Ewing's Sarcoma

INTRODUCTION

- Ewing's sarcoma is believed to arise from endothelial cells in the bone marrow.
- It occurs most commonly between the ages of 10 and 20 years, usually in a tubular bone and especially in the tibia, fibula or clavicle.
- Macroscopically the tumour is lobulated and often fairly large.
- Microscopically, sheets of small, dark polyhedral cells with no regular arrangement and no ground substance are seen.

INTRODUCTION

- Affect young age- 80% of patients are <20yrs. Rare after 40 yrs.
- Slightly male preponderance
- Aetiology- distinct genetic link

PATHOLOGY

- Tend to arise in the diaphysis of long bones
- Other places

1. - Pelvis
2. - Ribs
3. - Skull
4. - Vertebrae
5. - Scapula

PATHOLOGY

- Can occur in the soft tissue without bone involvement
- Metastasize to chest (25%) & bone (5%)

CLINICAL FEATURES

- The patient presents with **pain** – often throbbing in character – and swelling.
- Generalized illness and pyrexia, together with a **warm, tender swelling** and a raised ESR, may suggest a diagnosis of **osteomyelitis**

IMAGING

- X-ray usually shows an **area of bone destruction** which, unlike that in osteosarcoma, is predominantly in the mid diaphysis.
- New bone formation may extend along the shaft and sometimes it appears as fusiform layers of bone around the lesion – the so-called **'onion-peel'** effect.
- CT and MRI will reveal any large extraosseous component and radio-isotope scans may disclose **multiple lesions** elsewhere in the skeleton.

TYPICAL RADIOLOGICAL FEATURES

Ill defined osteolytic lesion

Wide zone of transition

Multilayered periosteal reaction(Onion skin)

Moth-eaten appearance

DIAGNOSIS

- The condition which should be excluded as rapidly as possible is **bone infection**.
- On biopsy the essential step is to recognize this as a malignant round-cell tumour, distinct from osteosarcoma.
- Other round-cell tumors that may resemble Ewing's are **reticulum-cell sarcoma** and **metastatic neuroblastoma**.

TREATMENT

- The prognosis is always poor and surgery alone does little to improve it.
- Radiotherapy has a dramatic effect on the tumour but overall survival is not much enhanced.
- Chemotherapy is more effective, offering a 5-year survival rate of about 50%.

TREATMENT

- The best results are achieved by a combination of **all three methods**: a course of preoperative **neoadjuvant chemotherapy**; then **wide excision** if the tumour is in a favorable site, or **radiotherapy** followed by local excision if it is less accessible; and then a further course of chemotherapy for 1 year