Orthopedics

lecture no.18

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Neoplastic conditions of bone

General principles

The definition of a neoplasm is a group of cells which continue to proliferate indefinitely in an uncontrolled fashion. The distinction between these cells and those which proliferate for a time, eventually becoming mature, is a blurred one. It is convenient to define the second group as hamartomata. True tumors are usually sub-divided into benign and malignant but, again, the distinction is not always clear either from the histological or behavioral point of view.

Causes, incidence, and risk factors:

The cause of bone tumors is unknown. They often arise in areas of rapid growth. Possible causes include:

· Inherited genetic mutations
· Radiation
· Trauma

The incidence of bone cancer is higher in families with familial cancer syndromes. The incidence of bone cancer in children is approximately 5 cases per million children each year.

Bone tumors

A classification of the commonest type of bone tumors is given below:

1. Hamartomas:

· Fibrous cortical defect
· Fibrous dysplasia
· Simple bone cyst

2. Benign bone tumors:

· Aneurysmal bone cyst.
· Enchondroma.
· Osteochondral.
· Chondroblastoma.
· Chondromyxoid fibroma.
· Osteoid osteoma/osteoblastoma.
· Giant cell tumor.

3. Malignant bone tumors:

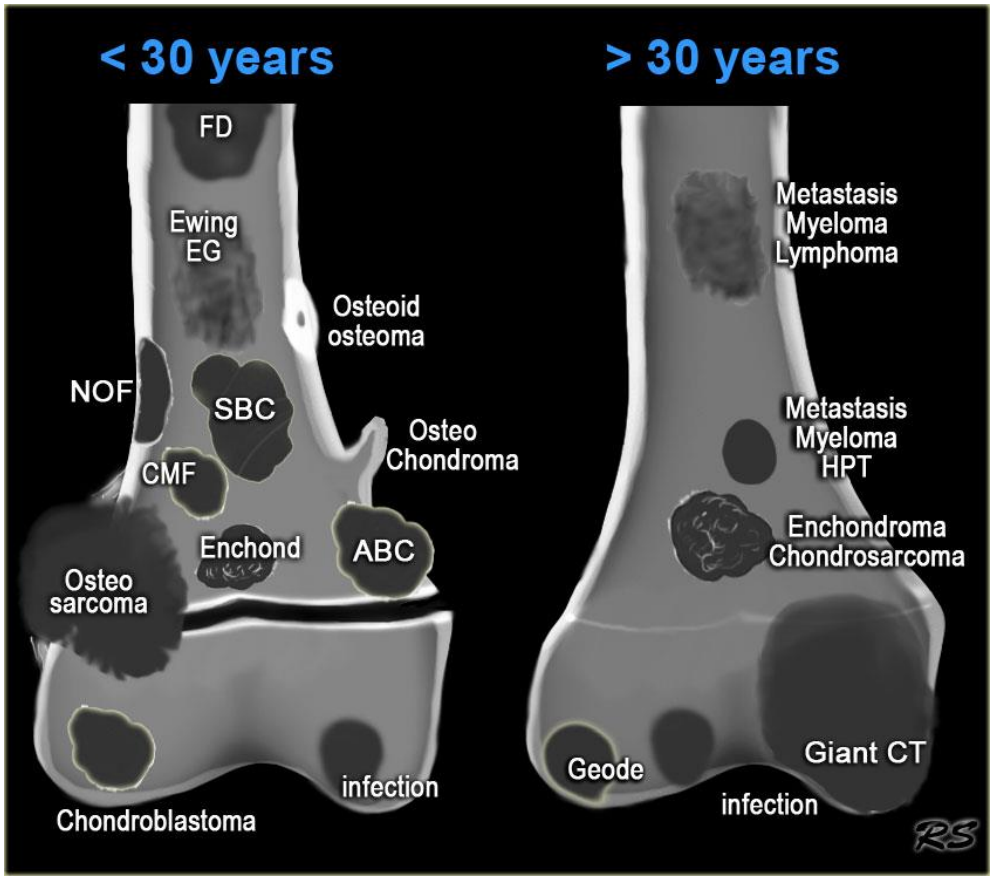
· Osteosarcoma.
· Ewing's sarcoma.
· Chondrosarcoma.
· Spindle cell sarcomas (including fibro-sarcoma., leiomyosarcoma, Malignant Fibrous Histiocytoma).

