



Soft Tissue Sarcomas

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What Is Soft Tissue Sarcoma?

Soft tissue sarcoma is a broad term for cancers that start in soft tissues (muscle, tendons, fat, lymph and blood vessels, and nerves). These cancers can develop anywhere in the body but are found mostly in the arms, legs, chest, and abdomen.

There are many types of soft tissue sarcoma. The cells of each type of sarcoma look different under a microscope, based on the type of soft tissue in which the cancer began:

- Childhood Soft Tissue Sarcoma
- Ewing Sarcoma
- Gastrointestinal Stromal Tumor (GIST)
- Kaposi Sarcoma
- Uterine Sarcoma

What Are the Risk Factors for Soft Tissue Sarcomas?

Anything that increases the chance of getting a disease is called a risk factor. Having a risk factor does not mean that you will get cancer; not having risk factors does not mean that you will not get cancer.

Risk factors for soft tissue sarcoma include the following inherited disorders:

- Retinoblastoma
- Neurofibromatosis type 1 (NF1; von Recklinghausen disease)
- Tuberous sclerosis (Bourneville disease)
- Familial adenomatous polyposis (FAP; Gardner syndrome)
- Li-Fraumeni syndrome
- Werner syndrome (adult progeria)
- Nevoid basal cell carcinoma syndrome (Gorlin syndrome)

Other risk factors for soft tissue sarcoma include the following:

- Past treatment with radiation therapy for certain cancers
- Being exposed to certain chemicals, such as Thorotrast (thorium dioxide), vinyl chloride, or arsenic
- Having swelling (lymphedema) in the arms or legs for a long time

What Are the Signs and Symptoms of Soft Tissue Sarcomas?

A sarcoma may appear as a painless lump under the skin, often on an arm or a leg. Sarcomas that begin in the abdomen may not cause signs or symptoms until they get very big. As the sarcoma grows bigger and presses on nearby organs, nerves, muscles, or blood vessels, signs and symptoms may include:

- Pain
- Trouble breathing

Other conditions may cause the same signs and symptoms.

What Tests Are Used to Detect (Find) and Diagnose Sarcomas?

If your doctor thinks you may have a soft tissue sarcoma, a biopsy will be done. The type of biopsy will be based on the size of the tumor and where it is in the body.

A pathologist views the tissue under a microscope to look for cancer cells and to find out the grade of the tumor. The grade of a tumor depends on how abnormal the cancer cells look under a microscope and how quickly the cells are dividing. High-grade tumors usually grow and spread more quickly than low-grade tumors.

What Is Staging?

The process used to find out if cancer has spread within the soft tissue or to other parts of the body is called staging. Staging of soft tissue sarcomas is also based on the grade and size of the tumor, and whether it has spread to the lymph nodes or other parts of the body. The information gathered from the staging process determines the stage of the disease. It is important to know the stage in order to plan treatment.

The following tests and procedures may be used:

- **Physical exam and history**
- **Chest x-ray:** An x-ray of the organs and bones inside the chest. An x-ray is a type of energy beam that can go through the body and onto film, making a picture of areas inside the body.
- **Blood chemistry studies:** A blood sample is checked to measure the amounts of certain substances released into the blood by organs and tissues in the body. An unusual (higher or lower than normal) amount of a substance can be a sign of disease.
- **Complete blood count (CBC):** A procedure in which a sample of blood is drawn and checked for the following:
 - The number of red blood cells, white blood cells, and platelets
 - The amount of hemoglobin (the protein that carries oxygen) in the red blood cells
 - The portion of the blood sample made up of red blood cells
- **CT scan (CAT scan):** A procedure that makes a series of detailed pictures of areas inside of the body, such as the lung and abdomen, taken from different angles. The pictures are made by a computer linked to an x-ray machine. A dye may be injected into a vein or swallowed to help the organs or tissues show up more clearly.

- **MRI (magnetic resonance imaging):** A procedure that uses a magnet, radio waves, and a computer to make a series of detailed pictures of areas inside the body.
- **PET scan (positron emission tomography scan):**
A procedure to find malignant tumor cells in the body. A small amount of radioactive glucose (sugar) is injected into a vein. The PET scanner rotates around the body and makes a picture of where glucose is being used in the body. Malignant tumor cells show up brighter in the picture because they are more active and take up more glucose than normal cells do.

The results of these tests are viewed together with the results of the tumor biopsy to find out the stage of the soft tissue sarcoma before treatment is given. Sometimes chemotherapy or radiation therapy is given as the initial treatment and afterwards the soft tissue sarcoma is staged again.

What Determines How Sarcomas Are Treated and Prognosis?

In addition to stage (how far the cancer has spread), the grade of the tumor is also important. You may hear the term mitotic rate which tells how quickly the cancer cells are dividing and growing.

The grade of the tumor describes how abnormal the cancer cells look under a microscope and how quickly the tumor is likely to grow and spread. Low grade, mid grade, and high grade are used to describe soft tissue sarcoma.

