CANCER FACTS

National Cancer Institute • National Institutes of Health
Department of Health and Human Services

Soft Tissue Sarcomas: Questions and Answers

1. What is soft tissue?

The term *soft tissue* refers to tissues that connect, support, or surround other structures and organs of the body. Soft tissue includes muscles, tendons (bands of fiber that connect muscles to bones), fibrous tissues, fat, blood vessels, nerves, and synovial tissues (tissues around joints).

2. What are soft tissue sarcomas?

Malignant (cancerous) tumors that develop in soft tissue are called *sarcomas*, a term that comes from a Greek word meaning "fleshy growth." There are many different kinds of soft tissue sarcomas. They are grouped together because they share certain microscopic characteristics, produce similar symptoms, and are generally treated in similar ways. (Bone tumors [osteosarcomas] are also called sarcomas, but are in a separate category because they have different clinical and microscopic characteristics and are treated differently.)

Sarcomas can invade surrounding tissue and can metastasize (spread) to other organs of the body, forming secondary tumors. The cells of secondary tumors are similar to those of the primary (original) cancer. Secondary tumors are referred to as "metastatic soft tissue sarcoma" because they are part of the same cancer and are not a new disease.

Some tumors of the soft tissue are benign (noncancerous). These tumors do not spread and are rarely life-threatening. However, benign tumors can crowd nearby organs and cause symptoms or interfere with normal body functions.

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

Scientists do not fully understand why some people develop sarcomas while the vast majority do not. However, by identifying common characteristics in groups with unusually high occurrence rates, researchers have been able to single out some factors that may play a role in causing soft tissue sarcomas.

Studies suggest that workers who are exposed to phenoxyacetic acid in herbicides and chlorophenols in wood preservatives may have an increased risk of developing soft tissue sarcomas. An unusual percentage of patients with a rare blood vessel tumor, angiosarcoma of the liver, have been exposed to vinyl chloride in their work. This substance is used in the manufacture of certain plastics.

In the early 1900s, when scientists were just discovering the potential uses of radiation to treat disease, little was known about safe dosage levels and precise methods of delivery. At that time, radiation was used to treat a variety of noncancerous medical problems, including enlargement of the tonsils, adenoids, and thymus gland. Later, researchers found that high doses of radiation caused soft tissue sarcomas in some patients. Because of this risk, radiation treatment for cancer is now planned to ensure that the maximum dosage of radiation is delivered to diseased tissue while surrounding healthy tissue is protected as much as possible.

Researchers believe that a retrovirus plays an indirect role in the development of Kaposi's sarcoma, a rare cancer of the cells that line blood vessels in the skin and mucus membranes. Kaposi's sarcoma often occurs in patients with AIDS (acquired immune deficiency syndrome). AIDS-related Kaposi's sarcoma, however, has different characteristics and is treated differently than typical soft tissue sarcomas.

Studies have focused on genetic alterations that may lead to the development of soft tissue sarcomas. Scientists have also found a small number of families in which more than one member in the same generation has developed sarcoma. There have also been reports of a few families in which relatives of children with sarcoma have developed other forms of cancer at an unusually high rate. Sarcomas in these family clusters, which represent a very small fraction of all cases, may be related to a rare inherited genetic alteration.

Certain inherited diseases are associated with an increased risk of developing soft tissue sarcomas. For example, people with Li-Fraumeni syndrome (associated with alterations in the p53 gene) or von Recklinghausen's disease (also called neurofibromatosis, and associated with alterations in the NF1 gene) are at an increased risk of developing soft tissue sarcomas.

4. Where do soft tissue sarcomas develop?

Soft tissue sarcomas can arise almost anywhere in the body. About 50 percent occur in the extremities (the arms, legs, hands, or feet), 40 percent occur in the trunk (chest, back, hips, shoulders, and abdomen), and 10 percent occur in the head and neck.

The tables on pages 6 and 7 list several types of sarcomas that occur in adults and children.

5. How often do soft tissue sarcomas occur?

Soft tissue sarcomas are relatively uncommon cancers. They account for less than 1 percent of all new cancer cases each year. In 2000, there will be an estimated 8,100 new cases of soft tissue sarcoma in the United States. Approximately 850 to 900 of these cases will occur among children and adolescents under age 20.

6. What are the symptoms of soft tissue sarcomas?

In their early stages, soft tissue sarcomas usually do not cause symptoms. Because soft tissue is relatively elastic, tumors can grow rather large, pushing aside normal tissue, before they are felt or cause any problems. The first noticeable symptom is usually a painless lump or swelling. As the tumor grows, it may cause other symptoms, such as pain or soreness, as it presses against nearby nerves and muscles.

7. How are soft tissue sarcomas diagnosed?

The only reliable way to determine whether a soft tissue tumor is benign or malignant is through a surgical biopsy. Therefore, all soft tissue lumps that persist or grow should be biopsied. During this procedure, a doctor makes an incision or uses a special needle to remove a sample of tumor tissue. A pathologist examines the tissue under a microscope. If cancer is present, the pathologist can usually determine the type of cancer and its grade. The grade of the tumor is determined by how abnormal the cancer cells appear when examined under a microscope. The grade predicts the probable growth rate of the tumor and its tendency to spread. Low-grade sarcomas, although cancerous, are unlikely to metastasize. High-grade sarcomas are more likely to spread to other parts of the body.