4. Hematological malignancies:
· Plasmacytoma /myeloma.
· Non-Hodgkin's lymphoma
5. Metastases to bone: mostly from epithelial cancers, the commonest being breast, bronchus, prostate, thyroid and kidney.

Presentation

· Age: bone tumors usually present within the first three decades except chondrosarcoma, myeloma and secondary's that occur later.
< 5 years: metastatic neuroblastoma.
5-20 years: Ewing sarcoma.
Teen's age: osteosarcoma.
· Sex: male > females
· Site: Epiphysis: Chondroblastoma, giant cell tumor. Diaphysis: Ewing, lymphoma. Metaphysis: any tumor e.g. osteosarcoma.
· Spine: Anterior column (mostly malignant), posterior column (mostly benign, but sometimes aggressive or malignant).
· Pelvis: myeloma, chondrosarcoma, Ewing.

Most bone tumors present with a history of increasing pain in the affected limb sometimes accompanied by a swelling and sometimes followed by a pathological fracture.



Symptoms of primary bone tumor:

- Bony swelling (before pain in benign lesions, & after pain in malignant lesions).
- Bone pain (may occur with or without movement and more intense at night).
- May experience dull and usually localized pain.
- Pressure manifestation e.g. paraplegia.
- May include a mass or tumor, which may be tender and may swell
- Pathologic fractures
- Cachexia (general ill health and malnutrition)
- Impaired mobility (which may occur during late stages)

The key investigation for any patient with non-specific pain, particularly non mechanical pain (i.e. not related to movement) is a plain X-ray of the affected part.

Radiological features indicative of pathological process.

- Bone destruction
- New bone formation
- Periosteal reaction
- Soft tissue swelling

Stepwise assessment of a suspected case:

- History
- Examination
- Simple tests: X-ray, bloods, chest x-ray
- Scan: bone scan, CT chest, angiography
- MRI
- Biopsy
- Staging of the tumor according to Enniking staging:

Investigation of a patient with an abnormal area of bone

The aim of investigating the patient is to reach a diagnosis in as short a time as possible. The most likely diagnosis will often be indicated by the patient's age, location of the lesion and the plain X-ray appearance, e.g. a lytic destructive tumor in an adolescent is likely to be a sarcoma, but in a 65-year-old it is likely to be a metastasis.

· **Step 1.** Take a detailed history from the patient, including past medical history (cancer) and current symptoms.
· **Step 2.** Examine the patient, including local examination (mass, limb) & general examination including sites of potential primary tumors (breast, bronchus, thyroid, kidney and prostate).
· **Step 3.** Arrange simple investigations, including chest X-ray and blood tests (full blood count, erythrocyte sedimentation rate and biochemical profile as a minimum, in older patients include prostate-specific antigen and myeloma screen).
· **Step 4.** If no diagnosis is apparent, arrange a bone scan to see if the lesion is solitary, and in patients over the age of 35 arrange for CT chest and abdomen.
· **Step 5.** If the lesion is solitary and no diagnosis is still apparent, arrange an MRI of the tumor and refer the patient to a tumor unit for biopsy & staging.

MRI:
Will show the full bony and soft tissue extent and relate the tumor to other nearby anatomic structures (e.g. vessels).

CT:
It can also be used to define the extra osseous extent of the tumor, especially in the skull, spine, ribs and pelvis. Both CT and MRI can be used to follow response to radiation and/or chemotherapy.

Biopsy:
Biopsies are usually carried out under X-ray or CT control using needles especially in suspected lesions. In benign lesions excisional biopsy is usually the rule. Samples must be sent for both histology and microbiology (to rule out infection) in every case, and in children should also be sent for cytogenetics. The biopsy should be done in a unit where there is experience of bone tumors and in particular where the pathologist is familiar with the very complex interpretation of bone tumor biopsies.

Grading of bone tumors:

According to Enniking:

Benign lesions: stage1 (latent), stage2 (active), stage3 (aggressive)

Malignant lesions:

· Stage IA..... Low grade intra compartmental.
· Stage IB..... Low grade extra compartmental.
· Stage IIA..... High grade intra compartmental.
· Stage IIB..... High grade extra compartmental.

