Soft Tissue Sarcoma Causes, Risk Factors, and Prevention

Risk Factors

A risk factor is anything that affects your chance of getting a disease such as cancer. Learn more about the risk factors for soft tissue sarcoma.

- Risk Factors for Soft Tissue Sarcomas
- What Causes Soft Tissue Sarcomas?

Prevention

The only way to prevent some soft tissue sarcomas is to avoid exposure to risk factors whenever possible. Still, most sarcomas develop in people with no known risk factors. At this time, there's no known way to prevent this cancer. And for people getting radiation therapy, there's usually little choice.

Risk Factors for Soft Tissue Sarcomas

A risk factor is anything that changes your chance of getting a disease like cancer. Different cancers have different risk factors. For example, unprotected exposure to strong sunlight is a risk factor for skin cancer. Smoking is a risk factor for cancers of the lung, and many other cancers. But risk factors don't tell us everything. Having a risk factor, or even many, doesn't mean that you will get the cancer. Also, many people get cancer without having a risk factor.
Injury and lifestyle factors such as smoking, diet, and exercise are NOT linked to the risk for soft tissue sarcoma. But the injury issue has caused some confusion in the past. One reason is that an injured area might swell. That swelling could look like a tumor, but it isn't one. Also, when you are injured, the pain may draw your attention to the injured area. A doctor may check the area, and x-rays or other imaging studies may be done. This can make it more likely that any sarcoma that's there will be discovered, even though it may have been there for some time.

Still, scientists have found a few risk factors that make a person more likely to develop soft tissue sarcomas:

**Radiation given to treat other cancers**

Radiation exposure accounts for less than 5% of sarcomas. But patients might develop sarcomas from radiation given to treat other cancers, like breast cancer or lymphoma. The sarcoma often starts in the part of the body that was treated with radiation. The average time between the radiation treatments and the diagnosis of a sarcoma is about 10 years.

Radiation therapy techniques have improved steadily over several decades. Treatments now target cancers more precisely, and more is known about selecting radiation doses. These advances are expected to reduce the number of cancers caused by radiation therapy. But because these cancers take so long to develop, the results of these changes may not be seen for a long time. Still, radiation therapy is used only when its benefits (improved survival rate and relief of symptoms) outweigh its risks. To learn more, see Second Cancers in Adults.

**Family cancer syndromes**

Family cancer syndromes are disorders caused by gene defects (mutations) that people are born with (often inherited from a parent) that are linked to a high risk of getting certain cancers. Some family cancer syndromes increase a person's risk of developing soft tissue sarcomas.

**Neurofibromatosis**

Neurofibromatosis is also known as von Recklinghausen disease. It usually runs in families and causes many benign (not cancer) tumors that form in nerves under the skin and in other parts of the body (These are called neurofibromas.) It’s caused by a defect (mutation) in genes called NF1 and NF2. About 5% of people with
neurofibromatosis will develop a sarcoma in a neurofibroma.

**Gardner syndrome**

Gardner syndrome is a disease caused by defects in the *APC* gene. This syndrome is a type of familial adenomatous polyposis (FAP), and people with it get many polyps in the colon (and intestines) and have a high risk of getting colon cancer. It also causes problems outside the colon, including desmoid tumors. (These are discussed in *What Is a Soft Tissue Sarcoma?*²)

**Li-Fraumeni syndrome**

Li-Fraumeni syndrome is caused by inherited defects in the *TP53* gene. People affected by this syndrome have a high risk of cancer, such as breast cancer³, brain tumors⁴, leukemia⁶, and sarcomas. Still, only 10 to 20 out of 100 people with Li-Fraumeni syndrome will develop a soft tissue sarcoma. People with this syndrome are sensitive to the cancer-causing effects of radiation. So if they have a cancer that’s treated with radiation, they have a very high chance of developing a new cancer in the part of the body that was treated.

**Retinoblastoma**

*Retinoblastoma*⁶ is an eye cancer in children that can be caused by defects in the *RB1* gene. Children with this gene defect also have a higher risk of developing bone or soft tissue sarcomas, especially if the retinoblastoma was treated with radiation.

