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Case Report

Endometrial Stromal Sarcoma in Term Pregnancy: A Unique Case Report and Review of the Literature

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Abstract

Background: Endometrial stromal sarcoma (ESS)is a rare malignancy described only in a limited number of case reports. It accounts for only 2% of all uterine tumors.

Case Report: We describe a case of a 29-year-old pregnant lady presenting at 37 weeks of gestation in labor. A huge intra-cavitary mass was seen on bedside ultrasound upon her presentation. The patient underwent an urgent repeat cesarean section where a twenty cm uterine mass was identified after delivering the baby and the placenta. The pathology exam of the group showed low-grade endometrial stromal sarcoma with anti-CD10 and anti-estrogen receptors. A radical hysterectomy with bilateral salpingo-oophorectomy was performed two days later as the ESS was staged 1B following the post-cesarean PET scan result.

Conclusion: ESS is infrequent in young women as well as pregnant patients. This is the first case of asymptomatic endometrial stromal sarcoma in pregnancy, giving birth to a full-term baby with successful treatment.

Keywords: Stromal sarcoma, Pregnancy, Endometrial, Anti-CD 10 receptor, Anti-estrogen receptor

Introduction

Endometrial stromal sarcoma (ESS) has always been challenging. Only a few cases have been reported in the literature [1,2]. It is a remarkably rare malignancy encountered in 1-2 points/100.000 women [3-6]. This rare tumor of endometrial stromal origin accounts for 2 % of all uterine tumors [2,3]. It is the second most common uterine mesenchymal tumor, following leiomyosarcoma [7]. WHO has classified endometrial stromal tumors as benign (ESSN: endometrial stromal nodules) or malignant (ESS of low and high grade) according to myometrial invasion [1,3]. Risk factors or genetic associations are largely unknown [3,8]. Some authors have listed obesity and diabetes to be significantly related to ESS [3,9]. ESS appears to increase with age and is seen most frequently between 42 and 52 [5,6]. It typically presents with abnormal uterine bleeding [2]. A few cases have been disclosed concerning ESS in pregnancy and its presentation. Here, we describe a case of endometrial stromal sarcoma with myometrial invasion in pregnancy diagnosed at the time of delivery.

Case Presentation

A 29-year-old lady G6P4A2L5, non-smoker nor alcohol consumer, without a remarkable medical or family history but obesity with a BMI of 30 Kg/m2 and four previous cesarean sections, presented with labor pain at 37 weeks of gestation and six days. The patient was not followed at our institution. She mentioned that, during a routine ultrasound exam, she was told to have an incidental finding of a uterine mass suggestive of a fibroid.

Her past surgical history is significant for two prior hysteroscopic resections of benign uterine masses of small size: two years before this pregnancy. Upon presentation, a bedside ultrasound showed a viable single intrauterine pregnancy, commensurate with gestational age, and a vast uterine mass of about 20*8 cm in dimensions, solid component, and hypoechoic echotexture, yet photos are not available. She is completely asymptomatic. The patient was sent to the operating theater for a repeat cesarean section. Following the baby's delivery and the placenta, a 20*8 cm uterine mass was extruded from the uterine cavity (Figure 1).

Figure 1: Spontaneous extrusion of the uterine mass after placental delivery

It was heterogeneous, of irregular contour, soft in consistency, and attached to the left lateral uterine wall by a fibrous band around 3 cm in diameter. The mass was excised from its base and was sent for pathological studies (Figure 2).

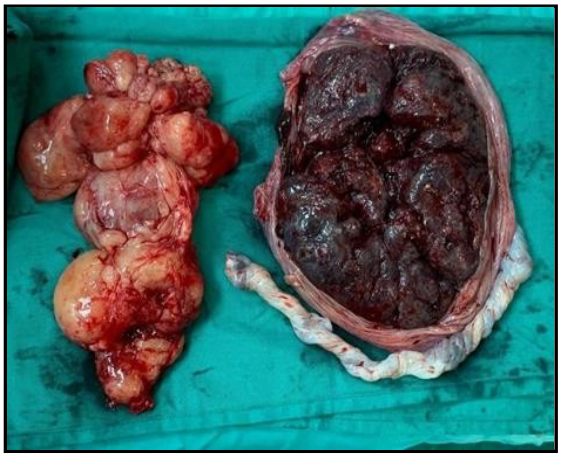

Figure 2: To the left: the 20*8 cm endometrial mass, heterogeneous with irregular borders of endometrial base found intra-operatively, to the right: The placenta.

