

Chapter 12

Tumours

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1) OSTEOSARCOMA:

Bone tumours are basically classified as benign and malignant lesions. Benign lesions are quite common and primary malignant are rare. Most classifications of bone tumours are based on the dominant tissue involved in lesions. This may help in both diagnosis and treatment.

The metastatic tumours are the commonest bone tumours, although they are not of true bone tumours means not of mesenchymal origin.

It is a highly malignant tumour of bone arising within bone (intramedullary) and spreads outwards to periosteum and surrounding soft tissues. It may affect any bone, but long bone metaphysis are most commonly involved around knee and the proximal end of humerus. It mainly occurs in children and adolescents.

C/F

Pain which is constant and worse at night and gradually increases in severity is the usually first symptom in these patients. In early cases, patient may present with lump and pain. There will be local tenderness. In later cases, overlying tissues may be swollen and inflamed with a palpable mass.

ESR and serum alkaline phosphatase are usually raised in these patients.

Imaging:

Plane X-rays are the most useful of all imaging techniques. They shows any obvious bony abnormality like cortical thickening, a lump or a cyst, or any ill defined lesion. If boundary of a cyst is sharply defined, it is probably a benign,

and if hazy and diffuse it suggests invasive tumours. Stippled calcification in a cyst is a characteristic of cartilage tumours. Periosteal new bone formation and extension of tumour into soft tissues are suggestive of its malignant nature.

In osteosarcoma, there are alternate hazy osteolytic and dense osteoblastic areas. Usually cortex is breached with poorly defined endosteal margins and tumour extends into surrounding soft tissues. This extension into soft tissues with streak of new bone formation radiating from cortex to outwards- is called as "Sunburst appearance". A reactive new bone formation at angles of periosteal elevation is called as "Codman's triangle". Both are typical for osteosarcoma. Diagnosis is made with certainty on typical X-rays appearance. However, other imaging studies are essential for staging purposes to define a probable prognosis. CT and MRI shows the extent of tumour. Chest X-ray, preferably Pulmonary CT should be done to detect metastasis. A biopsy is indicated before starting a treatment in these patients.

Pathology: This tumour located in metaphysis of long bones. It destroys and replaces the normal bone. Tumour extends intramedullary and across the physeal plate. There is alternate bone loss and abnormal new bone formation. Tumour extends into soft tissues with ossification at periosteal margins. There are streaks of new bone extending to extraosseous tissues producing sunburst appearance.

Histologically, there are characteristic spindle cells with pink staining osteoid matrix. In some they contain cartilage cells or fibroblastic tissues with little or no osteoid.

Principles of Treatment:

Once clinical and radiological diagnosis is made, a multidisciplinary approach is of paramount importance.

- 1) **Benign but asymptomatic:** These lesions never required treatment, if diagnosis is beyond doubt. However, if appearance is non pathognomic, the biopsy is indicated.
- 2) **Benign but Symptomatic or Enlarging:** These lesions require biopsy to confirm the diagnosis. These can be removed by local (marginal) excision or by curettage.

3) **Suspected Malignant tumours:** If lesion is of primarily malignant, then a thorough detailed diagnostic evaluation is required. A surgical staging is established. The various treatment options can be discussed with patients and after all pros and cons, a choice is to be made between amputation, limb sparing operations and adjuvant therapy.

Surgical staging: Bone sarcomas are broadly divided into following stages :

* **Stage -1** All low grade sarcomas

***Stage -2** Histologically high grade

***Stage-3** Sarcomas with metastasis

Enneking has described surgical staging to know prognosis of these tumours: a) how the tumour behaves and b) how far it has spread.

