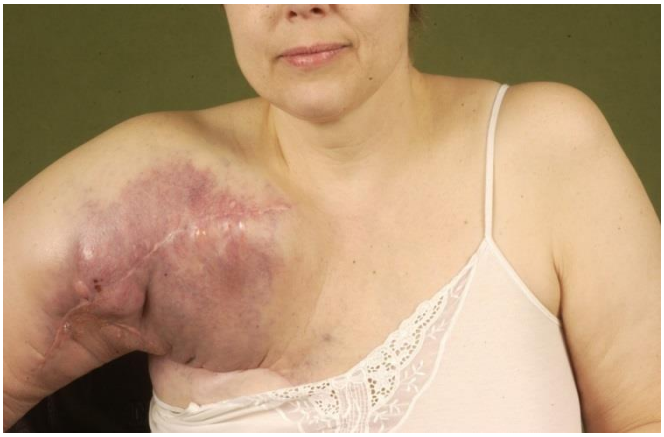
Cancer Association of South Africa (CANSA)

Research • Educate • Support

Fact Sheet
on
Spindle Cell Sarcoma

Introduction

A sarcoma (from the Greek σάρξ *sarx* meaning 'flesh') is a cancer that arises from transformed cells of mesenchymal origin. Thus, malignant tumours made of cancellous, cartilage, fat, muscle, vascular, or haematopoietic tissues are, by definition, considered sarcomas. This is in contrast to a malignant tumour originating from epithelial cells, which are termed carcinoma. Human sarcomas are quite rare.



[Picture Credit: Soft Tissue Sarcoma]

Cells of mesenchymal origin are multipotent stromal cells that can differentiate into a variety of cell types, including: osteoblasts (bone cells), chondrocytes (cartilage cells), myocytes (muscle cells) and adipocytes (fat cells). Mesenchymal cells can migrate easily, in contrast to epithelial cells, which lack mobility.

Sarcomas are rare cancers that develop in the muscle, bone, nerves, cartilage, tendons, blood vessels and the fatty and fibrous tissues.

There are about 100 different types of sarcoma that fall into three main types:

- Soft tissue sarcoma
- Bone sarcoma
- Gastro-intestinal stromal tumours (GIST)

Sarcomas can affect almost any part of the body, on the inside or the outside. Sarcomas commonly affect the arms, legs and trunk. They also appear in the abdomen as well as behind the abdomen (retroperitoneal sarcomas) and the female reproductive system (gynaecological sarcomas).

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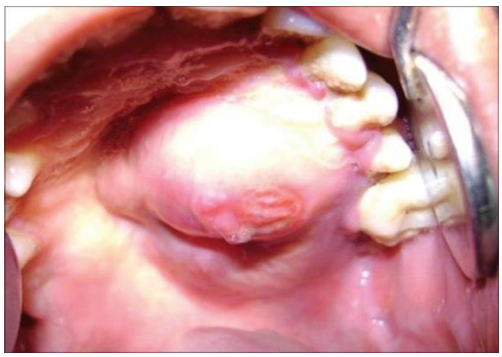
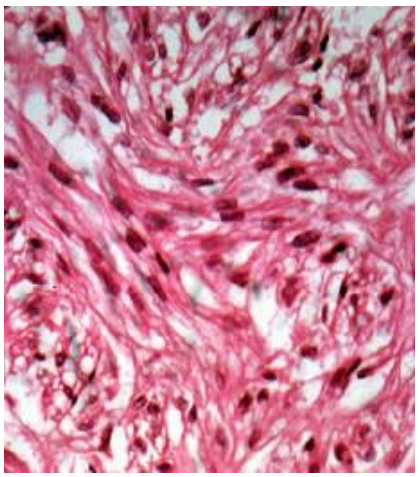
Spindle Cell Sarcoma

Spindle cell sarcoma is a type of connective tissue cancer in which the cells are spindle-shaped when examined under a microscope (see image below). The tumours generally begin in layers of connective tissue such as that under the skin, between muscles, and surrounding organs, and will generally start as a small lump with inflammation that grows.



[Picture Credit: Spindle Cell Sarcoma]



[Picture Credit: Spindle Cell Sarcoma Histopathology]

Classification of Spindle Cell Sarcoma

Adults above the age group of 40 years are more prone to spindle cell sarcoma. and it has also been traced in dogs, cats and even younger humans. Four various differentiations of spindle cell sarcoma arising from the connective tissue have been identified.