What Are the Stages Used For Soft Tissue Sarcoma?

There is no standard staging system for soft tissue sarcoma of the head, neck, chest, or abdomen. Other types of soft tissue sarcoma may be staged from I (1) to stage IV (4), with stage 4 being the most advanced.

The treatment options and prognosis (chance of recovery) depend on the following:

- The type of soft tissue sarcoma
- The size, grade, and stage of the tumor
- How fast the cancer cells are growing and dividing
- Where the tumor is in the body
- Whether all of the tumor is removed by surgery
- The patient's age and general health
- Whether the cancer has recurred (come back)

When cancer spreads to another part of the body, it is called metastasis. The metastatic tumor is the same type of cancer as the primary tumor. For example, if soft tissue sarcoma spreads to the lung, the cancer cells in the lung are actually soft tissue sarcoma cells. The disease is soft tissue sarcoma, not lung cancer.

How Is Sarcoma Treated?

Different types of treatments are available for patients with adult soft tissue sarcoma. Some treatments are standard (the currently used treatment), and some are being tested in clinical trials.

Surgery

Surgery is the most common treatment for adult soft tissue sarcoma. For some soft-tissue sarcomas, removal of the tumor in surgery may be the only treatment needed. The following surgical procedures may be used:

- **Mohs microsurgery:** A procedure in which the tumor is cut from the skin in thin layers. During surgery, the edges of the tumor and each layer of tumor removed are viewed through a microscope to check for cancer cells. Layers continue to be removed until no more cancer cells are seen. This type of surgery removes as little normal tissue as possible and is often used where appearance is important, such as on the skin.

- **Wide local excision:** Removal of the tumor along with some normal tissue around it. For tumors of the head, neck, abdomen, and trunk, as little normal tissue as possible is removed.
- **Limb-sparing surgery:** Removal of the tumor in an arm or leg without amputation, so the use and appearance of the limb is saved. Radiation therapy or chemotherapy may be given first to shrink the tumor. The tumor is then removed in a wide local excision. Tissue and bone that are removed may be replaced with a graft using tissue and bone taken from another part of the patient's body, or with an implant such as artificial bone.
- **Amputation:** Surgery to remove part or all of a limb or appendage, such as an arm or leg. Amputation is rarely used to treat soft tissue sarcoma of the arm or leg.
- **Lymphadenectomy:** A surgical procedure in which lymph nodes are removed and a sample of tissue is checked under a microscope for signs of cancer. This procedure is also called a lymph node dissection.

Radiation Therapy

Radiation therapy is a cancer treatment that uses high-energy x-rays or other types of radiation to kill cancer cells or keep them from growing.

Chemotherapy

Chemotherapy is a cancer treatment that uses drugs to stop the growth of cancer cells, either by killing the cells or by stopping them from dividing.

Targeted Therapy

Targeted therapy is a type of treatment that uses drugs or other substances to identify and attack specific cancer cells without harming normal cells.

Follow-Up Tests May Be Needed

Some of the tests that were done to diagnose the cancer or to find out the stage of the cancer may be repeated. Some tests will be repeated in order to see how well the treatment is working. Decisions about whether to continue, change, or stop treatment may be based on the results of these tests.

Support is available for coping with changes that may have happened because of cancer treatment. Your healthcare team can offer ideas as well as a plan of care for long-term follow-up.

Clinical Trials

Clinical trials are done to find out if new cancer treatments are safe and effective or better than the standard treatment.

People who take part in a clinical trial may receive:

- The standard drugs alone or
- The standard drugs plus the new treatment being studied

Many of today's standard treatments for cancer are based on earlier clinical trials.

Ask if there is a clinical trial right for you.

Taking part in a clinical trial helps improve the way cancer will be treated in the future. Even when clinical trials do not lead to effective new treatments, they often answer important questions and help move research forward.

Some clinical trials only include people who have not yet received treatment. Other trials test treatments for those whose cancer has not gotten better. There are also clinical trials that test new ways to stop cancer from coming back or reduce the side effects of cancer treatment.

To Learn More About Soft Tissue Sarcomas

American Cancer Society

<https://www.cancer.org/>

National Cancer Institute

<https://www.cancer.gov/>

National Comprehensive Cancer Network Guidelines for Patients

<https://www.nccn.org/patients/guidelines/cancers.aspx>

MedlinePlus

<https://medlineplus.gov/>

Common Questions

What does the pathology report say?

What is the stage of my cancer?

What are my goals for treatment?

What are my treatment choices?

What kind of support services are available for me about finances, emotions, spiritual questions, etc.?

My Health Care Team	Contact Information
Surgeon:	
Medical Oncologist:	
Radiation Oncologist:	
Primary Care Doctor:	
Navigator:	
Nurse:	
Registered Dietitian Nutritionist:	
Other:	
Other:	

Adapted from: PDQ Soft Tissue Sarcoma Treatment. Bethesda, MD: National Cancer Institute.

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