Ewing's Sarcoma

Ewing's sarcoma is the second most common bone sarcoma in children. It is a malignant, small round cell tumor caused by an 11;22 translocation of the EWS gene on chromsome 22 and the ETS transcription factor, which results in abnormally increased cell proliferation. It most frequently affects boys between 10-15 years old. It tends to present as extreme bone pain with a rapidly enlarging mass in the diaphysis of long bones, like the femur, as well as the pelvis, scapula and ribs. There may be an elevated ESR, CRP, or leukocytosis from an inflammatory response. Radiographically, it appears as a lytic lesion with an onion-skin appearance of the periosteum. This characteristic onion-skinning is from reactive bone formation in a lamellated, concentric pattern. It is an aggressive tumor with hematogenous spread of metastases, but it is very responsive to chemotherapy with or without surgical excision.



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Epidemiology

Most Common in Boys < 15 years old
Little league <15 years old
Ewing's sarcoma is most common in boys less than 15 years of age, and is the second most common bone sarcoma in children.

Pathophysiology

11;22 Translocation
(11) Double Wand; (22) Double Tutu
There is a translocation of the EWS gene on chromosome 22 and the ETS transcription factor on chromosome 11, which causes abnormal cell proliferation and survival.

Characteristics

Long bones, Pelvis, Ribs
Long bone, pelvis, ribs
Ewing's sarcoma tends to affect the long bones, as well as the flat bones of the pelvis, scapula and the ribs.

Diaphysis
Dice-fist
Ewing's sarcoma tends to present in the long shaft of the bone, known as the diaphysis.

Small Blue Cells
Small Blue-spots
Ewing's sarcoma histologically appears as sheets of small blue cells that are slightly larger than lymphocytes and contain little cytoplasm.

Malignant
Malignant-man
Ewing's sarcoma is a malignant tumor that often metastasizes hematogenously to the lungs, bone marrow and other bones.

Aggressive
Aggressive-expression
Ewing's sarcoma is an aggressive tumor that can invade beyond the bone into the soft tissue.

Onion-skin Appearance
Onion
Radiographically, the tumor has an onion- skin appearance, and is observed as a periosteal reaction. A periosteal reaction is the formation of new bone in response to injury, or other stimuli of the periosteum surrounding the bone. This occurs because the body treats this tumor as a permeative lytic lesion. This

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characteristic onion-skinning is from reactive bone formation in a lamellated, concentric pattern.

TREATMENT

Responsive to Chemotherapy

Chemotherapy head-wrap

Ewing's sarcoma responds very well to chemotherapy with or without surgical resection.

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