Treatment

Benign lesions

Treatment for benign bone lesions:

- Observation; for latent lesions.
- Curettage & graft: for active lesions.
- Excision reconstruction: for aggressive lesions.

Malignant lesions

Treatment for primary bone cancer is surgery either before or after chemotherapy (and/or radiation therapy). Trans-arterial selective embolization can be used preoperatively to diminish blood loss during resection aggressive & malignant lesions.

· Patients may be started on a course of radiation therapy or chemotherapy first to shrink their tumors so that they are amenable to surgery.
· Surgery in these cases depends on the type of cancer, its location, how advanced it is, how aggressive it is and the age and functional status of the patient.
· Limb reconstructive surgery following treatment for cancer is now possible and improves the functional results in many instances.

Limb Salvage Surgery (Limb Sparing Surgery for Bone Tumors)

· Limb salvage surgery includes all of the surgical procedures designed to accomplish removal of a malignant tumor and reconstruction of the limb with an acceptable oncologic, functional, and cosmetic result.

· In limb-sparing surgery, the surgeon removes the along with a margin of healthy tissue. A metal rod or plate (called an endoprosthesis) is used to support the bone. If a knee or other joint is removed, an artificial joint will be placed to help you regain use of the arm or leg.

· Chemotherapy is often given both before and after surgery.
- Before surgery, can help shrink the tumor and kill cancer cells that may have traveled to other parts of the body & to determine the effectiveness of a given regimen of chemotherapy.
- After surgery, chemo is repeated to help reduce the risk of the cancer coming back. Treatment may continue for up to a year.

· Rehabilitation after limb-sparing surgery is intense and long-term. It often takes six to 12 months of rehab to regain strength and movement in the affected limb.

· A doctor may recommend amputation if:
- The tumor is large and doesn't respond to chemotherapy
- The tumor has grown into the skin near the bone
- The tumor surrounds important nerves or blood vessels
- The patient is a very young child

· People who have limb-sparing surgery usually need several more surgeries throughout their lives. A person who's had an amputation probably won't need any more surgery.

· There are four phases of limb-sparing procedures:
- 1. Preoperative planning.
- 2. Resection of tumor.
- 3. Reconstruction of bone.
- 4. Reconstruction of soft tissue.

· Resection should follow strictly the principles of oncologic surgery. The Enneking's treatment recommendations when surgery alone is to be used for local control of a lesion is valid and generally accepted.

Bone secondaries

Osteolytic Osteosclerotic
Bone destruction then pathological fracture increasing density at the site of metastasis
Bronchogenic carcinoma
Thyroid follicular carcinoma
Kidney (hypernephroma)
Breast carcinoma

Prostatic carcinoma
Some breast carcinoma
.More common than primary tumors. Occurs in bones that contain red Marrow e.g. vertebrae, ribs and pelvis.
· The most commonly affected bones are the spine, pelvis, ribs and femur.

Plain X-ray

· *Osteolytic*: area of destruction without bone formation with ill-defined margins (mouth-eaten).
· *Osteoplastic*: area of sclerosis

MRI
CT (local & chest)
Bone scintigraphy

Management of patients with bone metastases

· The presence of bone metastases usually indicates the start of widespread tumor dissemination and treatment is mostly palliative, i.e. not curative.
· The aim of orthopedic management is to relieve symptoms, restore function and ensure that the patient has received appropriate supporting treatment from an oncologist.
· If a metastasis is detected prior to fracture, then the patient will usually receive oncological treatment depending on the primary (hormonal therapy and/or chemotherapy for some tumors, radiotherapy and bisphosphonates for most). Prophylactic fixation is indicated for persistent pain, particularly in a weight bearing bone accompanied by destruction of the cortex.
· If the bone has already fractured or if prophylactic fixation is used, it is essential to ensure that the fixation method used is sufficient to allow immediate restoration of function and should outlive the patient. More than half of pathological fractures will not heal and so whatever fixation is used, this must be taken into account. The use of joint replacements or custom prostheses is indicated for replacement of large areas of destroyed bone at bone ends. If