These are:

- Undifferentiated sarcoma of the bone: In this type of sarcoma the cells do not show proper specialisation. Since these cells do not belong to any other category of connective tissue cancerous cells they are termed as undifferentiated sarcoma cells.
- Malignant fibrous histiocytoma: This type of spindle cell sarcoma mostly affects legs and arms and is very rare. It is more common amongst middle aged adults.
- Fibrosarcoma: This type of sarcoma affects the thigh bone
- Leiomyosarcoma: Not much is known about this type of sarcoma.

Schwitzer, D., Kim, R., Williams, F. & Hammer, D. 2020.

"Spindle cell sarcoma (SCS) is a malignancy, with the most recent Surveillance, Epidemiology, and End Results (SEER) data citing a total of 250 reported cases occurring in the head and neck. Of these cases, none originated in the maxillofacial hard tissue. To the best of our knowledge, only 2 cases of primary osseous SCS of the maxillofacial region have been reported. These cases were not accounted for in the SEER data. The diagnosis of SCS requires its differentiation from other sarcomas and spindle cell neoplasms. Therefore, a comprehensive review to reinforce its inclusion in oral and maxillofacial surgeons' differential diagnosis for osseous neoplastic pathology is desired. In the present case report, we have described a maxillary SCS in a patient with an initial diagnosis of a spindle cell lesion of uncertain biologic behavior. We reviewed the data for SCS, including the epidemiologic data, diagnostic challenges, clinical and radiographic presentations, prognostic indicators, and treatment."

Incidence of Spindle Cell Sarcoma in South Africa
The National Cancer Registry (2016) does not provide any information regarding the incidence of Spindle Cell Sarcoma.

Signs and Symptoms of Spindle Cell Sarcoma
Spindle Cell Sarcoma is usually discussed under the umbrella of Soft Tissue Sarcoma.

More than half of sarcomas begin in an arm or leg. Most people simply notice a lump that has grown over a period of time (weeks to months). Although the lump is often not painful, in some cases it will hurt. When sarcomas grow in the back of the abdomen (the retroperitoneum), the symptoms they cause more often come from other problems. Sometimes the tumours cause pain.
The tumours may also cause blockage or bleeding of the stomach or bowels. It can grow large enough for the tumour to be felt in the abdomen. About 20% of sarcomas begin in the abdomen (stomach) area. Sarcomas can also begin on the outside of the chest or abdomen (about 10%) or in the head or neck area (around 10%).

Individuals who have any of the following problems, should see a doctor right away:

- A new lump or a lump that is growing (anywhere on the body)
- Abdominal pain that is getting worse
- Blood in stool or vomit
- Black, tarry stools (when bleeding happens in the stomach or bowels, the blood can turn black as it is digested, and it may make the stool very black and sticky)

These symptoms are more often caused by things other than sarcoma, but still needs to be checked out by a doctor.

Diagnosis of Spindle Cell Sarcoma
There are about 50 different types of soft tissue sarcomas. Diagnosis depends on where in the body the sarcoma is situated.

Imaging tests - Some tests, such as a computed tomography (CT) scan or a magnetic resonance imaging (MRI) scan, are often done to look for the cause of symptoms and to find a tumour (such as a sarcoma).

Other tests that may be done after a sarcoma is diagnosed to look for cancer spread.

Plain X-ray - a regular X-ray of the area with the lump may be the first test ordered. A chest X-ray may be done after the patient has been diagnosed to see if the sarcoma has spread to the lungs.

Computed Tomography Scans - the CT scan is an x-ray procedure that produces detailed, cross-sectional images of the body. Instead of taking one picture like a conventional x-ray, a CT scanner takes many pictures as it rotates around the body. A computer then combines these pictures into an image of a slice of the body. The machine will create multiple images of the part of the body being studied.

Magnetic Resonance Imaging Scans - magnetic resonance imaging (MRI) scans use radio waves and strong magnets instead of X-rays to take pictures of the body. The energy from the radio waves is absorbed and then released in a pattern formed by the type of tissue and by certain diseases. A computer translates the pattern of radio waves given off by the tissues into a very detailed image of parts of the body. A contrast material might be injected, just as with CT scans, but is used less often. MRI scans are often part of the work-up of any tumour that could be a sarcoma.

