17th World Congress in Fetal Medicine

# Low grade endometrial stromal sarcoma during pregnancy

Matalliotakis M, Velegrakis A, Koliarakis I, Matalliotaki Ch, Zervou M, Giannikaki E, Kokkinakis Th, Niraki E
Obstetrics & Gynecology, Surgery and Histopathology Departments of Venizeleio General Hospital, Crete, Greece, Heraklion, Greece

## Objective

Endometrial stromal (ESS) sarcoma is more common in perimenopausal and postmenopausal women and accounts for less than 1% of all uterine tumors. Endometrial sarcomas are a rare finding during pregnancy since most of the cases are misdiagnosed as benign uterine leiomyomas. Previously, Matsuo et al reported 40 cases with genital sarcomas, with mean age at diagnosis at 27. 8 years and a five-year survival rate at 22%.

## Methods

A 33-year old nulliparous woman was admitted to our Unit at 20 weeks of gestation due to sudden severe right abdominal pain. In 2013, she had undergone laparoscopy for myomectomy. In 2015 she had total thyroidectomy in combination with radio-iodine therapy due to papillary thyroid microcarcinoma. First trimester screening test: low risk for trisomy. At the time of the admission, Ultrasound revealed an abnormal mass next to the right ovary which infiltrated the posterior uterine wall. MRI results: large infiltrating myometrial mass with irregular and ill-defined margins, on the right side of pelvis and localized lymphadenopathy. CA 125 was 41 U/ml.

## Results

On laparotomy we observed several necrotized masses in the peritoneal wall and multiple small solid masses extending to the omentum and intestine. Biopsy indicated Low grade ESS with direct metastases to the peritoneal cavity and omentum. The patient was referred to a specialized Gynecologic Oncology Center where decided to schedule a planned elective C-Section at 32 weeks of gestation followed by a radical hysterectomy, bilateral salpingo-oophorectomy and adjuvant chemotherapy.

## Conclusion

ESS is an aggressive tumor, which mostly metastasizes via the hematogenous route and rarely through lymphatics. Risk factors are conditions as unopposed estrogen treatment, exposure to tamoxifen, radiation and chromosomal translocations [t (7; 17) (p15; q21)]. Low grade ESS has a tendency for recurrence and is more common in pre-menopause patients. Histopathology shows fewer than 10 mitoses per 10 HPFs, and the cell nuclei appeared with mild atypia. Signs and symptoms are nonspecific: menorrhagia, abdominal distension and pain due to hemorrhage and tissue necrosis. Ultrasound imaging is not specific. On MRI, sarcomas show higher enhancement than normal myometrium and appear as a sudden overgrowth mass in the uterine cavity. The tumor margins are not regular, usually with marginal nodular lesions. Tissue biopsy identify the diagnosis. In general, the main treatment of ESS is total abdominal hysterectomy with bilateral salpingo-oophorectomy. In early disease stages, ovaries may be preserved to retain hormonal function. Conservative therapy includes chemotherapy, radiation and hormonal therapy; progestin therapy lowers the chance of reoccurrence. In case of diagnosis during pregnancy, the time of delivery or termination of pregnancy depends on the risk of the cancer progression, the uterine involvement and the risk of prematurity. Treatment during pregnancy is more challenging - chemotherapy can be administered after organogenesis, in the second or third trimester, and is rarely associated with stillbirth, fetal toxicity and intrauterine growth restriction. A multidisciplinary medical team is recommended to decide the management and the follow up treatment. More research is needed to evaluate the risk of cancerous cells to bypass the placenta barrier and further effects on the embryo. Moreover, scientists should further evaluate the recurrent gene fusions such as JAZF1/SUZ12 and/or EPC1-PHF1, which result in the creation of fusion transcripts which have anti-apoptotic activities and represent a specific diagnostic tool to differentiate LG and HG-ESS.