MSS

Musculoskeletal System

Doctor 2019 | Medicine | JU

5

Pathology

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Lecture 5, we have finished half of the material lol!

BONE TUMORS AND TUMOR LIKE CONDITIONS:

These 6 points are important, they give us the concepts which we have understand:

- **Primary bone tumors are rare:** if you are going to compare for osteosarcoma with breast cancer, lung carcinoma, you will find it less common comparing them.
- **Benign >>>> malignant tumors:** the chance of having benign is much common.
- **First 3 decades mostly have benign lesions; adults more to be malignant**
- **Trx: aims to optimize survival while maintaining function:** the function of bones and joints is extremely important.

  For instance if you have a guy with proximal chondrosarcoma of the femur, and the head or the neck of the femur, you stage it and if it localized you can just go and remove that part, put prosthesis, the patient walks in one week after surgery, it's called limb salvage resection of the tumor
- **Age & location help narrow ddx:** a classical pathology training and teaching never make the diagnosis on a bone biopsy without getting detailed history evaluating the imaging of X-Ray, MRI…etc
- **S&S: asymptomatic, pain, path #:** if the lesions is big and involves an area which is very active, sometimes there will be a fracture due to this tumor which is called pathological fracture.

COMMON BONE TUMORS TABLE:

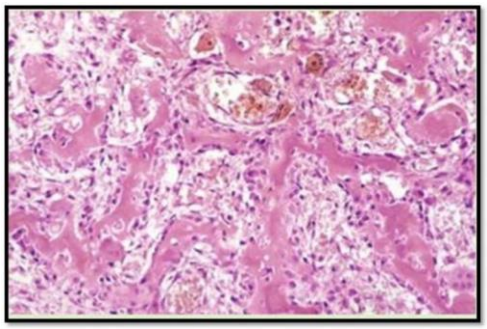
Category	Behavior	Tumor Type	Common Locations	Age (yr)	Morphology
Cartilage forming	Benign	Osteochondroma	Metaphysis of long bones	10–30	Bony excrescence with cartilage cap
—	—	Chondroma	Small bones of hands and feet	30–50	Circumscribed hyaline cartilage nodule in medulla
—	Malignant	Chondrosarcoma (conventional)	Pelvis, shoulder	40–60	Extends from medulla through cortex into soft tissue, chondrocytes with increased cellularity and atypia
Bone forming	Benign	Osteoid osteoma	Metaphysis of long bones	10–20	Cortical, interlacing microtrabeculae of woven bone
—	—	Osteoblastoma	Vertebral column	10–20	Posterior elements of vertebra, histology similar to osteoid osteoma
—	Malignant	Osteosarcoma	Metaphysis of distal femur, proximal tibia	10–20	Extends from medulla to lift periosteum, malignant cells producing woven bone
Unknown origin	Benign	Giant cell tumor	Epiphysis of long bones	20–40	Destroys medulla and cortex, sheets of osteoclasts
—	—	Aneurysmal bone cyst	Proximal tibia, distal femur, vertebra	10–20	Vertebral body, hemorrhagic spaces separated by cellular, fibrous septae
—	Malignant	Ewing sarcoma	Diaphysis of long bones	10–20	Sheets of primitive small round cells

BONE-FORMING TUMORS

-General features for:

Osteoid osteoma (more common)	Osteoblastoma
< 2 cm	**> 2 cm**
Young men	**Posterior vertebrae; no rim of bone reaction by radiology**
Femur & tibia; nidus with surrounding bone reaction	**Pain unresponsive to aspirin**
Severe nocturnal pain (PGE2) relieved by aspirin & NSAIDS	**Treated by curetting**
Treated by: radiofrequency ablation or surgery	

- We can compare them by radiology.
- Once the osteoid osteoma is diagnosed either the patient will live with it and just whenever he has pain, he will take aspirin or NSAIDs or treated by radiofrequency ablation or surgery.



Histologically, both of them look like reactive bone with some hemorrhage and reactive giant cells, you don't see atypia.

OSTEOSARCOMA

- Malignant osteogenic tumor
- Excluding hematopoietic malignancies; it is the most common primary malignant tumor of bone
- 75% adolescents between (10-20) ; another peak in older (55-65) (secondary osteosarcoma, because they occur on top of a predisposing condition like Puget disease)
- Males > females (1.6:1.0)
- Location: Metaphysis of long bones (distal femur & proximal tibia)

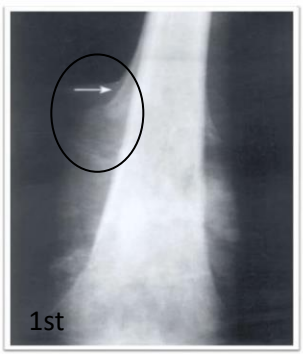
How do they patients present?

