Cancer Association of South Africa (CANSA)

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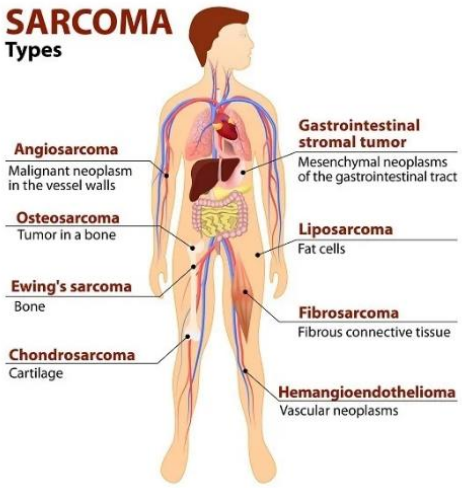
Fact Sheet on Vascular Sarcoma

Introduction

Sarcomas are rare tumours of connective tissue, and as a result, they can affect any part of the body. These are tumours of fat, nerves, bone, tendons, muscle and skin. They account for about 1% of all adult cancers and approximately 15% of paediatric tumours. In addition to the wide distribution of potential primary sites and the rarity, these are also very heterogeneous tumours with over 80 different histological subtypes.



[Picture Credit: Sarcoma Types]

These factors make sarcomas extremely challenging to treat. Consequently, it is very important that sarcoma patients are managed by an experienced multi-disciplinary team, including surgeons, pathologists, radiologists, oncologists, specialist nurses, physiotherapists and pharmacists.

Soft Tissue Sarcomas

Soft tissue sarcoma types are determined by the way a sample of cells from the tumour looks under a microscope. A surgeon will obtain the sample during a biopsy or during surgery to remove the tumour.

There are more than 80 types of soft tissue sarcoma or sarcoma-like growths. Knowing which type you have helps doctors predict how the tumour will respond to specific treatments. It also allows them to personalize your treatment.

Soft tissue sarcoma is usually classified based on where in the body the cancer started:

- If sarcoma starts in smooth muscle, it is called leiomyosarcoma.
- Gastrointestinal tract sarcoma is called a gastrointestinal stromal tumour.
- Soft tissue sarcoma in fatty tissue (also called adipose tissue) is called liposarcoma.
- Malignant schwannoma or malignant peripheral nerve sheath tumours start in the peripheral nervous system.

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- Soft tissue sarcoma starting in the blood vessels can be angiosarcoma, haemangioendothelioma, hemangiopericytoma, or a solitary fibrous tumour.
- Fibrosarcoma, dermatofibrosarcoma, low-grade fibromyxoid sarcoma, and fibromatosis are sarcomas of the connective tissue.

Soft tissue sarcoma tumours can affect more than one type of body tissue. They also may have no clear origin. This is the case for alveolar soft part sarcoma, clear cell sarcoma (malignant melanoma of soft parts), epithelioid sarcoma, synovial sarcoma, and undifferentiated soft tissue sarcoma.

Vascular sarcoma is one of the most common soft tissue sarcoma types.

Vascular Sarcoma

Vascular Sarcoma is a subtype of sarcoma of sarcoma which develops in the lining of blood vessels and more commonly affects adults versus children. This can form anywhere in the body and in some cases, blood vessel sarcomas can affect the lymphatic system. There are some more aggressive vascular sarcomas and some less aggressive vascular sarcomas which dictates the manner of treatment

Ravi, V. & Patel, S. 2013.

"Vascular sarcomas are soft-tissue tumors that arise from the endothelium with a malignant potential. This review discusses the management of epithelioid hemangioendothelioma (EHE) and angiosarcoma. EHE is a vascular tumor of intermediate malignant potential with an indolent course. EHE arising from the liver, lung, or bone tends to be multifocal and the rate of progression is slow and often unpredictable. Treatment should be considered in patients with significant symptomatic deterioration and/or progressive disease on imaging studies. Various cytotoxic and targeted therapies are available for management, with disease stabilization as the most common outcome. Angiosarcoma is an aggressive vascular tumor with a high malignant potential. Multidisciplinary care is critical for the management of localized disease, and the best outcomes are often observed in patients when a combination of systemic and local therapy options is used. Metastatic angiosarcoma is treated primarily with systemic therapy, and several cytotoxic and targeted therapies are available, alone or in combination. The choice of therapy depends on several factors, such as cutaneous location of the tumor, performance status of the patient, toxicity of the treatment, and patient goals."

Incidence of Vascular Sarcoma

The National Cancer Registry (2017) does not provide any information on vascular sarcoma.

Signs and Symptoms of Vascular Sarcoma

A soft tissue sarcoma may not cause any signs and symptoms in its early stages. As the tumour grows, it may cause:

- A noticeable lump or swelling
- Pain, if a tumour presses on nerves or muscles

Risk Factors and Causes of Vascular Sarcoma
In most cases, it's not clear what causes soft tissue sarcoma.

In general, cancer occurs when cells develop errors (mutations) in their DNA. The errors make cells grow and divide out of control. The accumulating abnormal cells form a tumour that can grow to invade nearby structures and the abnormal cells can spread to other parts of the body.

Diagnosis of Vascular Sarcoma
The diagnosis of vascular diagnosis is largely dependent on exactly where in the body it occurs.

Machado, I., Giner, F., Lavernia, J., Cruz, J., Traves, V., Requena, C., Llombart, B., López-Guerrero, J.A. & Llombart-Bosch, A. 2021.
"Angiosarcomas (AS) represent a heterogenous group of tumors with variable clinical presentation. AS share an important morphologic and immunohistochemical overlap with other sarcomas, hence the differential diagnosis is challenging, especially in poorly-differentiated tumors. Although molecular studies provide significant clues, especially in the differential diagnosis with other vascular neoplasms, a thorough hematoxylin and eosin analysis remains an essential tool in AS diagnosis. In this review, we discuss pathological and molecular insights with emphasis on implications for differential diagnosis in cutaneous, breast, soft tissue and visceral AS."

Treatment of Vascular Sarcoma
The best chance to cure a soft tissue sarcoma is to remove it with surgery, so surgery is part of the treatment for all soft tissue sarcomas whenever possible. It is important that your surgeon and other doctors are experienced in the treatment of sarcomas.

Stage I soft tissue sarcomas are low-grade tumours of any size. Small (less than 5 cm across) tumours of the arms or legs may be treated with surgery alone. The goal of surgery is to remove the tumour with some of the normal tissue around it.

For all stage II and III sarcomas, removing the tumour with surgery is the main treatment. Lymph nodes will also be removed if they contain cancer.

Radiation may be given after surgery.

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/

Medical Disclaimer

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Sources and References Consulted and/or Utilised

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Sarcoma Types
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Vascular Sarcoma
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