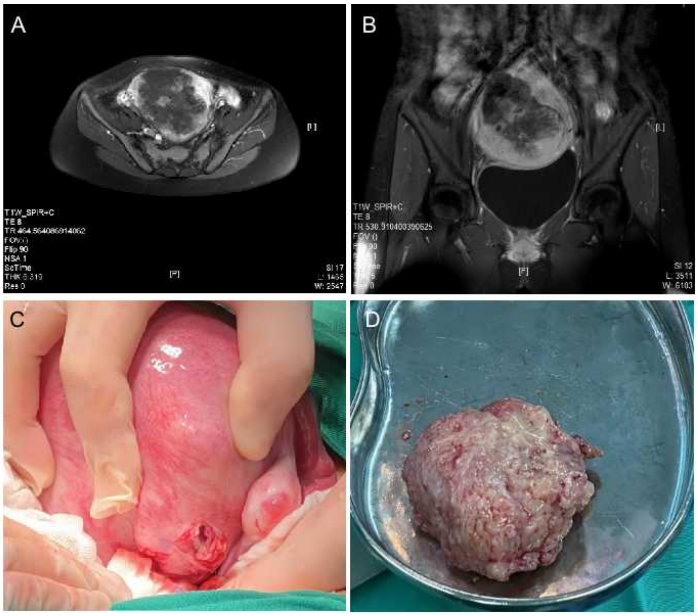

Figure 1: The radiographic and pathological images of the tumor. (A) The MRI of pelvic transverse section; (B) The MRI of pelvic coronal plane; (C) The tumor tissue penetrated the uterine serous layer; (D) The intact tumor tissue

Surgical Treatment

On February 20, 2021, intraoperative exploration exhibited that the uterus was enlarged to the size of more than 4 months of gestation. A mass about 9cm was found on the right anterior wall of the uterus and a local area about 3×2cm of the mass penetrated the serosal layer and adhered to the anterior abdominal wall. Another mass about 8cm was seen on the posterior wall of the uterus, and a local area about 2×1.5cm of the mass penetrated the serosal layer and adhered to the omentum. The tumor had invaded the lumen of the right oviduct, the right ovarian ligaments and ovarian vessels. One lymph node on the surface of the right external iliac artery was slightly enlarged (Fig-

ure 1C, D).

Finally, the uterus, right adnexa, left fallopian tube, right parauterine tissue, peritoneal lesions, and part of the omentum of the patient were removed by transabdominal and a biopsy of the right external iliac lymph node was also performed. The postoperative pathological was well-differentiated endometrial stromal sarcoma with low-grade malignant (Figure 2) and the tumor infiltrated the whole thickness of the uterine muscle wall with locally penetrating the serosal surface. The cervical canal, the fibromuscular wall of the cervix and the right parauterine tissue were involved, and the tumor thrombus was found in multiple vessels and the surrounding adipose connective tissue. The tumor tissues were also observed in the wall of the right fallopian tube, the surface tissue of the right ovary, the lesion tissue of the abdominal wall and the free blood clot. There was no metastasis in the right external iliac lymph node (0/1) and the removed omentum. On the basis of the postoperative pathological results, the patients were diagnosed as stage IIIA LGESS.

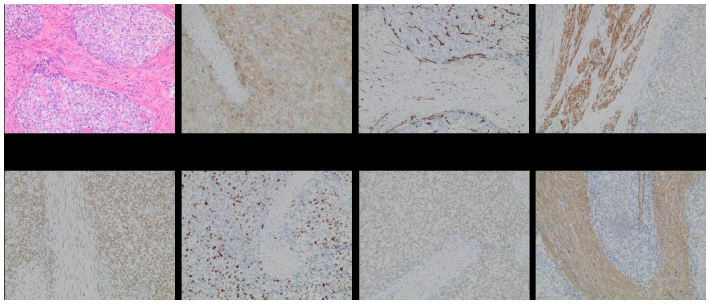

Figure 2: The HE staining and the immunohistochemistry of CD10, CD34, DES, ER, Ki67, PR and SMA of the tumor (400×)

Adjuvant Therapy

Oral megestrol 80mg Bid was administered on the 9th day after operation. Pelvic external irradiation (DT: 50-Gy/25F/ 25D) was administered from the 12th week after operation. Megestrel was changed to letrozole 2.5mg Qd on the 18th weeks after surgery. At the same time, chest CT was reexamined regularly after operation, and the size of pulmonary nodules did not change significantly, which meant the nodules may be benign rather than metastatic. In order to guide the postoperative medication, the patients were tested for tumor individualized medication gene, and the results showed that no gene mutations related to guiding targeted medication were detected (TMB:0.0mut/Mb level was low; PD-L1 immunohistochemistry: tumor cell staining ratio < 1%, PD-L1 protein expression was negative; Mismatch repair defect (dMMR) MLH1/MSH2/MSH6/PMS2 gene mutation (-); POLE/POLD1 gene mutation (-); BRCA2 gene mutation (-); MSI: MSI-L/MSS; TAP1/TAP2 gene mutation (-); B2M gene mutation (-); JAK1/JAK2 gene mutation (-); HLA-I heterozygosity: HLA-I heterozygous, etc.). Therefore, the patient was informed to continue oral letrozole treatment, and informed consent was obtained to use the case for medical research. The timeline of the case was exhibited in Figure 3. Now the patient is closely followed up.

