Bone Tumors

When cells divide abnormally and uncontrollably, they can form a mass or lump, this lump is called a tumor. As the tumor grows, abnormal tissue can displace healthy tissue. It is either be benign or malignant, which is subdivided into primary and secondary. Primary neoplasms of the skeleton are rare, amounting to only 0.2% of the overall human tumor burden. However, children are frequently affected.

CAUSES:

The causes of bone tumors are unknown. A few possible causes are *genetics*, *radiation*, and *injuries* to the bones. Osteosarcoma has been linked to radiation treatment (particularly high doses of radiation) and other *anticancer drugs*, especially in children. However, a direct cause hasn't been identified.

SYMPTOMS:

A *dull ache* in the affected bone is the most common symptom of bone cancer. The pain starts off as occasional and then becomes severe and constant. The pain may be severe enough to wake the patient up in the night.

Sometimes, when people have an undiscovered bone tumor, what seems like an insignificant injury breaks the already weakened bone, leading to severe pain, this is known as a *pathologic fracture*. Sometimes there may be *swelling* at the site of the tumor and the patient might not have any pain. *Loss of appetite* and *weight loss* are dangerous signs of malignant type of tumor.Tumors can also cause *night sweats*, *fever*, or both.

People with benign tumors might not have any symptoms. The tumor might not be detected until an imaging scan reveals it while receiving other medical testing (*incidentally* found).

DIAGNOSIS :

It is important to exclude *Fractures*, *infections*, and other conditions which might resemble tumors. To be sure it is a bone tumor, variety of tests may be ordered. History and physical exam focused on the suspected area to start with followed by non-invasive as well as invasive investigations if needed.

Laboratory investigations:

Tests, including blood or urine samples(CBP, ESR, L.F.T, R.F.T, urine for; Bence-Jones protein and plasma M protein in Multiple myeloma). A lab will analyze these fluids to detect different proteins that may indicate the presence of a tumor or other medical problems.

An alkaline phosphatase test is one common tool use to diagnose bone tumors. When bone tissue is especially active in forming cells, large quantities of this enzyme show up in the blood. This could be because a bone is growing, such as in young people, or it could mean a tumor is producing abnormal bone tissue. This test is more reliable in people who've stopped growing.

Imaging tests:

- X-rays to determine the size and exact location of the tumor. Depending on the X-ray results, these other imaging tests may be necessary:
- CT scan is a series of detailed X-rays that are taken from several angles to figure out the outlines of the lump and its exact site and size.
- MRI scan provide detailed pictures of the area in question with the extension to the surrounding soft tissu .
- Positron emission tomography (PET) scan, by injecting of a small amount of radioactive sugar I.V. since cancer cells use more glucose than regular cells, this activity helps to locate the site of the tumor and skip lesions.

- Arteriogram is a contrast X-ray of the arteries and veins may be required.
- Abdominal and pelvic U/S, pulmonary C-T scan, may be needed to detect the spread of tumor in cases of metastasis to chest or abdominal and pelvic organs.

Biopsies:

In this test, a sample of the tissue that makes up the tumor will be removed for histopathological study. The main types of biopsies are a needle biopsy (FNA) and true cut biopsy including an incisional or an excisional biopsy, it is important to make a definite diagnosis of the condition.

Staging and grading of tumour:

Tumor can be spread by blood steam or rarely lymphatics as well as direct extension especially if the tumor is malignant and of high grade.

Benign tumor: graded as Latent, active and aggressive.

Malignant tumor: staging is to determine the anatomical extent of the tumor.

Intra-compartmental tumor is called A

Extra-compartmental tumor is called B

Stage I : Low grade

Stage II :High grade

Stage III : Metastasis whether low or high grade.