Methods of Treatment:

1) **Excision:** Tumour excision depends upon its aggressiveness and local spread.

a) Intracapsular excision and curettage are incomplete procedures. These are indicated in benign lesions with very low risk of recurrences. Bone cementing after curettage decreases the risk of local recurrence.

b) Marginal excision: It is just only beyond the tumour margins. It is suitable for benign lesions.

c) Wide excision: In this lesion excised is carried through normal tissues. This can be done in conjunction with chemotherapy for grade II A lesions.

d) Radical excision: In this entire compartment in which tumour lies is removed En bloc without exposing the lesion.

2) LIMB SALVAGE:

The dreadful prognosis that was formerly considered in these patients, has been markedly improved as result of better diagnostic and staging procedures. These methods have made limb salvage as the treatment of choice for many

patients. Advanced surgical procedures like endoprosthetic replacement or bone grafting are available these days. In these cases, the first step is wide excision of tumour while preserving neurovascular bundles. The small defect produced during this is filled with vascularized or non vascularized bone graft. The longer defect may require custom made implants. In growing children, extensible implants have been used to avoid repeated procedures.

These are associated with various complications.

2) AMPUTATION:

Amputation may be curative in cases which are resistant to chemotherapy and radiation therapy.

3) MULTI AGENT CHEMOTHERAPY

Neoadjuvant and adjuvant are now considered as preferred for malignant bone and soft tissue tumours. Modern chemotherapy regimens effectively decrease the size of primary tumour, prevent metastatic seedings and increased survival ship.

Doxorubicin, methotrexate, cyclophosphamide, vincristine and cis-platinum are commonly used agents in chemotherapy. Treatment is started 8-120 weeks preoperatively. Chemotherapy is considered as successful, if 90% of resected tumour tissues shows necrosis.

4) RADIOTHERAPY

It gives an alternative to amputation in radiosensitive tumours. Adjuvant treatment is used in inoperable lesions, in high grade tumours, inaccessible sites, proximity to vessels and for marrow cell tumours.

Post-irradiation spindle cell carcinoma and pathological fractures are the main complications associated with radiotherapy.

2) CHONDROSARCOMA:

These are slow growing and one of the commonest malignant tumours of bone. These are seen in fourth and fifth decades

having male dominance. Patients complain of dull aching pain or a gradually enlarging lump mostly in lower limbs. Pelvis and ribs are next common sites.

Chondrosarcomas fall into basically two categories: a) centrally tumours occupying medullary cavity of bone, and b) peripheral tumours growing from cortex. Juxtacortical chondrosarcoma, clear cell chondrosarcoma, and mesenchymal chondrosarcoma are least common varieties.

1) **Central chondrosarcoma:** These involves medullary cavity of tubular or flat bones, most commonly at the proximal end of femur. There may be expanded radiolucent area on bone with flecks of increased density. These lesions may have scalloping appearance or destruction of cortex.

2) **Peripheral chondrosarcoma:** These arises in cartilage cap of an exostosis, (osteochondroma) which are present since childhood. X rays shows bony exostosis, surmounted by patchy calcifications. A tumour which is very large and have fluffy calcifications with poorly outlined are suspicious features. MRI is the best investigation to detect it's size and internal features of cartilage cap.

3) **Juxta cortical chondrosarcoma:** It arises from outermost layer of the cortex of bone, usually femur. X- rays shows flecks of calcification, sunray streaks and new bone formation at the margins of the stripped periosteum.

These are slow growing tumours and metastasize late, so these are ideal case for wide excision and prosthetic replacement.

Prognosis is determined largely by the cellular grade and resection margins.

1) **OSTEOCHONDROMA (EXOTOSIS):**

This is one of the commonest benign tumour of bone characterized as lump, mostly on the fast growing ends of long bones and crest of iliac bones. It starts as a small overgrowth of cartilage at the edge of physal plate and develops by endochondral ossification into bony protuberance covered by cartilage cap. Patient is usually teenager or a young adult

having pain due to an overlying bursa or impingement of soft tissues. It stops growing after the arrest of normal adult growth. Any change or enlargement after the end of normal growth period suggests malignant transformation of lesion. On X-rays, there will be a well defined exostosis emerging from metaphysis with base on parent bone. It looks smaller than it feels, because the cartilage cap is invisible on X-rays.

