

EWING SARCOMA– GUIDELINES

Suspicious signs suggestive of a sarcoma:

- The commonest symptom of a primary bone sarcoma is non mechanical pain
- The presence of pain or a palpable mass arising from any bone should be viewed with suspicion
- The presence of any of the following on the X-ray is suggestive, but not diagnostic of a bone sarcoma:
 - ♦ bone destruction
 - ♦ new bone formation
 - ♦ periosteal swelling
 - ♦ soft tissue swelling

EWING SARCOMA

- Ewing's sarcoma (including primitive neuroectodermal tumor of bone/PNET) second most common primary malignant bone cancer in children and adolescents, but is also seen in adults
- Most frequent sites of involvement are the long bones and pelvis
- All forms of Ewing's sarcoma are high-grade tumors

Biopsy

- Biopsy diagnosis is mandatory
- Biopsy to be done only after all local imaging is completed
- In most cases a core needle biopsy is adequate (it may need to be image guided depending on anatomical location of lesion)
- Ideally performed at centre which will do definitive management of disease
- Immunohistochemistry confirmation desirable , may need additional cytogenetic and molecular studies

Serological Investigations

- Though there are no specific laboratory tests for diagnosis some maybe of prognostic value; e.g. alkaline phosphatase (ALP) and lactate dehydrogenase (LDH)

Staging

- Local X Ray
- MRI

- PET CT Scan / or if PET not available : Bone scan + CT Thorax & Bone marrow aspiration & biopsy

EWING SARCOMA– NON METASTATIC AT PRESENTATION

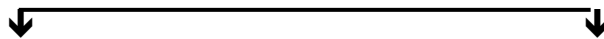
Induction chemotherapy (chemotherapy is multiagent) for at least 9 weeks prior to local therapy



Evaluation for local therapy between week 9 and 12 (reimaging with MRI recommended)



Limb sparing surgical resection possible with adequate oncologic margins



Yes

No

Limb sparing surgery

Extremity Lesion

CentroAxial Lesion

Definitive Radiotherapy vs Ablative surgery
(Discuss with patient and multidisciplinary treating team)

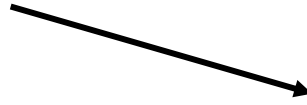
Radiotherapy

Indications for post-operative radiotherapy

	Negative margins	Positive margins
≥ 90 % necrosis	No adjuvant RT	Adjuvant RT
< 90 % necrosis	Discuss in multidisciplinary clinic	Adjuvant RT

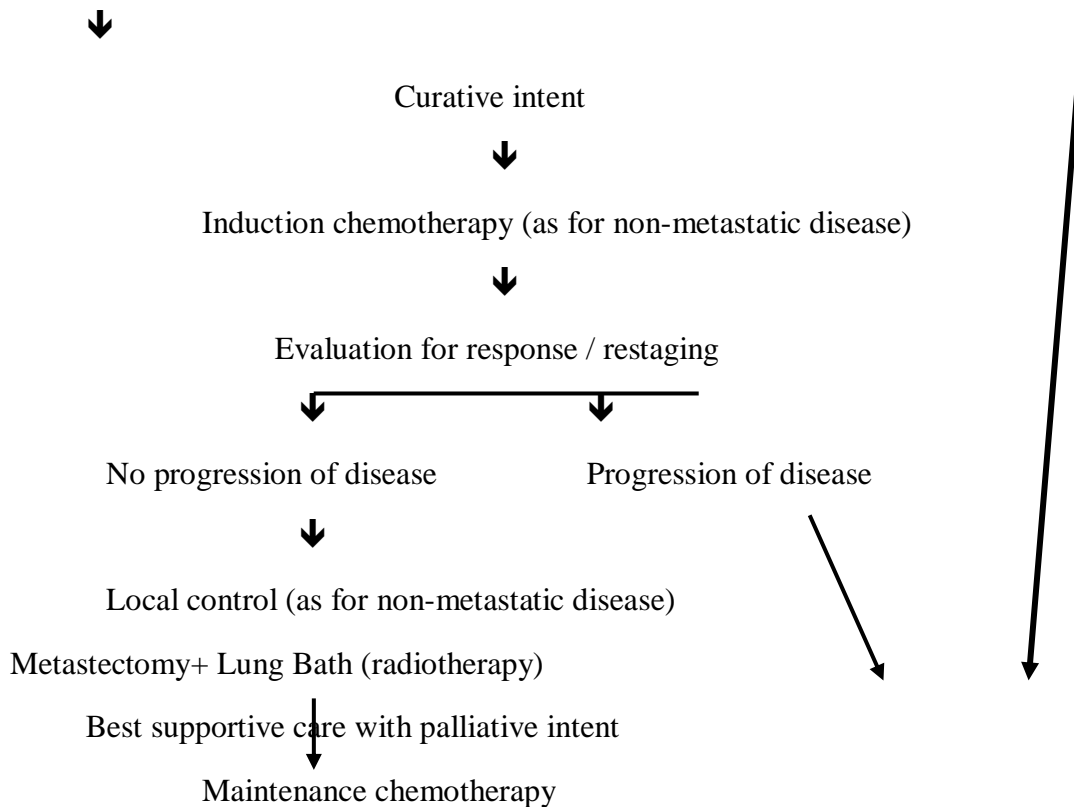


Maintenance chemotherapy



EWING'S SARCOMA – METASTATIC AT PRESENTATION

To evaluate for intent of treatment based on site and number of metastasis



Post treatment surveillance:

- Relapses most often occur to the lungs
- Risk assessment based on tumor grade, tumor size and tumor site may help in choosing the most suitable follow-up policy
- MRI to detect local relapse and CT scan for lung metastases is likely to pick up recurrence earlier but it is yet to be demonstrated that this is beneficial or cost effective compared with clinical assessment of the primary site and regular chest X-rays
- Local examination, chest and local imaging every 3 to 6 months for first 2 years, every 6 months for next 3 years and annually after year 5 is suggested
- Extended surveillance may be necessary to identify and address potential late effects of surgery, radiation and chemotherapy for long term survivors.