8. How are soft tissue sarcomas treated?

In general, treatment for soft tissue sarcomas depends on the stage of the cancer. The stage of the sarcoma is based on the size and grade of the tumor, and whether the cancer has spread to the lymph nodes or other parts of the body (metastasized). Treatment options for soft tissue sarcomas include surgery, radiation therapy, and chemotherapy.

- **Surgery** is the most common treatment for soft tissue sarcomas. If possible, the doctor may remove the cancer and a safe margin of the healthy tissue around it. Depending on the size and location of the sarcoma, it may occasionally be necessary to remove all or part of an arm or leg (amputation). However, the need for amputation rarely arises; no more than 10 percent to 15 percent of individuals with sarcoma undergo amputation. In most cases, limb-sparing surgery is an option to avoid amputating the arm or leg. In limb-sparing surgery, as much of the tumor is removed as possible, and radiation therapy and/or chemotherapy are given either before the surgery to shrink the tumor or after surgery to kill the remaining cancer cells.

- **Radiation therapy** (treatment with high-dose x-rays) may be used either before surgery to shrink tumors or after surgery to kill any cancer cells that may have been left behind.

- **Chemotherapy** (treatment with anticancer drugs) may be used with radiation therapy either before or after surgery to try to shrink the tumor or kill any remaining cancer cells. If the cancer has spread to other areas of the body, chemotherapy may be used to shrink tumors and reduce the pain and discomfort they cause, but is unlikely to eradicate the disease. The use of chemotherapy to prevent the spread of soft tissue sarcomas has not been proven to be effective. Patients with soft tissue sarcomas usually receive chemotherapy intravenously (injected into a blood vessel).

Doctors are conducting clinical trials in the hope of finding new, more effective treatments for soft tissue sarcomas, and better ways to use current treatments. Clinical trials are in progress at hospitals and cancer centers around the country. Clinical trials are an important part of the development of new methods of treatment. Before a 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.

Patients who are interested in learning more about participating in clinical trials can call NCI's Cancer Information Service (see below) or access NCI's Cancer.gov Web site at http://cancer.gov/clinical_trials on the Internet.

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Sources of National Cancer Institute Information

Cancer Information Service

Toll-free: 1–800–4–CANCER (1–800–422–6237)
TTY (for deaf and hard of hearing callers): 1–800–332–8615

NCI Online

Internet

Use http://cancer.gov to reach the NCI's Web site.

LiveHelp

Cancer Information Specialists offer online assistance through the *LiveHelp* link on the NCI's Web site.

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Table 1
Major Types of Soft Tissue Sarcomas in Adults

Tissue of Origin	Type of Cancer	Usual Location in the Body
Fibrous tissue	Fibrosarcoma	Arms, legs, trunk
	Malignant fibrous hystiocytoma	Legs
Fat	Liposarcoma	Arms, legs, trunk
Muscle		
Striated muscle	Rhabdomyosarcoma	Arms, legs
Smooth muscle	Leiomyosarcoma	Uterus, digestive tract
Blood vessels	Hemangiosarcoma	Arms, legs, trunk
	Kaposi's sarcoma	Legs, trunk
Lymph vessels	Lymphangiosarcoma	Arms
Synovial tissue (linings of joint cavities, tendon sheaths)	Synovial sarcoma	Legs
Peripheral nerves	Neurofibrosarcoma	Arms, legs, trunk
Cartilage and bone-forming tissue	Extraskeletal chondrosarcoma	Legs
	Extraskeletal osteosarcoma	Legs, trunk (not involving the bone)

Table 2
Major Types of Soft Tissue Sarcomas in Children

Tissue of Origin	Type of Cancer	Usual Location in the Body	Most common ages
Muscle			
Striated muscle	Rhabdomyosarcoma		
	Embryonal	Head and neck, genitourinary tract	Infant–4
	Alveolar	Arms, legs, head, and neck	Infant–19
Smooth muscle	Leiomyosarcoma	Trunk	15–19
Fibrous tissue	Fibrosarcoma	Arms and legs	15–19
	Malignant fibrous histiocytoma	Legs	15–19
	Dermatofibrosarcoma	Trunk	15–19
Fat	Liposarcoma	Arms and Legs	15–19
Blood vessels	Infantile hemangio-pericytoma	Arms, legs, trunk, head, and neck	Infant–4
Synovial tissue (linings of joint cavities, tendon sheaths)	Synovial sarcoma	Legs, arms, and trunk	15–19
Peripheral Nerves	Malignant peripheral nerve sheath tumors (also called neurofibrosarcomas, malignant schwannomas, and neurogenic sarcomas)	Arms, legs, and trunk	15–19
Muscular nerves	Alveolar soft part sarcoma	Arms and legs	Infant–19
Cartilage and bone-forming tissue	Extraskeletal myxoid chondrosarcoma	Legs	10–14
	Extraskeletal mesenchymal	Legs	10–14