Multiple lesions may develop in a hereditary disorder called hereditary multiple exostosis.

Pathologically, a cartilage cap is surmounted on a narrow base or pedicle of bone. The cartilage cap consists of simple hyaline cartilage. Larger lesions may have cauliflower appearance, with degeneration and calcification in centre of cartilage cap.

Complications: Malignant transformation is difficult to assess in these cases. However, 1) enlargement of cartilage cap 2) a bulky cartilage cap (more than 1cm in thickness) 3) irregular scattered flecks calcifications and spread into surrounding tissues are suggestive of malignant change. MRI can be done in these patients to rule out malignant changes.

Treatment: All symptomatic lesions should be excised.

2) GIANT CELL TUMOUR (GCT):

This tumour of uncertain origin usually appears in mature bones. Distal femur, proximal tibia, proximal humerus, distal radius are most commonest sites involved. It doesn't appear before the closure of epiphysis and it is characterized by its extend up to subarticular bone plate. Clinically, a young patient usually complains of pain at the end of long bones. A history of trauma is not uncommon. Pathological fracture is seen in 10-15% of these patients. These patients may have "egg shell crackling" on examination.

On X-rays, there will be an eccentrically situated radiolucent area at the end of long bones and bounded by subchondral bone plate. The cortex is thin and ballooned producing "soap bubble appearance" of lesion. It has to be differentiated from brown tumour of hyperparathyroidism. MRI and CT scans tell us about extent of tumour. Biopsy is essential to rule out malignant change.

The tumour has a reddish, fleshy gross appearance. Histologically, there are abundance of multinucleated giant cells on back ground of stromal cells with little or no visible intercellular tissues.

Treatment:

It consists of thorough curettage and striping of cavity with burr and gouges, followed by swanning with hydrogen peroxide or application of liquid nitrogen and packing of cavity with bone chips. This tumour has the potential to transform into osteosarcoma.

3) OSTEIOD OSTEOMA:

This is a tiny bone tumour having less than 1 cm in diameter and occurs usually under 30years of age with male dominance. All bones are involved except skull. The patient complains of persistent well localized pain and typically relieved by salicylates. Small radiolucent area called nidus is typical feature on X-rays. Long bone diaphysis shows dense sclerosis and cortical thickening. It has to be differentiated from Brodie's abscess.

It appears dark brown or reddish nucleus surrounded by dense bone with central area having unorganized sheets of osteoid tissue and bone cells.

Treatment:

Complete removal or destruction by radio ablation is only effective treatment.

4) SIMPLE BONE CYST:

This benign tumour is also called a solitary cyst or unicameral bone cyst. It is seen in childhood, typically in metaphysis of long bones most commonly in proximal humerus or femur. It heals spontaneously and seldomly seen in adults. On X-rays, there will be a well demarcated radiolucent area in the metaphysis. It extends upto physeal plate, cortex may be thinned with bone expanded.

Asymptomatic lesions in older children can be left alone. Active cysts should be treated with aspiration of fluid and injection of 80-160 mg of Methylprednisolone or autogenous bone marrow.

5) ANEURYSMAL BONE CYST:

This benign expansile tumour occurs in any age and in any bone is involved. But commonly seen in young adults in long bone metaphysis areas. It is a expanding lesion and patient complains of pain and swelling on affected part. X-rays shows a well defined radiolucent cyst which is often trabeculated and eccentrically placed in bone. It doesn't extend up to the articular margin. It has soap bubble appearance on X-rays.

It contains clotted blood on opening it. Histologically, it consists of fibrous tissue with vascular spaces, deposits of hemosiderin and multinucleated giant cells. Malignant transformation doesn't occur.

Treatment consists of thorough curettage and bone grafting. Packing with bone cement (methylmethacrylate) can be done.