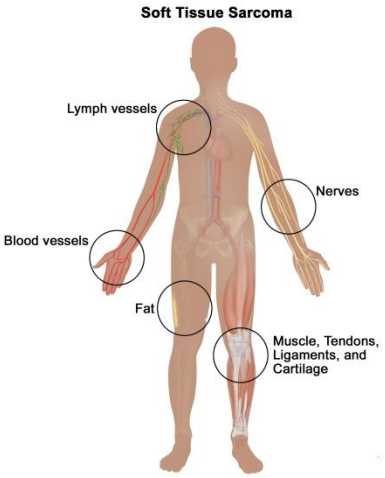
Cancer Association of South Africa (CANSA)

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]

Gurria, J.P. & Dasgupta, R. 2018.
"Ewing sarcoma (ES) is a round cell tumor, highly malignant and poorly differentiated that is currently the second most common malignant bone tumor in children. In rare instances, it develops from an extraskeletal origin, classified as extraosseous Ewing sarcoma (EES)."

Incidence of Soft Tissue Ewing's Sarcoma

The National Cancer Registry (2014) does not provide any information on Soft Tissue Ewing's Sarcoma.

Martin, R.C. 2nd, & Brennan, M.F. 2003.

BACKGROUND: Ewing sarcoma (ES) is the second most common primary osseous malignancy in childhood and adolescence. The improvement in survival is primarily associated with the combination of surgery and chemotherapy.

HYPOTHESIS: Little is known about the outcome of adults with soft tissue ES or primitive neuroectodermal tumors (PNET). Certain prognostic factors from soft tissue sarcomas (tumor size, tumor location, margin status, and initial presentation) in adults (>16 years) with ES/PNET will help to identify factors associated with outcome.

METHODS: Between July 1, 1982, and June 30, 2000, we identified 59 adult patients with primary soft tissue ES/PNET. Clinicopathologic factors were correlated with the end points studied: patient factors, tumor factors, pathologic factors, status of surgical margins, adjuvant chemotherapy, and radiation therapy.

RESULTS: There were 41 male and 18 female patients, with a median age of 27 years (range, 16-72 years). Median tumor size was 8 cm, with all lesions being high grade. The most common site was the trunk (n =

22), with an even distribution of retroperitoneal, pelvis, buttock, and lower extremity (all n = 5). The median follow-up was 29 months (range, 6-222 months), with local recurrence identified in 13 patients (22%), with a median time to recurrence of 15 months (range, 5-200 months). Overall 5-year survival was 60%. Initial presentation was the only predictor of long-term survival, with primary tumor-only presentation having a 5-year survival of 60% (median not reached) compared with primary tumor plus metastatic disease having a 5-year survival of 33% (median, 17 months) (P =.02).
CONCLUSION: Initial presentation of disease represents the only predictor of survival identified in this small group of adult patients with ES/PNET.

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.

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/

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Sources and References Consulted and/or Utilised

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Soft Tissue Ewing's Sarcoma
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Soft Tissue Ewing's Sarcoma Picture
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