CONDITION

Solitary fibrous tumor

A solitary fibrous tumor is a rare growth of connective-tissue cells that can develop almost anywhere in the body.

WHAT IT IS
Rare tumor arising from connective-tissue cells

BODY SYSTEM
Connective tissues anywhere in the body

MAIN SYMPTOMS
Painless lump, pressure pain, cough, shortness of breath

WHAT CAUSES IT
Acquired genetic change; trigger unknown

HOW IT IS DIAGNOSED
Imaging and biopsy with STAT6 testing

MAIN TREATMENT
Surgery, sometimes with radiation or systemic treatment

CAN IT BE CURED?
Sometimes - complete removal can be curative

IS IT AN EMERGENCY?
Sometimes - severe breathing trouble, seizure, or fainting

LONG-TERM OUTLOOK
Often favorable, but late recurrence can occur

Overview

A solitary fibrous tumor, or SFT, is a rare growth that starts in cells supporting and connecting the body's tissues. Despite the name, "solitary" does not describe a particular body part. These tumors can develop almost anywhere.

SFTs were first recognized in the pleura, the thin lining around the lungs. They can also occur in the abdomen, pelvis, arms, legs, head, neck, or tissues covering the brain and spinal cord.

Many SFTs grow slowly and remain in one place. Others can return after treatment or spread to another part of the body. A tumor's behavior cannot always be predicted from its appearance alone. For this reason, doctors may avoid simply calling an SFT "benign," even when it appears to have a low risk of spreading.

SFT is different from common cancers of organs such as the lung, bowel, or brain. Care is often coordinated by a team experienced in sarcomas, which are tumors of connective and supporting tissues.

Symptoms

A small SFT may cause no symptoms. It may be found during a scan performed for another reason. Symptoms usually develop when the tumor becomes large enough to press on nearby organs, nerves, blood vessels, or muscles.

Possible symptoms include:

Rarely, an SFT produces an abnormal form of a hormone-like protein called insulin-like growth factor 2. This can lower blood sugar, a complication called Doege-Potter syndrome. Symptoms may include sweating, shaking, hunger, confusion, fainting, or seizures. These symptoms need prompt medical assessment.

Symptoms do not show whether an SFT is low or high risk. A small tumor can occur in a sensitive location, while a larger tumor may cause few problems if it has room to grow.

- A painless or slowly enlarging lump
- Pain, pressure, fullness, or swelling near the tumor
- Cough, chest discomfort, or shortness of breath from a chest tumor
- Abdominal swelling, constipation, or feeling full quickly
- Headache, vision changes, weakness, or seizures from a tumor near the brain
- Numbness, weakness, or reduced movement if nerves are compressed

Causes and risk factors

SFT develops because of changes within the tumor cells. Most contain a change that joins parts of two genes, called NAB2 and STAT6 . This gene fusion affects how the cells control growth.

This is usually an acquired change found only in the tumor. It is not normally inherited from a parent or passed to children. Having an SFT does not usually mean that relatives face an increased risk.

The event that causes the genetic change is unknown. No specific lifestyle, food, injury, infection, or environmental exposure has been established as the cause. SFT can occur across

a wide age range but is diagnosed most often in adults.

Diagnosis

Imaging can locate the tumor and show its size, blood supply, and relationship to nearby structures. Depending on its location, tests may include ultrasound, CT, or MRI. Imaging alone cannot reliably confirm that a mass is an SFT.

Confirmation usually requires a biopsy. During a biopsy, a clinician removes a small tissue sample so a pathologist can examine it under a microscope. When possible, biopsy planning is coordinated with the team that would perform surgery, because the biopsy path may affect the later operation.

SFT cells often have a spindle shape, meaning they look long and narrow under a microscope. The pathologist also uses tests called immunohistochemistry to look for proteins in the cells. Strong staining for STAT6 inside the cell nucleus is a key finding. Testing for the NAB2-STAT6 gene fusion may help when the diagnosis remains uncertain.

After confirming SFT, the team assesses its risk of returning or spreading. Features considered may include:

Additional scans may look for tumors elsewhere, particularly in the lungs. The exact tests depend on the original site and biopsy findings.

- Tumor size and location
- Your age at diagnosis
- How quickly the cells are dividing
- Areas of dead tumor tissue, called necrosis
- Whether the tumor has already spread

Treatment and management

Treatment is planned around the tumor's location, size, risk features, symptoms, and whether it has spread. Your general health and the possible effects of treatment also matter.

Surgery is the main treatment for a localized SFT. The aim is to remove the whole tumor with a rim of unaffected tissue, known as a clear or negative margin. Surgery can be curative when complete removal is possible, but it does not remove every risk of recurrence.

Some tumors are difficult to remove because they involve major blood vessels, nerves, or vital

organs. SFTs can also have a rich blood supply. Detailed imaging helps the surgical team prepare for these issues.