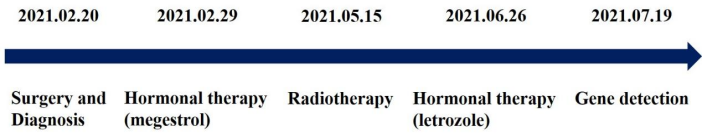

Figure 3: The timeline of the presented case

Discussion

LGESS is a rare gynecological malignancy with an incidence of less than 1% of all uterine malignancies. It is more common in women aged 40-55 years and rarely occurs in young women [1,2]. At present, literature review shows that only a few cases are reported for young women,

among which the lowest age in foreign countries is 16 years old [3], and the lowest age in China is 13 years old with only stage I [4]. This patient is 14 years old with stage IIIA LGESS, which is very rare. We wondered if this patient had t (7; 17) (p15; q21) balanced translocation or other genetic abnormalities because of the young age [5]. Therefore, the chromosome karyotype analysis was performed on the patient after surgery, and the result was 46XY without fusion gene, which excluded the cause of early onset of the patient, was caused by genetic factors.

LGESS is highly malignant, 25% of patients are asymptomatic. The clinical manifestations of LGESS are nonspecific; mainly lower abdominal and pelvic pain, abnormal vaginal bleeding, or progressive menorrhagia [6]. In this case, the patient experienced irregular vaginal bleeding for up to 2 months which resulted in severe anemia, but the symptoms such as lower abdominal pain and pelvic pain did not appear. LGESS has a high propensity for metastasis, not only through lymphatic vessels, but also through blood vessels. The metastatic sites of LGESS are mainly confined to the pelvis and lower reproductive tract, but distant metastasis to lungs may also occur after a few years [7]. In this case, the main metastatic sites were ovary, fallopian tube, parauterine tissue, peritoneum and omentum.

The current treatment for LGESS is total hysterectomy and bilateral salpingo-oophorectomy [8]. Simple tumor tissue resection had been proposed in the literature in order to preserve fertility especially in young women, however, most of these cases are stage I LGESS [9]. In this case, the tumor infiltrated the full thickness of the uterine muscle wall. In addition, the tumor broke through the serosal layer in the local uterus and transferred to the pelvis and abdomen. Therefore, fertility preserving surgery is out of the question. But one ovary was preserved for the patient during the operation, considering the intraoperative freezing result only suggested malignancy and the patient's young age. For lymph nodes, patients with positive lymph nodes had significantly worse survival than those with negative lymph nodes. A study had shown that ESS had a high lymph node involvement rate [10]. Therefore, the pathological results of lymph nodes provide clear prognostic information and treatment guidance. However, the potential value of lymph node dissection remains to be determined. But it is necessary to remove swollen or suspected metastatic lymph nodes during the operation [11]. In this case, only a swollen lymph node on the surface of the right external iliac artery was removed during the operation. Fortunately, the postoperative pathology of this lymph node was negative.

Hormonal therapy for LGESS include aromatase inhibitors, high-dose progesterone, and gonadotropin-releasing hormone analogues (GnRH-α). Progesterone is generally considered to be the first-line hormonal drug for treating LGESS [12]. In this case, megestrol was the first choice for postoperative hormone therapy. However, considering the side effects of long-term use of progesterone the result of the genetic test and the studies that the aromatase inhibitors have a longer survival time and fewer side effects for patients with LGESS than progesterone [13-14]. Therefore, the hormone therapy was changed from megestrol to letrozole at 18th weeks after surgery. Studies had shown that postoperative radiotherapy could reduce the local recurrence rate and was also suitable for patients with advanced or recurrent tumors [15]. For this patient with stage IIIA, pelvic radiation therapy was performed in the hope of prolonging disease-free survival and overall survival as much as possible.

Conclusion

In conclusion, we report a rare case of stage IIIA LGESS in a young teenager who underwent surgery and radiation therapy, as well as letrozole maintenance therapy. No case of advanced LGESS in such a young age has been reported, and the results of genetic testing did not suggest meaningful medication guidance. Up to now, the patient was followed up for nearly 30 months, and there was no sign of recurrence. Considering the high recurrence rate of advanced LGESS, we need to carry out continuous follow-up for this teenager in the future, and pay close attention to the changes of the patient's condition. In the course of treatment, the patient agreed with our treatment plan and would continue to follow up closely in the future.

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