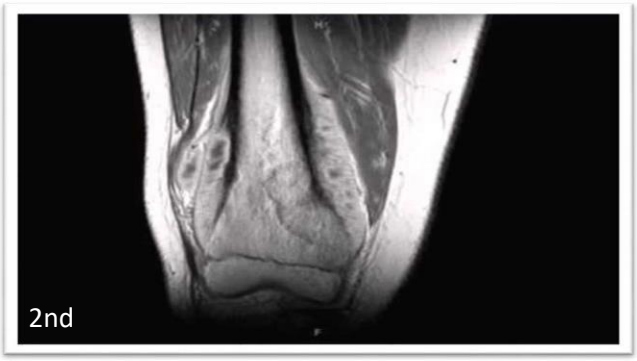
- Progressive pain or pathologic fracture
- Imaging: large destructive and infiltrative lesions with Codman triangle: which means they start in the bone and they go deep into the medulla and outside into the periosteum, when they go into the periosteum, infiltration of the periosteum and the surrounding soft tissue, they elevate the periosteum forming an angle which is called Codman triangle.

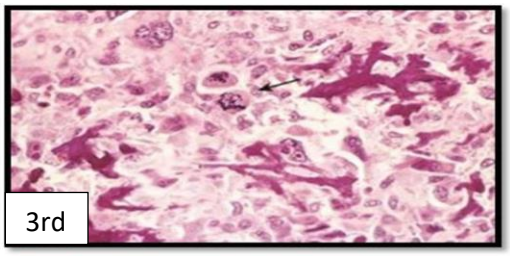
*Codman triangle is not specific for osteosarcomas, we can see it in any other infiltrative bone lesion whether it's infectious or other tumors like ewing sarcoma

- Genetic abnormalities: mutations in RB gene, TP53 gene, CDKN2A (p16 & p14), MDM2 & CDK2

OSTEOSARCOMA FEATURES:







1st : simple x-ray, distal femur showing a process which started in medulla of the bone and infiltrate the surrounding tissue elevating the periosteum (→) and the angle between actual bone and the periosteum is called Codman's triangle.

2nd: MRI showed you the processes, tumor arises at the metaphysis and extends to the soft tissue, the skeletal muscle infiltrated by this tumor, and the periosteal elevation causing Codman triangle. (back to video)

3rd: histologically: we saw malignant osteoid haphazard abnormal mitosis, atypia of the osteoblast with frequent abnormal mitosis again.

4th: the gross specimen, we cut it longitudinally and articular cartilage of the distal femur and below it a tumor area.

OSTEOSARCOMA TREATMENT:

What is the current modality for osteosarcoma treatment, there was progress in the management and the survival of those patients in the old days, that was very bad prognosis nowadays patients live 5 year.

- **Multimodality approach (MDTeam)**
- **1. Neoadjuvant chemotherapy 2. Surgery 3. Chemotherapy and radiation**

  Chemotherapy → to prevent or to kill any metastatic possibility

  Radiation → to control the local diseases

  Surgery → to the bulk or remove the tumor
- **Hematogenous spread to lungs:** through bloodstream, and the most common location is to the lungs. There's some exception, they don't usually go to the lymph nodes.
- **5 year survival reaches 60-70%:** probably, it's improving now 75-80%.
- **Presence of metastasis at diagnosis is a bad prognostic factor:** if the previous patient we talked about when he came and he was stage and multiple lung lesions, this is a bad sign and the 5 year survival will be dropped, so early diagnosis and early management are extremely important.

CARTILAGE-FORMING TUMORS:

- Osteochondroma (most common - benign exostoses):benign cartilaginous tumor composed of benign bone covered by benign cartilage, and it's exophytic from long bones, solitary (85%); part of multiple hereditary exostoses (MHE):characterized by EXT1, EXT2 gene mutations
- Rare (<3-5%) transformation to chondrosarcoma, if they do, usually they do in cases when they are MHE.
- If you diagnose a patient with MHE, you have to follow up closely to watch those osteochondromas to avoid transforming to malignant chondrosarcoma