**Werner syndrome**

Werner syndrome is caused by defects in the *RECQL2* gene. Children with this syndrome have problems like those seen in the elderly. These include cataracts, skin changes, and clogged heart arteries (arteriosclerosis) which can lead to heart attacks. They also have an increased risk of cancer, including soft tissue sarcomas.

**Gorlin syndrome**

Gorlin syndrome is also called *nevoid basal cell carcinoma syndrome* (NBCCS). It's caused by defects in the *PTCH1* gene. People with this syndrome have a high risk of developing many basal cell skin cancers⁷. They also have an increased risk of fibrosarcoma and rhabdomyosarcoma⁸.
Tuberous sclerosis

Tuberous sclerosis can be caused by a defect in the TSC1 and/or TSC2 gene. People with this syndrome often have seizures and learning problems. They get benign (not cancer) tumors in many different organs. They also have kidney problems, often along with a kidney tumor called angiomyolipoma. People with tuberous sclerosis have an increased risk of rhabdomyosarcoma.

Damaged lymph system

Lymph is a clear fluid containing immune system cells that’s carried throughout the body by a series of lymph vessels. These vessels connect lymph nodes (small bean-shaped collections of immune system cells). When lymph nodes have been removed or damaged by radiation therapy, lymph fluid can build up and cause swelling. This is called lymphedema.

Lymphangiosarcoma (a malignant (cancer) tumor that develops in lymph vessels) is a very rare complication of chronic lymphedema.

Chemicals

Exposure to vinyl chloride (a chemical used in making plastics) is a risk factor for developing sarcomas of the liver, but it hasn’t been proven to cause soft tissue sarcomas. Arsenic has also been linked to a type of liver sarcoma but not soft tissue sarcoma. Exposure to dioxin and to herbicides that contain phenoxyacetic acid at high doses (such as might occur in people who work on farms) may also be risk factors, but this isn’t known for certain. There’s no evidence that herbicides (weed killers) or insecticides, at levels encountered by the general public, cause sarcomas.

Hyperlinks


References


What Causes Soft Tissue Sarcomas?

Scientists don't know exactly what causes most soft tissue sarcomas, but they have found some risk factors that can make a person more likely to develop these cancers. And research has shown that some of these risk factors affect the genes in cells in the soft tissues.

Researchers have made great progress in understanding how certain changes in DNA (pieces of genes) can cause normal cells to become cancer. DNA carries the instructions for nearly everything our cells do. We usually look like our parents because they are the source of our DNA. But DNA affects more than just the way we look.

The DNA is made of genes. Genes carry the recipes for making proteins, the molecules that control all cell functions. Some genes contain instructions for proteins that control when our cells grow and divide.

- Certain genes that promote cell division are called oncogenes.
- Others that slow down cell division or cause cells to die at the right time are called tumor suppressor genes.

Cancers can be caused by DNA mutations (defects) that turn on oncogenes or turn off tumor suppressor genes.

Many family cancer syndromes have been found in which inherited DNA mutations cause a very high risk of developing breast, colon, kidney, eye, or other cancers. Some of these syndromes are also linked to an increased risk of developing soft tissue sarcomas. (These syndromes were noted in Risk Factors for Soft Tissue Sarcomas\(^1\)) The syndromes are caused by defects (mutations) in genes that can be inherited (passed on) from a parent. Some of these gene defects can be found through testing. For more on this, see Family Cancer Syndromes\(^2\) and Genetics and Cancer\(^3\).
DNA mutations in soft tissue sarcoma are common. But they're usually acquired during life rather than having been inherited before birth. Acquired mutations may result from exposure to radiation or cancer-causing chemicals. In most sarcomas, they occur for no apparent reason.

Researchers still don't know why most soft tissue sarcomas develop in people who have no apparent risk factors.

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**Can Soft Tissue Sarcomas Be Prevented?**

The only way to prevent some soft tissue sarcomas is to avoid exposure to risk factors whenever possible. But most sarcomas develop in people with no known risk factors, so, at this time, there's no known way to prevent most cases. And for people getting radiation therapy, there's usually little choice.

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