STAGING SYSTEMS

Musculo Skeletal Tumor Society Classification

Stage	*Grade*	*Site*
IA	Low (G1)	Intracompartmental (T1)
IB	Low (G1)	Extracompartmental (T2)
IIA	High (G2)	Intracompartmental (T1)
IIB	High (G2)	Extracompartmental (T2)
III	Any G Regional or distant metastasis (M1)	Any (T)

Enneking et al

Stage IA (G1, T1, M0): Low-grade intracompartmental lesion, without metastasis
Stage IB (G1, T2, M0): Low-grade extracompartmental lesion, without metastasis
Stage IIA (G2, T1, M0): High-grade intracompartmental lesion, without metastasis
Stage IIB (G2, T2, M0): High-grade extracompartmental lesion, without metastasis
Stage IIIA (G1 or G2, T1, M1): Intracompartmental lesion, any grade, with metastasis
Stage IIIB (G1 or G2, T2, M1): Extracompartmental lesion, any grade, with metastasis

TREATMENT FOR BENIGN TUMORS :

If the tumor is benign, it may or may not require action. Sometimes doctors just keep an eye on benign bone tumors to see if they change over time. This requires coming back periodically for follow-up X-rays.

Bone tumors can grow, stay the same, or eventually disappear. Children have a higher likelihood of having their bone tumors disappear as they mature. Benign tumors can sometimes spread or transform into malignant tumors and can also lead to a pathological fractures, for these reasons complete excision or curettage with bone graft maybe needed.

TREATMENT FOR MALIGNANT TUMORS:

The treatment will depend on what type of bone cancer we have and whether it's spread or not. If the cancer cells are confined to the tumor and its immediate area, this is called the localized stage. In the metastatic stage, cancerous cells have already spread to other parts of the body, this makes curing the cancer more difficult.

Surgery, radiation, and chemotherapy are the main strategies for treating cancer.

Surgery:

Malignant bone cancer is usually treated with surgery. In surgery, the entire tumor is removed. The surgeon carefully examines the margins of the tumor to make sure no cancer cells are left after surgery.

Surgical techniques have improved greatly including excisional and radical resection depending on the grade and stage of tumor. Nowadays are much more likely able to spare the limbs by reconstructive surgery to retain as much limb function as possible. If bone cancer is in an arm or leg, a surgery may be done known as limb salvage surgery. This means that while the cancerous cells are removed, the tendons, muscles, blood vessels, and nerves are spared. The surgeon will replace the cancerous bone with a metal implant but if the tumor is not excisable or widely spread to the surrounding soft tissues amputation maybe needed.
Advances in chemotherapy have greatly improved recovery and survival.

Radiation therapy:

Radiation is often used in conjunction with surgery. High-dose X-rays are used to shrink tumors before surgery and kill cancer cells. Radiation can also reduce pain and decrease the chance of bone fractures.

Chemotherapy:

If the cancer cells are likely to spread or if they already have, chemotherapy may be recommended. Chemotherapy uses anticancer drugs to kill the rapidly growing cancer cells.

Cryosurgery:

This treatment involves killing cancer cells by freezing them with liquid

nitrogen. A hollow tube is inserted into the tumor, and liquid nitrogen or argon gas is pumped in. In some cases, cryosurgery can be used to treat bone tumors instead of regular surgery.

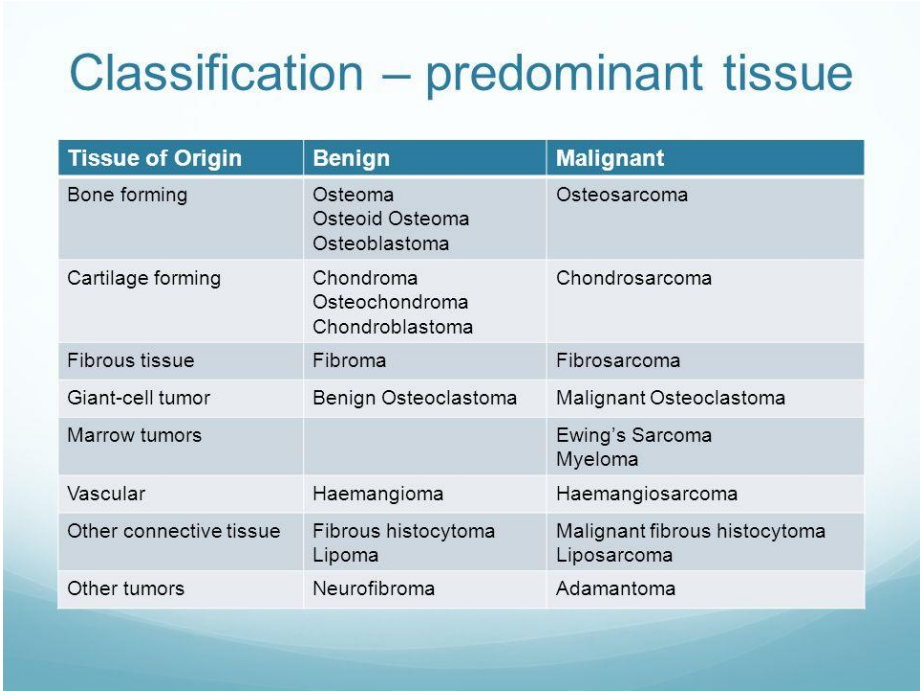
Prognosis:

