

Cancer Association of South Africa (CANSA)



Fact Sheet on Undifferentiated Pleomorphic Sarcoma

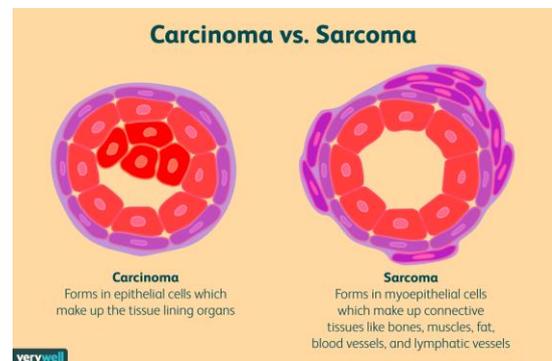
Introduction

A carcinoma forms in the skin or tissue cells that line the body's internal organs, such as the kidneys and liver. A sarcoma grows in the body's connective tissue cells, which include fat, blood vessels, nerves, bones, muscles, deep skin tissues and cartilage.

[Picture Credit: Carcinoma vs Sarcoma]

Sarcomas are categorized in two ways:

- Soft tissue sarcoma, which forms in soft tissues
- Bone sarcoma (or osteosarcoma), which develops in bone tissue, cartilage or bone marrow



New research, though, has found that some sarcomas have a greater immune response than others, and may respond to certain checkpoint inhibitors. These drugs are a type of immunotherapy that works by disrupting the communication signals that allow cancer cells to hide from the immune system, exposing the tumour to the body's defenses. The study identified immune response patterns in common sarcoma subtypes, including leiomyosarcoma and liposarcoma, both soft tissue sarcomas.

Undifferentiated Pleomorphic Sarcoma (UPS)

Undifferentiated pleomorphic sarcoma (UPS) is a rare type of cancer that begins mostly in the soft tissues of the body. Soft tissues connect, support and surround other body structures. UPS usually occurs in the arms or legs. Less often it can happen in the area behind the abdominal organs (retroperitoneum).

The name undifferentiated pleomorphic sarcoma (UPS) comes from the way the cancer cells appear under the microscope. Undifferentiated means the cells don't look like the body tissues in which they develop. The cancer is called pleomorphic because the cells grow in multiple shapes and sizes.

Undifferentiated Pleomorphic Sarcoma (UPS) used to be called malignant fibrous histiocytoma.

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Incidence of Undifferentiated Pleomorphic Sarcoma (UPS)

The South African National Cancer Registry (2017) does not provide any information on Undifferentiated Pleomorphic Sarcoma.

Risk Factors for Undifferentiated Pleomorphic Sarcoma (UPS)

It's not clear what causes undifferentiated pleomorphic sarcoma.

Doctors know this cancer begins when a cell develops changes in its DNA. A cell's DNA contains the instructions that tell a cell what to do. The changes tell the cell to multiply rapidly, creating a mass of abnormal cells (tumour). The cells can invade and destroy nearby healthy tissue. In time, the cancer cells can break away and spread (metastasize) to other parts of the body, such as the lungs and bones.

Signs and Symptoms of Undifferentiated Pleomorphic Sarcoma (UPS)

Signs and symptoms of Undifferentiated Pleomorphic Sarcoma may include:

- Growing lump or area of swelling.
- If it grows very large, there may be pain, tingling and numbness.
- If it occurs in an arm or leg, there may be swelling in the hand or foot of an affected limb.
- If it occurs in the abdomen, there may be pain, loss of appetite and constipation.
- Fever.
- Weight loss.

Diagnosis of Undifferentiated Pleomorphic Sarcoma (UPS)

This cancer is often diagnosed after other types of cancer have been ruled out.

Tests and procedures may include:

- **Physical examination** - Your doctor will ask you questions about when your symptoms began and if they've changed over time. He or she will examine the area to better understand the size and depth of the growth, whether it's connected to nearby tissues, and whether there are any signs of swelling or nerve damage.
- **Imaging tests** - Your doctor may recommend imaging tests to create pictures of the affected area and understand more about your condition. Imaging tests may include X-rays, CT, MRI and positron emission tomography (PET) scans.
- **Removing a sample of tissue for testing (biopsy)** - To make a definitive diagnosis, your doctor collects a sample of the tumour tissue and sends it to a lab for testing. Depending on your particular situation, the tissue sample may be collected with a needle inserted through your skin or during an operation.

In the lab, doctors trained in analysing body tissues (pathologists) examine the sample to determine the types of cells involved and whether the cells are likely to be aggressive. This information helps rule out other types of cancer and guides your treatment.

Allen, A.H. 2019. Large undifferentiated pleomorphic sarcoma of the posterior thigh. *Am J Case Rep.* 2019 Mar 11;20:318-322.

BACKGROUND: Sarcomas account for less than 1% of all cancers. Undifferentiated Pleomorphic Sarcoma, formerly called Malignant Fibrous Histiocytoma, is a rare subtype identified by a lack specific immunohistochemical markers for a specific lineage of differentiation. These soft tissue tumors are aggressive and rapidly enlarge. Risk for metastasis increases almost linearly as the tumor increases in size, emphasizing the importance of early detection, treatment, and post-resection monitoring.

CASE REPORT: This article reports a case of a large undifferentiated pleomorphic sarcoma of the posterior thigh in a 62-year-old female. Given the patient's history of thrombotic thrombocytopenic purpura, her initial mass was thought to be a hematoma following a hernia repair surgery. After diagnosis of undifferentiated pleomorphic sarcoma, she underwent radical excision revealing a 24x9.5x7cm lesion - one of the largest reported in the literature.

CONCLUSIONS: Sarcomas are very rare soft tissue neoplasms, but they should not be excluded in a physician's differentials when a patient presents with an enlarging soft tissue mass. Because sarcomas enlarge rapidly, delay in evaluation and management should be avoided and these patients should be quickly referred to a center specializing in sarcoma treatment. Magnetic Resonance Imaging (MRI) is the recommended initial imaging for all soft tissue masses of the extremities, trunk, and head and neck while Computed Tomography (CT) is the recommended imaging choice for retroperitoneal and visceral masses. After successful surgical excision with clean margins, patients should undergo serial monitoring by CT or MRI for surveillance of recurrence or late pulmonary metastases.

Treatment of Undifferentiated Pleomorphic Sarcoma (UPS)

Treatment for undifferentiated pleomorphic sarcoma usually involves surgery to remove the cancer cells. Other options include radiation therapy and drug treatments (systemic therapies), such as chemotherapy, targeted therapy, and immunotherapy.

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](https://www.sanctr.gov.za/) 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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Sources and References Consulted and/or Utilised

Allen, A.H. 2019. Large undifferentiated pleomorphic sarcoma of the posterior thigh. *Am J Case Rep.* 2019 Mar 11;20:318-322.

Carcinoma vs Sarcoma

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Sarcoma

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Undifferentiated Pleomorphic Sarcoma

[https://www.mayoclinic.org/diseases-conditions/undifferentiated-pleomorphic-sarcoma/symptoms-causes/syc-20389554#:~:text=Undifferentiated%20pleomorphic%20sarcoma%20\(UPS\)%20is,the%20abdominal%20organs%20\(retroperitoneum\).](https://www.mayoclinic.org/diseases-conditions/undifferentiated-pleomorphic-sarcoma/symptoms-causes/syc-20389554#:~:text=Undifferentiated%20pleomorphic%20sarcoma%20(UPS)%20is,the%20abdominal%20organs%20(retroperitoneum).)

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