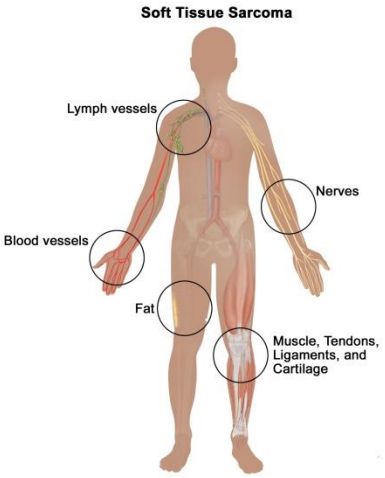
Cancer Association of South Africa (CANSA)

Research • Educate • Support

Fact Sheet
on
Soft Tissue Ewing's Sarcoma

Introduction

Soft Tissue Ewing's Sarcoma, also known as Extraosseous Ewing's Sarcoma (EES) or Extraskeletal Ewing's Sarcoma. It develops in the soft tissue around the bone. It is found in the trunk, limbs and the brain and mostly affects individuals under 25 years of age. These soft tissue tumours behave a bit differently to other types of soft tissue sarcoma. They're usually treated in the same way as Ewing's sarcoma that start in the bone.



[**Picture Credit**: Soft Tissue Ewing's Sarcoma Picture]

Gargallo, P., Yáñez, Y., Juan, A., Segura, V., Balaguer, J., Torres, B., Oltra, S., Castel, V. & Cañete, A. 2020.

"Ewing sarcoma is a rare tumor developed in bone and soft tissues of children and teenagers. This entity is biologically led by a chromosomal translocation, typically including EWS and FLI1 genes. Little is known about Ewing sarcoma predisposition, although the role of environmental factors, ethnicity and certain polymorphisms on Ewing sarcoma susceptibility has been studied during the last few years. Its prevalence among cancer predisposition syndromes has also been thoroughly examined. This review summarizes the available evidence on predisposing factors involved in Ewing sarcoma susceptibility. On the basis of these data, an integrated approach of the most influential factors on Ewing sarcoma predisposition is proposed."

Durer, S. & Shaikh, H. 2020.

Ewing sarcoma (ES) is an aggressive tumor of adolescents and young adults, which constitutes 10% to 15% of all bone sarcomas. James Ewing first described it in 1921, and it represents 'classic' Ewing sarcoma of bone, extra-skeletal Ewing sarcoma, malignant small cell tumor of the chest wall (Askin tumor), and soft tissue-based primitive neuroectodermal tumors (PNET). These sarcomas originate from unique mesenchymal progenitor cells due to their similar histologic and immunohistochemical characteristics. Ewing sarcoma family tumors (ESFT) are characterized by the presence of non-random chromosomal translocations producing fusion genes that encode aberrant transcription factors. The t(11;22)(q24;q12) translocation is associated with 85% of tumors and leads to EWS-FLI-1 formation, whereas t(21;12)(22;12) and other less common translocations induced EWS-ERG fusion comprises the remaining 10% to 15% of cases. The most common anatomical sites include the pelvis, axial skeleton, and femur; however, it may occur in almost any bone or soft tissue. Typically, patients present with pain and swelling over the site of involvement. Over the last 40 years, both local therapy and multiagent adjuvant chemotherapy have achieved considerable

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progress in the treatment of localized disease that improved 5–year survival rate from less than 20% to greater than 70% but the recurrence rate remains high. Although most present locally, subclinical metastatic disease is present in almost all. Approximately 25% of patients with initially localized disease ultimately relapse. No standard therapy exists for relapsed and refractory ES, with survival rates being less than 30% in those with isolated lung metastases and less than 20% in those with bone and bone marrow involvement. Given these considerations of toxicity and suboptimal survival from metastatic disease, there is an urgent unmet need to develop novel therapies for ES.

Incidence of Soft Tissue Ewing's Sarcoma

The National Cancer Registry (2016) does not provide any information on Soft Tissue Ewing's Sarcoma.

Signs and Symptoms of Soft Tissue Ewing's Sarcoma

Signs and symptoms may include:

- A lump in the bone of the arm or leg
- Pain in a bone
- Swelling and warmth near a bone
- Bone pain that does not get better over time or lasts longer than expected for a minor injury
- Pain may come and go
- Pain may be worse or better at night
- Limping
- Fever with no known cause
- Unintended weight loss
- Broken bone with no known cause

If the cancer spreads, or metastasizes, it usually goes to the lungs, other bones, or to the bone marrow (the spongy material inside the bone).

Causes and Risk Factors of Soft Tissue Ewing's Sarcoma

Doctors and researchers do not know what causes most cancers in children and teens, but the following factors may raise a person's chance of developing any form of Ewing's sarcoma:

- Genetic changes. Changes in a tumour cell's chromosomes appear to be responsible for Ewing sarcoma - the disease is not inherited. The genetic changes occur for no known reason.
- Age. Soft Tissue Ewing's Sarcoma can occur at any age. More than half of people 10 and 20, with a median age of 15 years.
- Gender. It is more common among boys than girls.
- Race/ethnicity. It occurs most frequently in white people and appears to be rare in black people.

Diagnosis of Soft Tissue Ewing's Sarcoma

The doctor usually diagnoses sarcoma through a series of tests. These may include:

- Physical examination

- A scan
  - Computed tomography (CT) scan
  - Magnetic Resonance Imaging (MRI)
    Features of Ewing's sarcoma are non-specific, and a radiological differential diagnosis should be considered
- A biopsy – taking and testing a tissue sample for examination by a specialist sarcoma pathologist. The results from a biopsy can inform the grade of the cancer

A diagnosis of bone sarcoma should be confirmed by a specialist sarcoma pathologist who will identify the type of sarcoma and the stage and grade of the tumour. Identifying the stage and grade of a cancer means the treating physician can advise on the best course of treatment. The stage of a cancer is measured by how much it has grown or spread which can be seen on the results of the tests and scans.

Houser, J.D. & Carow, S.D. 2020.
"A 21-year-old male Army basic trainee was evaluated in a direct-access physical therapy clinic for left-sided low back pain. Following examination, radiographs and a bone scan were ordered. The findings prompted referral for a computed tomography scan of the pelvis, which characterized an infiltrative soft tissue mass. The therapist immediately contacted an orthopaedic surgeon, who recommended that the physical therapist order immediate magnetic resonance imaging (STAT MRI). The patient underwent a tissue biopsy and subsequently was diagnosed with Ewing's sarcoma. *J Orthop Sports Phys Ther 2020;50(5):276. doi:10.2519/jospt.2020.9109*."

Treatment of Soft Tissue Ewing's Sarcoma
Treatment of Soft Tissue Ewing's Sarcoma may include:
- Chemotherapy
- Surgery
- Radiation Therapy
- Stem cell transplant – currently still as clinical trial
- Bone marrow transplant – currently still as clinical trial

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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Soft Tissue Ewing's Sarcoma Picture
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