The first pathology report stated possible endometrial sarcoma, which was then confirmed by immunohistochemistry as endometrial stromal sarcoma massively

infiltrating the myometrium with tumoral tissues strongly expressing anti-CD10 and anti-estrogen receptions, in turn confirming the diagnosis of low grade-ESS.

Postoperatively, a PET scan showed mild non-specific activity associated with the central aspect of the uterus without metastatic activity. ESS was thus staged IB according to FIGO classification for endometrial malignancies. Accordingly, the patient underwent a radical hysterectomy with bilateral salpingo-oophorectomy with an intraoperative finding of a 3*3 cm right simple ovarian cyst and no other gross lesion. The final pathology result showed endometrial stromal sarcoma massively infiltrating the myometrium yet not extending beyond the uterus.

Discussion

ESS is a tumor that affects mainly perimenopausal women between the ages of 45 and 55 [1-3,5]. High-grade ESS affects postmenopausal women while Low-grade ESS affects younger patients around 45 years old. Our case is among a few patients described in the literature about ESS affecting younger women. The symptomatology is not specific [4,11]. Affected women most frequently present with abnormal uterine bleeding, abdominal pain, and pressure due to the enlarging tumor. A small subset of patients does not exhibit symptoms as in our case. Diagnosis of ESS is often challenging [5]. Usually, the preoperative diagnosis of ESS is difficult and is misdiagnosed as uterine leiomyoma in 75% of cases [2,3]. Ultrasound findings are non-specific; a tissue biopsy is required to establish a definite diagnosis. According to specific histologic criteria such as cellular atypia, mitotic activity, and vascular invasion, ESS can be further divided into low and high-grade ESS types. [1,3]

Low-grade ESS carries a better prognosis than high grade-ESS as they are associated with slight nuclear atypia, minimal necrosis, less

Then 10 MF/10 HPF, and the presence of estrogen and progesterone receptors [1-3,6,10,11]. Whereas high-grade ESS is composed exclusively of high-grade round cells with uniform nuclear features but with a permeative pattern of infiltration or a cytomorphology featuring enlarged round to ovoid cells with smooth nuclear membranes and distinct chromatin clearing but lacking prominent nucleoli. It also exhibits diffuse moderate to strong nuclear cyclin D1 staining [1,3,10]. Our histopathology findings were convincing of low-grade ESS.

Low-grade ESS has an excellent prognosis with a 5-year survival rate of 90% for low-stage (I, II) and 50% for high-stage (III, IV) whereas that of high-grade ESS is only 32% [1,2]. Today, our patient is 33 years old. She is doing well and has regular follow-ups. The pathogenesis of ESS is not clear. Some risk factors have been identified such as pelvic radiation therapy, Long-term estrogen use, and obesity [3,8,9,11]. The latter is the only risk factor found in our patients. Only a few cases in the literature have been reported about ESS with concurrent pregnancy, but this is the first case of low-grade ESS with a pregnancy that yielded a delivery of a full-term baby with subsequent successful treatment of the sarcoma. Both cases described in the literature were associated with symptoms during pregnancy for which the patients sought medical advice, and with further workup, ESS was diagnosed [6,11]. Our case had an asymptomatic and smooth pregnancy course, a rare presentation in patients with ESS. Moreover, the two cases described in the literature were for pregnant patients presenting at 12 [11] and 15 weeks [6] of gestation, and these patients had a subsequent interruption of pregnancy to pursue treatment of sarcoma. Our case is the first in the literature to describe a pregnant woman delivering at term with incidental finding of ESS upon delivery. Surgery is the definitive management of ESS to establish both diagnosis and treatment [3,4]. Treatment for low-grade-ESS consists of total abdominal hysterectomy with bilateral salpingo-oophorectomy to decrease the likelihood of extension into the adjacent structures and eliminate the possible stimulating effect of

estrogen on the tumoral cells. Adjuvant treatment consists of radiotherapy and hormonotherapy [3-5]. The tumor metastasizes mainly through hematogenous spread with low lymph node metastasis rates, suggesting that lymph node dissection is not necessary at the time of operation [3,11].

Conclusion

We describe a case of Low-grade ESS diagnosed incidentally in a 29-year-old asymptomatic pregnant woman. ESS is a rare tumor and the preoperative diagnosis is usually a leiomyoma clinically and radiologically. The final diagnosis is obtained through biopsy. We would like to highlight the necessity for a high degree of suspicion of ESS even in young patients.

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