

Soft Tissue Sarcomas: Questions and Answers

Key Points

- Sarcomas are malignant (cancerous) tumors that develop in tissues which connect, support, or surround other structures and organs of the body (see Question 1).
- Soft tissue sarcomas can arise almost anywhere in the body, with most occurring in the extremities and in and around organs (see Question 2).
- The cause of most cases of soft tissue sarcoma is not known. However, exposure to radiation and certain chemicals, and some inherited diseases, are known risk factors for the development of soft tissue sarcomas (see Question 3).
- Surgery is the primary treatment for soft tissue sarcomas (see Question 7).
- People are encouraged to enroll in clinical trials (research studies) that explore new treatments (see Question 8).

1. What are soft tissue sarcomas?

Sarcomas are malignant (cancerous) tumors that develop in tissues which connect, support, or surround other structures and organs of the body. Muscles, tendons (bands of fiber that connect muscles to bones), fibrous tissues, fat, blood vessels, nerves, and synovial tissues are types of soft tissue. Soft tissue sarcomas are grouped together because they share certain microscopic characteristics, have similar symptoms, and are generally treated in similar ways (1). They are usually named for the type of tissue in which they begin.

Examples of soft tissue sarcomas and the type of tissue in which they begin include the following (2):

- **Fibrous tissue** (tissue that holds bones, muscles, and organs in place)—fibrosarcoma, malignant fibrous histiocytoma
- **Fatty tissue**—liposarcoma



- **Smooth muscle** (e.g., uterus)—leiomyosarcoma
- **Skeletal muscle**—rhabdomyosarcoma
- **Blood and lymph vessels**—epithelioid hemangioendothelioma, angiosarcoma, lymphangiosarcoma, Kaposi sarcoma
- **Perivascular tissue** (near or around blood vessels)—glomangiosarcoma, malignant hemangiopericytoma
- **Synovial tissue** (tissue that lines joints, tendon sheaths, and fluid-filled sacs between tendons and bones)—synovial sarcoma
- **Peripheral nerves**—malignant granular cell tumor, malignant peripheral nerve sheath tumor (also called malignant schwannoma or neurofibrosarcoma)
- **Mesenchymal cells** (cells that develop into connective tissue, blood vessels, and lymphatic tissue)—gastrointestinal stromal tumor (GIST), malignant mesenchymoma

Other types of soft tissue sarcomas include alveolar soft part sarcoma, epithelioid sarcoma, desmoplastic small cell tumor, and clear cell sarcoma. At this time, scientists do not know the types of tissue in which these sarcomas begin (2). Many sarcomas have specific chromosomal alterations, which are used to help classify the tumors (2).

Sarcomas that develop in the bone and cartilage (osteosarcoma, Ewing sarcoma, and chondrosarcoma) are not classified as soft tissue sarcomas and are not described in this resource. (More information about these types of cancer is available in the National Cancer Institute (NCI) fact sheet *Bone Cancer: Questions and Answers* at <http://www.cancer.gov/cancertopics/factsheet/Sites-Types/bone> on the Internet.)

2. **Where in the body are soft tissue sarcomas more likely to develop?**

Soft tissue sarcomas can arise almost anywhere in the body. About 43 percent occur in the extremities (e.g., arms, legs); 34 percent occur in and around the internal organs (e.g., uterus, heart); 10 percent occur in the trunk (e.g., chest, back); and 13 percent occur in other locations (2). In very rare cases, these tumors develop in the gastrointestinal tract. A small percentage of these are GISTs. Malignant GISTs occur most commonly in the stomach and small intestine.

3. **What are the possible causes of soft tissue sarcomas?**

Although most soft tissue sarcomas do not have a clearly defined cause, researchers have identified several factors that increase the likelihood of developing these tumors (2). External radiation therapy is the most well-established risk factor for soft tissue sarcomas. Patients treated with radiation therapy for cancers of the retina, breast, cervix, ovary, testes, or lymphatic system have a much higher chance of developing soft tissue sarcomas than the general population (1). The risk appears to be related to the dose of radiation. To limit this risk, radiation treatment for cancer is planned to ensure that the greatest amount of radiation is delivered to diseased tissue while surrounding healthy tissue is protected as much as possible.

Another risk factor for soft tissue sarcomas is exposure to certain chemicals in the workplace, including vinyl chloride, arsenic, herbicides such as phenoxyacetic acids, and wood preservatives that contain chlorophenols (2). Chronic lymphedema (a condition in which excess fluid collects in the tissue and causes swelling) following radiation to, or surgical removal of, lymph nodes is also a risk factor.