Ultrasound - ultrasound uses sound waves and their echoes to produce pictures of parts of the body. A small instrument called a transducer emits sound waves and picks up the echoes as they bounce off the organs. A computer converts the sound wave echoes into an image that is displayed on a computer screen.

Positron Emission Tomography Scan - in this test, radioactive glucose (sugar) is injected into the patient's vein to look for cancer cells. Because cancers use glucose (sugar) at a higher rate than normal tissues, the radioactivity will tend to concentrate in the cancer. A scanner can spot the radioactive deposits. A positron emission tomography (PET) scan is useful when the doctor thinks the cancer has spread but doesn't know where.

Biopsy - a biopsy is a procedure that removes a sample of tissue from a tumour to see if it is cancer. The piece of tissue is looked at under a microscope and, some other tests may be done on the sample as well. A physical examination or imaging test may suggest that a tumour is a sarcoma, but a biopsy is the only way to be certain that it is a sarcoma and not another type of cancer or a benign disease.

Fine Needle Aspiration (FNA) Biopsy - in FNA, the doctor uses a very thin needle and a syringe to withdraw small pieces of tissue from the tumour mass. The doctor can often aim the needle while feeling the mass near the surface of the body.

Core Needle Biopsy - core needle biopsies use a needle that is larger than the FNA needle. Sometimes this needle is called a Tru-Cut needle. It removes a cylindrical piece of tissue about 1/16 inch across and 1/2 inch long. It usually removes enough tissue to see if a sarcoma is present. Like FNA, CT scan and ultrasound can be used to guide the needle into tumours of internal organs.

Surgical Biopsy - in a surgical biopsy, the entire tumour or a piece of the tumour is removed during an operation. There are 2 types of surgical biopsies, excisional and incisional. In an excisional biopsy, the surgeon removes the entire tumour. In an incisional biopsy, only a piece of a large tumour is removed. An incisional biopsy almost always removes enough tissue to diagnose the exact type and grade of sarcoma. If the tumour is near the skin surface, this is a simple operation that can be done with local or regional anaesthesia (numbing medication given near the mass or into a nerve).

Grading of Soft Tissue Sarcomas

If a sarcoma is present, the biopsy will be used to determine what type it is and its grade. The grade of a sarcoma is based on how the cancer cells look under the microscope. In grading a cancer, the pathologist considers how closely the tumour resembles normal tissue (differentiation), how many of the cells appear to be dividing, and how much of the tumour is made up of dying tissue.

Each factor is given a score, and the scores are added to determine the grade of the tumour.

Treatment of Spindle Cell Sarcoma
The first line treatment for spindle cell sarcoma is surgical removal, cryotherapy, radiotherapy, hormone therapy and chemotherapy. The treatment of spindle cell sarcoma is selected independently depending on the location and size of the tumour. Combination therapy may also be decided upon for best results.

The most common spindle cell sarcoma treatment is surgical removal. The surgical removal treatment for spindle cell sarcoma has two types: conservative removal and wide surgical removal.

So, Y.K., Chow, C., To, K.F., Chan, J.K.C. & Cheuk, W. 2020.
"Neurotrophic tyrosine receptor kinase (*NTRK*)-rearranged spindle cell neoplasm is a recently described soft tissue tumor entity that occurs predominantly in children and young adults. The diagnosis of this tumor is difficult due to the nonspecific and highly variable morphology, yet it is of clinical relevance because of the availability of highly effective TRK inhibitors. In this article, we report the case of a 40-year-old female who presented with a mass in the left calf. Histologic examination revealed a low-grade sarcoma consisting of monomorphic spindle cells accompanied by abundant myxoid stroma, a feature that had not been emphasized in the reported cases of *NTRK*-rearranged tumors. The tumor cells expressed CD34 and S100 but not SOX10, and they showed positive staining for pan-TRK. Next-generation sequencing showed the presence of *LMNA*-*NTRK1* fusion. The patient developed several episodes of lung metastases that eventually became unresectable. TRK inhibitor was given that led to near-complete resolution of the tumors."

About Clinical Trials
Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.
Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The **South African National Clinical Trials Register** provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

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