Follow-up X-rays and blood tests will be necessary to make sure the whole tumor is gone and that it doesn't recure and follow-up tests every few months maybe required.

Prognosis will depend on what type of bone tumor, how big it was, and where it was located as well as its grade and stage. If the tumor is benign, the long-term outcome will probably be good. However, benign bone tumors can grow, recur, or turn into cancer, so this can be discover by regular checkups.

Classification:

Bone tumor can be classified whether benign of malignant according to the type of tissue that it is originated from.



Benign tumors

Benign bone tumors typically stay in place and are unlikely to be fatal, they are still abnormal cells and may require treatment. They can grow and could compress healthy bone tissue and cause future symptoms. They are more common than malignant ones.

Osteoid osteoma:

Benign tumor of young adult <30 year, male predominant, affecting all bones except skull, painful lesion (in spite of very small size) characteristically respond to Aspirin, spinal lesion gives scoliosis, no risk of malignancy.
X-ray: Small radiolucent area (nidus) in the shaft with extensive periosteal reaction and new bone formation, with cortical thickening and obliteration of medullary cavity. The nidus may be only visible by CT-scan. Metaphyseal lesion with less cortical thickening gives picture similar to Brodies abscess
Treatmen: Excision and bone graft

Osteoblastoma: spine, giant osteoid osteoma, risk of malignant changes.
Compact (ivory) osteoma….in the skull

Osteochondromas(cartilage-capped exostosis):

The most common type of benign bone tumor is an osteochondroma. This type accounts for between 35 and 40 percent of all benign bone tumors. They are develop in adolescents and teenagers.

These tumors form near the actively growing ends of long bones, such as arm or leg bones. Specifically, affect the lower end of the femur, the upper end of tibia, and the upper end humerus.

These tumors are made of bone and cartilage. Osteochondromas have been considered to be an abnormality of growth. A child may develop a single osteochondroma or many of them.

Non-ossifying fibroma (Fibrous cortical defect):

Non-ossifying fibroma is a simple solitary bone cyst. It's the only true cyst of bone. It's usually found in the leg and occurs most often in children and adolescents.

Giant cell tumour:

Represent 5% of all primary bone tumours, disease of mature bone, mainly at the end of femur, proximal tibia, distal radius and proximal humerus.
C/F: Pain, swelling at bone ends and pathological fracture in 10-15%
x-ray: Radiolucent area eccentrically located at bone end pointing to the sub-articullar area, either well-defined or ill-defined according to its aggressiveness, its center contain ridges of surrounding bone giving it soap-bubble appearance, with thinning and ballooning of the cortex, and soft tissues extension sometimes.
CT-scan and MRI are needed for staging and needle or incisional biopsy are needed for grading
Treatment:

- Benign slow growing….curettage and bone graft.
- Benign aggressive and local recurrent…excision &bone graft &/or prosthesis.
- Malignant giant cell tumour….radical resection

Enchondroma

An enchondroma is a cartilage cyst that grows inside the bone marrow. When they occur, they begin in children and persist as adults. Malignant changes occur in 2% of solitary lesion and in 10% 0f multiple lesion (Olliers disease) and in 30% if associated with hemangiomas (Muffuccis syndrome). Enchondromas occur in the hands and feet as well as the long bones of the arm and thigh.

Chondroblastoma: well demarcated radiolucent area in the epiphysis

Chondromyxoid fibroma: is an extremely rare benign cartilaginous neoplasm which accounts for < 1% bone tumours.

Typical x-ray: Ovoid radiolucent area in the metaphysis of long bones eccentrically placed, the endosteal margin scalloped and bound by dense bone extending as tongue towards diaphysis.

