Uterine Sarcoma

Patient Information Series – What you should know, what you should ask.

What is uterine sarcoma?

Uterine sarcomas are rare malignant tumors arising from the uterus. Population-based estimates of uterine sarcoma incidence range from 1.5 to 2 per 100,000 women per year. The patients operated for a leiomyoma have an estimated risk to have a sarcoma of 0.1-0.3%.
Uterine sarcomas are classified as leiomyosarcomas, endometrial stromal sarcoma or undifferentiated endometrial sarcoma. Leiomyosarcoma has been reported to be the most common type of sarcoma (41-60%). Women presenting with uterine sarcomas are in median in their 5-6 decades of life.

What are the symptoms?

Women with uterine sarcoma often present with symptoms of abnormal vaginal bleeding either in the pre- or post-menopausal period, a palpable pelvic mass, or abdominal pain, while up to 25% will be asymptomatic. Signs and symptoms resemble those of benign leiomyomas, and preoperative distinction between the two on the basis of clinical information may be difficult. Malignancy should be suspected in case of tumor growth in post-menopausal women. Occasionally, the presenting symptoms are explained by tumor rupture, or the presence of metastases.

What are the risk factors?

- advanced age and postmenopausal status
- black women
- exposure to tamoxifen, unopposed estrogen and polycystic ovary syndrome
- previous pelvic radiation for the endometrial stromal sarcomas

Is a screening test available for uterine sarcoma?

A screening test is a test that is done when no symptoms are present. Currently, no screening method has been shown to significantly affect mortality in women with uterine sarcomas.

How is it diagnosed?

Diagnosis can difficult as both the clinical presentation and imaging findings may be overlapping with benign fibroids. Recent studies have shown that the sonographic presentation of uterine sarcomas have characteristics that are uncommon in benign lesions. Thus, the diagnostic work up starts with clinical examination and an expert ultrasound or MRI assessment to look at the tumor. A computed tomography scan (CT) of abdomen and pelvis may be offered to evaluate the presence of metastases. Tumor markers (proteins that are sometimes increased in women

with peritoneal involvement) such as for example Ca-125, LDH may be elevated, but the finding is non-specific and the clinical role is unclear, although it may have a roll in follow-up after treatment if initially elevated. In some case (intracavitary endometrial stromal sarcoma) a biopsy can confirm the diagnosis, while it may be contraindicated in others due to the risk of spreading the tumor.

What is the prognosis?

Leiomyosarcomas are very aggressive tumors with poor prognosis. Endometrial stromal sarcomas are indolent tumors with a favorable prognosis. Undifferentiated endometrial sarcomas have very poor prognosis, and most patients die of the disease within 2 years from diagnosis.

What are the treatment options?

The cornerstone in the treatment of uterine sarcomas is surgery; including hysterectomy , removal of the ovaries and fallopian tubes, and removing any tumor outside the uterus. Lymph node dissection is controversial. Young patients affected by early stage endometrial stromal sarcoma who wish to preserve reproductive function, remove only the lesion with sparing of uterus and ovaries, is an option.

In women with leiomyosarcomas additional radiation therapy does not seem to affect the prognosis or improve survival. The benefits of chemotherapy for patients with early stage sarcoma, are also controversial. Occasionally, hormone sensitive tumors will respond to hormonal treatment, for example progestin, aromatase inhibitor, Gonadotrophin Releasing Hormone (GnRH) analogues or GnRH analogues plus megestrol acetate.

Endometrial stromal sarcomas are usually hormone sensitive and progestins or aromatase inhibitors may be used as additional treatment for this disease.

Undifferentiated endometrial sarcomas: Treatment is primarily surgical with or without adjuvant radiotherapy or chemotherapy.

What is the follow up after treatment?

Women treated for uterine sarcomas need to have regular follow-up appointments. A checkup after cancer treatment usually includes a review of symptoms and a physical exam. The checkup also may include blood samples (to monitor CA 125 blood levels – or other tumors markers - if they were positive before surgery) and imaging tests. These may include ultrasound, chest X-ray, **magnetic resonance imaging,** or **computed tomography.** For the first couple of years follow-up appointments will be scheduled every 2–3 months. Afterwards the visits will then become less frequent, once or twice a year for up to 5 years (depending on Institutional policies).

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