



Uterine Leiomyosarcoma

WHAT SHOULD I KNOW?



BACKGROUND

The uterus has two major components that include a thick smooth muscle shell wrapped over a thin endometrial lining. Cancers can arise from either of these distinct components and be called “uterine cancer,” however greater than 90% of “uterine cancer” arises from the endometrial component leaving only a small fraction arising from the smooth muscle component. These rare smooth muscle cancers of the uterus are uterine sarcomas, which can be further classified based on the cellular appearances and genetic makeup of the cancer cells. The most common of these uterine sarcomas is the leiomyosarcoma, which accounts for 60% of all uterine sarcomas. Leiomyosarcomas are the malignant relative of the very common and benign uterine fibroid (leiomyoma), which exist in nearly 75% of all women – often without symptoms. Most leiomyosarcomas are diagnosed at an early stage while confined to the uterus, however they are aggressive tumors that often get into the blood stream leading to frequent recurrence. Due to the rarity of these tumor, it is difficult to study them or develop novel therapies and treatment strategies. There does not appear to be any established hereditary link to most leiomyosarcomas.

SIGNS & SYMPTOMS

Leiomyosarcomas are most commonly diagnosed in the perimenopausal years (age 45-60). Patients typically complain of abnormal uterine bleeding, pelvic pain, or pelvic pressure leading to an evaluation that identifies one or more uterine smooth muscle masses. These are often assumed to be benign fibroids and the diagnosis of leiomyosarcoma is often given after hysterectomy because preoperative biopsy is rarely pursued or indicated given the rarity of leiomyosarcoma. Less commonly a patient may have cough, shortness of breath, abdominal distention, and upper abdominal pain from tumors in the lungs, liver, and upper abdomen.

SCREENING

Currently there is no standardized screening for leiomyosarcoma. You should seek medical evaluation from a gynecologist if you are experiencing abnormal uterine bleeding, pelvic pain or pressure.

TREATMENT

Surgery: Hysterectomy and surgical excision of any metastatic leiomyosarcoma is the cornerstone of therapy.

Chemotherapy: Chemotherapy is typically recommended for stage II-IV leiomyosarcoma. There is uncertainty about the benefit of chemotherapy in stage I disease. Commonly used drugs include doxorubicin, gemcitabine, docetaxel, dacarbazine, and ifosfamide.

Radiation: In some cases pelvic radiation may be prescribed after initial diagnosis and hysterectomy with or without chemotherapy. Radiation is more commonly used to treat inoperable metastatic..

Targeted Therapy: Larotrectinib or entrectinib for NTRK gene fusion containing tumors

Immunotherapy – Pembrolizumab may be offered in tumors classified with high tumor mutational burden or other genetic markers suggesting potential benefit from immune therapy.

Hormonal Therapy: Some leiomyosarcomas overexpress estrogen or progesterone receptors and hormonal therapy could be an adjunct in these cases.

QUESTIONS YOU SHOULD ASK

Am I eligible for any clinical trials? What stage is my cancer? Is my doctor a gynecologic oncologist? Should I see a sarcoma specialist or get a second opinion? Can genetic testing be done on my tumor?



Society of Gynecologic Oncology

The Foundation for Women's Cancer (FWC) is a 501(c)3 nonprofit organization dedicated to supporting research, education and public awareness of gynecologic cancers.

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