Fibrous dysplasia:

Developmental lesion, tumor contains fibrous tissue, osteoid and woven bone, affecting metaphysis of proximal femur and tibia, either in one bone (monostotic) or multiple (polystotic) leads to deformity
X-ray: Radiolucent lesion as ground-glass appearance as femoral deformity as *shepherds crook*, there is risk of malignancy in polystotic.
Treatment: If large curettage and bone graft.

Simple bone cyst:

Also called solitary or unicameral bone cyst seen in children rare in adult, mainly in the proximal humerus and femur, usually symptomless accidentally diagnosed by x-ray and sometimes pathological fracture.
X-ray: well demarcated radiolucent area in the metaphysis with thinning of the cortex and bone expansion ,sometimes extended to the epiphysis.
D.Diagnosis: fibrous cortical defect, fibrous dysplasia and enchondroma. Aspiration shows straw colored fluid,

Treatment

Asymptomatic…..follow-up , Active lesion….aspiration and methyl prednisolone injection or bone graft, Pathological fracture.....curettage + bone graft + internal fixation.

Aneurysmal bone cyst

An aneurysmal bone cyst is an abnormality of blood vessels that begins in the bone marrow. It can grow rapidly and can be particularly destructive because it affects growth plates.

malignant bone tumors

Primary bone cancer means that the cancer originated in the bones, they account for less than 1 % of all types of cancer.
The three most common forms of primary bone cancers are osteosarcoma, Ewing sarcoma family of tumors, and chondrosarcoma.

Osteosarcoma:

Commonest primary malignant bone tumor in exception of myeloma, affect children and adolescents, in the long bones around knee and proximal humerus

C/F: Painful bony lump with localized tenderness, pathological fractures are rare. High ESR and alkaline phosphatase .
X-ray: Combined osteolytic and osteoblastic areas, with cortical erosion and ill-defined edges and soft tissue invasion gives *sunburst appearance*. Reactive new bone formation at the angles of periosteal elevations leads to *Codman's triangle*.
D.dx: post-traumatic swelling, stress #, aggressive cyst and infection
Dx: x-ray, CT-scan and MRI and isotope scan for staging and to detect any pulmonary metastases (in 10% at presentation)
Incisional biopsy for diagnosis and grading .
Treatment:
 Preoperative chemotherapy (for 10-12weeks) followed by wide local resection + bone graft or prosthesis.
 Radical resection=amputation through proximal joint

Prognosis: 5years survival rate 50%.
Variants: parosteal, periosteal, and Paget's sarcomas.

Ewing's sarcoma

Originate from bone marrow endothelial cells, age incidence 10-20 years old, in the long bones(tibia and fibula) and flat bones like clavicle, patient presented with pain, fever, localized swelling, tenderness and warmth.
D.dx :osteomyelitis.
X-ray: diaphyseal lesion with bony destruction and new bone formation due to excessive periosteal reaction giving to *onion-peel appearance*, *sun-ray* and *Codman's triangle* are common.
C-T scan and MRI and isotope scan used for staging

Treatment :
Poor prognosis even with treatment and the radio sensitivity does not improve survival; however chemotherapy does improve 5 years survival to 50%.
 Preoperative chemotherapy followed by wide-excision and postoperative chemotherapy.
 Preoperative radiotherapy followed by excision and chemotherapy
 Proximal amputation

Chondrosarcoma:

One of commonest primary malignant bone tumors, in the 4th -5th decades, occur in male more than female, slow growing tumor,50% in the long bones of the lower limbs and in pelvis.
Primary (central) chondrosarcoma….in the medullary cavity
Secondary (peripheral)…occur in exostosis

X-ray: radiolucent lesion contains flecks of calcifications
Staging needs MRI & CT-scan before biopsy.

Treatment:
slow growing tumor…wide excision and prosthesis and even sometimes amputation. Isolated pulmonary secondaries needs excision.
Chondrosarcoma usually poorly responded to chemo. Or radiotherapy

Secondary bone cancer:

The term “secondary bone cancer” means that the cancer started somewhere else in the body and then spread to the bone. It usually affects older adults. The types of cancer most likely to spread to the bones are: kidney, breast, prostate, lung, thyroid gland.

Multiple myeloma:

The most common type of secondary bone cancer is called multiple myeloma. This bone cancer shows up as tumors in the bone marrow. Multiple myeloma most commonly affects older adults.