Certain inherited diseases are associated with an increased risk of developing soft tissue sarcomas. Studies have focused on genetic changes that may lead to the development of soft tissue sarcomas. For example, people with Li-Fraumeni syndrome (associated with alterations in the p53 tumor suppressor gene), von Recklinghausen disease (also called neurofibromatosis type 1 and associated with alterations in the NF1 gene), hereditary leiomyomatosis and renal cell cancer syndrome (with alterations in the FH gene), and hereditary retinoblastoma (with alterations in the RB1 gene) are at increased risk of developing soft tissue sarcomas.

Kaposi sarcoma is a soft tissue sarcoma that sometimes develops in people with human immunodeficiency virus (HIV) infection. The primary cause of Kaposi sarcoma is infection with Kaposi sarcoma-associated herpesvirus (KSHV), or human herpesvirus-8. However, people infected with KSHV, but not HIV, rarely develop Kaposi sarcoma (3).

4. How often do soft tissue sarcomas occur?

Soft tissue sarcomas are rare. About 9,500 new cases were diagnosed in the United States in 2006 (4), which is less than 1 percent of all new cancer cases. However, sarcomas occur more often in children and young adults. For example, soft tissue sarcomas account for about 7 percent of all childhood cancers. The most common soft tissue sarcomas are leiomyosarcoma, malignant fibrous histiocytoma, and liposarcoma. By site of origin, leiomyosarcoma is the most common sarcoma of the organs, while liposarcoma and malignant fibrous histiocytoma are the most common sarcomas of the extremities (5). Rhabdomyosarcoma is the most common soft tissue sarcoma in children.

5. What are the symptoms of soft tissue sarcomas?

Soft tissue sarcomas usually appear as a lump or mass, but they rarely cause pain, swelling, or other symptoms (1). A lump or mass might not be a sarcoma; it could be benign (noncancerous), a different type of cancer, or another problem. It is important to see a doctor about any physical change, such as a lump or mass, because only a doctor can make a diagnosis.

6. How are soft tissue sarcomas diagnosed?

The doctor performs a physical exam and may use the following procedures and tests to diagnose soft tissue sarcoma:

- **X-rays** create images of areas inside the body on film.

- **Computed tomography (CT)**, a procedure that uses special x-ray equipment to obtain cross-sectional pictures of the body, can determine whether a soft tissue tumor has metastasized (spread) to the lung or abdomen (1). CT scans, also called CAT scans, can also be helpful in determining the size of the tumor and whether the tumor can be accessed through surgery.
- **Magnetic resonance imaging (MRI)** uses a powerful magnet linked to a computer to create detailed pictures of areas inside the body. MRI scans can aid in diagnosis, particularly in helping to distinguish soft tissue sarcomas from benign tumors, as well as showing the extent of the tumor. MRIs are also used to monitor the patient after treatment to see if the tumor has recurred (come back) (1).
- A **biopsy** is the removal of cells or tissue for examination by a pathologist. The pathologist studies tissue samples under a microscope or performs other tests on the cells or tissue. A biopsy is the only sure way to tell whether a person has cancer.

Specialized testing of the tumor cells for chromosomal alterations may also be conducted to aid in diagnosis (5).

7. **How are soft tissue sarcomas treated?**

Treatment for soft tissue sarcomas is determined mainly by the stage of the disease. The stage depends on the size of the tumor, the grade, and whether the cancer has spread to the lymph nodes or other parts of the body. The most important component of the stage is the tumor grade (how abnormal the cancer cells look under a microscope and how quickly the tumor is likely to grow and spread). Treatment options for soft tissue sarcomas include surgery, radiation therapy, and chemotherapy. A multidisciplinary team of cancer specialists can help plan the best treatment for patients with soft tissue sarcomas.

- **Surgery** is the usual treatment for soft tissue sarcomas. For surgery to be effective, the surgeon must remove the entire tumor with negative margins (no cancer cells are found at the edge or border of the tissue removed during surgery). The surgeon may use special surgical techniques to minimize the amount of healthy tissue removed with the tumor. Some patients need reconstructive surgery.
- **Radiation therapy**, also called radiotherapy, involves the use of high-energy x-rays to kill cancer cells. This therapy may be used before surgery to shrink the tumor, after surgery to kill any cancer cells that may remain in the body, or both before and after surgery. Radiation may come from a machine outside the body (external radiation therapy). It can also come from radioactive materials placed directly into or near the area where the cancer cells are found (internal radiation therapy or radiation implant) (1).