Radiation therapy uses high-energy beams to damage tumor cells. It may be considered when an SFT cannot be fully removed, has higher-risk features, returns in the same area, or is in a location where another operation would be difficult. It is not needed after every operation.

Systemic treatment travels through the bloodstream and may be used for an SFT that cannot be removed or has spread. Options can include medicines that limit the tumor's blood supply, sometimes called anti-angiogenic or targeted treatments. Traditional chemotherapy may be considered in selected cases. Because SFT is rare, there is no single drug plan that is right for everyone, and a clinical trial may be an option.

In selected situations, a small tumor that is not causing symptoms may be watched with scans rather than treated immediately. This is more likely when treatment would carry substantial risks. Any observation plan needs scheduled follow-up.

If an SFT causes low blood sugar, the immediate problem is treated while the team addresses the tumor responsible for it.

Complications

An SFT can return where it first developed. Recurrence may happen several years or, occasionally, decades after treatment. Higher-risk tumors are more likely to return, but no assessment can predict the future with certainty.

Some SFTs spread through the bloodstream. Common sites include the lungs, liver, and bones. A spreading tumor is called metastatic SFT.

Other complications depend on location. A tumor may compress an organ, nerve, airway, or blood vessel. Treatment itself can also affect nearby structures, particularly when the tumor is large or in a complex area. Your team should explain the expected benefits and possible effects before treatment.

Prevention

There is no known way to prevent a solitary fibrous tumor. The genetic change is usually acquired by chance and has not been tied to a preventable exposure or behavior.

Routine screening is not recommended for people without an SFT or symptoms. After diagnosis, scheduled imaging is follow-up rather than prevention. Its purpose is to find growth

or recurrence early enough to discuss treatment options.

Living with it and what to expect

Many people with a completely removed, localized SFT return to their usual activities after recovery. The time needed depends on the operation's location and extent. Rehabilitation may help if treatment affects strength, movement, breathing, speech, or nerve function.

Follow-up commonly continues for many years because SFT can return late. Visits may include a physical examination and scans of the original area, the chest, or both. The timing is based on the tumor's risk features, location, treatment, and previous scan results. There is no single schedule that fits every person.

A stable scan does not mean follow-up is unimportant. It is evidence that there is no visible change at that point. Keep appointments even when you feel well, and tell the team about new symptoms between visits.

People with recurrent or metastatic SFT may live with the condition over a longer period. Treatment may focus on controlling growth, relieving symptoms, and protecting organ function. Plans can change as scan results, symptoms, and treatment effects change.

Uncertainty about recurrence can be difficult. Clear information about your follow-up plan, access to your pathology report, and support from family, counseling services, or sarcoma organizations may make long-term care easier to manage.

When to get medical help

Arrange a routine medical appointment if you notice a new lump, an unexplained lump that is growing, or ongoing pressure or pain. These symptoms have many possible causes and do not mean that you have an SFT.

If you have been treated for SFT, contact your treating team before your next planned appointment if you develop a new lump, worsening pain, persistent cough, increasing breathlessness, unexplained weight loss, or new nerve-related symptoms.

Seek urgent medical care for repeated episodes of sweating, shaking, confusion, or faintness, especially if low blood sugar has been linked to your tumor.

Call emergency services for:

These emergency symptoms can have causes other than SFT, but they should not wait for routine follow-up.

- Severe or rapidly worsening breathing difficulty
- Fainting or inability to wake normally
- A first seizure or a seizure lasting several minutes
- Sudden weakness, facial drooping, or trouble speaking
- Severe confusion associated with suspected low blood sugar

Common questions

Is a solitary fibrous tumor cancer?

Some SFTs remain localized and behave in a low-risk way. Others can invade nearby tissue, return, or spread. Doctors assess the risk using the pathology findings, tumor size, age, and other features rather than relying only on the labels "benign" or "malignant."

Can a solitary fibrous tumor be cured?

Sometimes. Complete surgical removal can be curative for a localized tumor. Recurrence is still possible, including many years later, so long-term follow-up is usually important.

How fast does a solitary fibrous tumor grow?

Many grow slowly, but growth rates differ. Comparing scans taken at different times is more reliable than estimating growth from symptoms. Faster growth, increased cell division, and tumor necrosis may suggest a higher-risk tumor.

Can it come back after surgery?

Yes. An SFT may return at the original site or appear elsewhere. The chance is influenced by whether it was completely removed and by its size and microscopic features. Late recurrence is one reason follow-up can extend for years.

Is solitary fibrous tumor hereditary?

Usually not. The characteristic NAB2-STAT6 change is normally present only in the tumor cells. It is not generally inherited or passed to children.

What does "STAT6 positive" mean?

It means a laboratory stain found STAT6 protein inside the nuclei of the tumor cells. This pattern strongly supports an SFT diagnosis, although the pathologist considers it alongside the tissue's overall appearance and other test results.

Sources

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