- **Chemotherapy** is the use of anticancer drugs to kill cancer cells. Chemotherapy may be used before or after surgery, and with or without radiation therapy. The effectiveness of current anticancer drugs depends on the type of sarcoma. Some sarcomas are very responsive to chemotherapy, while others do not respond to current anticancer drugs (1). Some sarcomas with specific chromosomal alterations can be treated with therapies targeted to the alteration. For example, imatinib mesylate (Gleevec®) is a targeted therapy used to treat GIST that has metastasized (1).

8. Are clinical trials (research studies) available? Where can people get more information about clinical trials?

Yes. Participation in clinical trials is an important treatment option for many people with soft tissue sarcomas. To develop new treatments, and better ways to use current treatments, the NCI, a component of the National Institutes of Health, is sponsoring clinical trials in many hospitals and cancer centers around the country. Clinical trials are a critical step in the development of new methods of treatment. Before any new treatment can be recommended for general use, doctors conduct clinical trials to find out whether the treatment is safe for patients and effective against the disease.

People interested in taking part in a clinical trial should talk with their doctor. Information about clinical trials is available from the NCI's Cancer Information Service (CIS) (see below) at 1-800-4-CANCER and in the NCI booklet *Taking Part in Cancer Treatment Research Studies*, which can be found at <http://www.cancer.gov/publications> on the Internet. This booklet describes how research studies are carried out and explains their possible benefits and risks. Further information about clinical trials is available at <http://www.cancer.gov/clinicaltrials> on the NCI's Web site. The Web site offers detailed information about specific ongoing studies by linking to PDQ®, the NCI's comprehensive cancer information database. The CIS also provides information from PDQ.

Selected References

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2. Brennan M, Singer S, Maki R, O'Sullivan B. Sarcomas of the soft tissue and bone. In: DeVita VT Jr., Hellman S, Rosenberg SA, editors. *Cancer: Principles and Practice of Oncology*. Vol. 2. 7th ed. Philadelphia: Lippincott Williams and Wilkins, 2004.
3. Yarchoan R, Little R. Immunosuppression-related malignancies. In: DeVita VT Jr., Hellman S, Rosenberg SA, editors. *Cancer: Principles and Practice of Oncology*. Vol. 2. 7th ed. Philadelphia: Lippincott Williams and Wilkins, 2004.
4. Ries LAG, Harkins D, Krapcho M, et al. *SEER Cancer Statistics Review, 1975-2003*. Bethesda, MD: National Cancer Institute, 2006.

5. Brennan MF. Soft tissue sarcoma: Advances in understanding and management. *Surgeon* 2005; 3(3):216–223.

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Related NCI materials and Web pages:

- National Cancer Institute Fact Sheet 5.9, *Tumor Grade: Questions and Answers* (<http://www.cancer.gov/cancertopics/factsheet/Detection/tumor-grade>)
- National Cancer Institute Fact Sheet 5.32, *Staging: Questions and Answers* (<http://www.cancer.gov/cancertopics/factsheet/Detection/staging>)
- National Cancer Institute Fact Sheet 6.1, *Synovial Sarcoma: Questions and Answers* (<http://www.cancer.gov/cancertopics/factsheet/Sites-Types/synovial>)
- National Cancer Institute Fact Sheet 6.20, *Metastatic Cancer: Questions and Answers* (<http://www.cancer.gov/cancertopics/factsheet/Sites-Types/metastatic>)
- National Cancer Institute Fact Sheet 6.26, *Bone Cancer: Questions and Answers* (<http://www.cancer.gov/cancertopics/factsheet/Sites-Types/bone>)
- National Cancer Institute Fact Sheet 7.49, *Targeted Cancer Therapies: Questions and Answers* (<http://www.cancer.gov/cancertopics/factsheet/Therapy/targeted>)
- *Chemotherapy and You: Support for People With Cancer* (<http://www.cancer.gov/cancertopics/chemotherapy-and-you>)
- *Radiation Therapy and You: Support for People With Cancer* (<http://www.cancer.gov/cancertopics/radiation-therapy-and-you>)
- *Taking Part in Cancer Treatment Research Studies* (<http://www.cancer.gov/clinicaltrials/Taking-Part-in-Cancer-Treatment-Research-Studies>)

For more help, contact:

NCI's Cancer Information Service

Telephone (toll-free): 1-800-4-CANCER (1-800-422-6237)

TTY (toll-free): 1-800-332-8615

LiveHelp[®] online chat: <https://cissecure.nci.nih.gov/livehelp/welcome.asp>

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