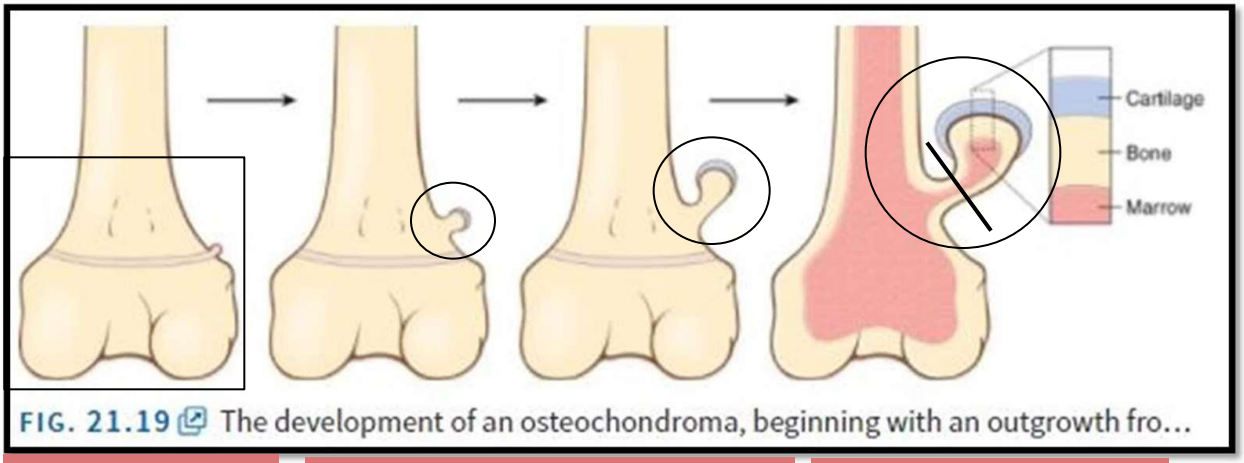


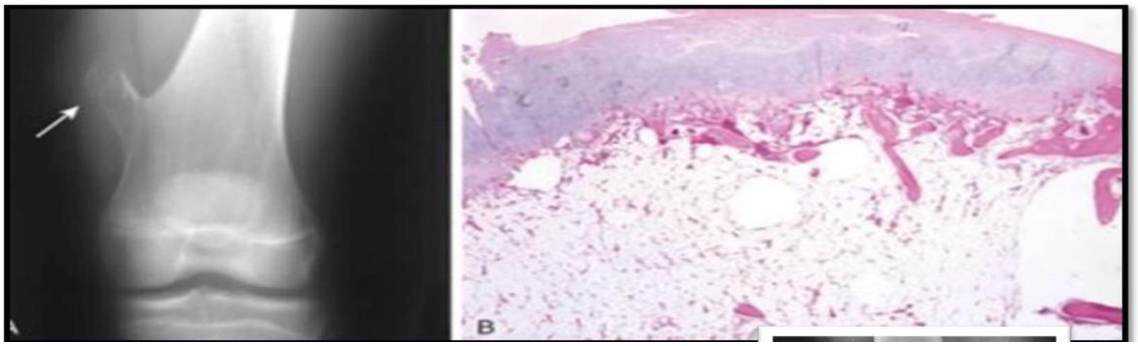
FIG. 21.19 The development of an osteochondroma, beginning with an outgrowth fro…

Most common location, around the cartilaginous plate

Extension of this bone and bone marrow goes out

Covering by benign cartilage, so if you cut there, what you will see, normal cartilage, normal periosteum, normal marrow

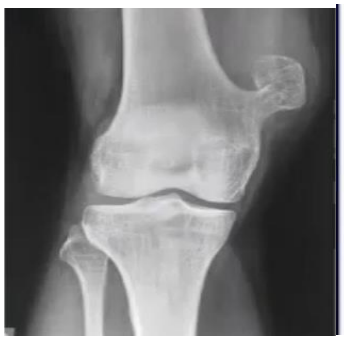
OSTEOCHONDROMA:

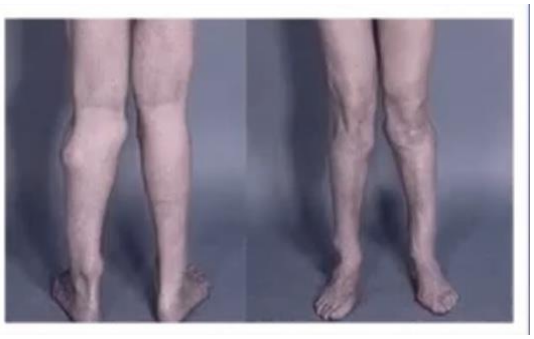


1st: X-Ray of the distal femur, very classic appearing exophatic pedunculated mass with normal cartilaginous cap, you don't see destruction of the tissue around it , infiltration and Codman's triangle.

2nd: when the surgeon cut it and give it to pathologist, and he sees normal cartilage, normal subchondral bone and bone marrow, this is all benign appearing as if he is looking at a normal articular piece of bone.

They cause pain and pathological fractures

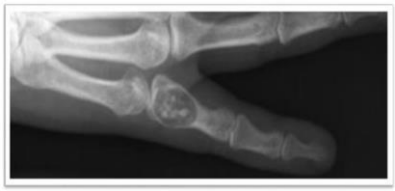


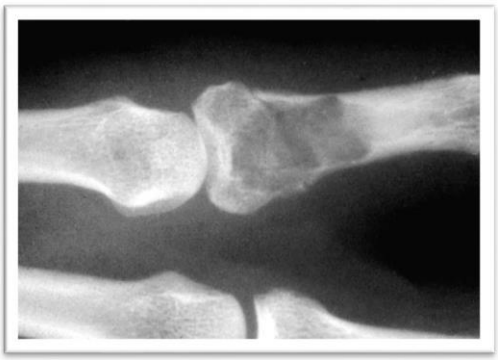


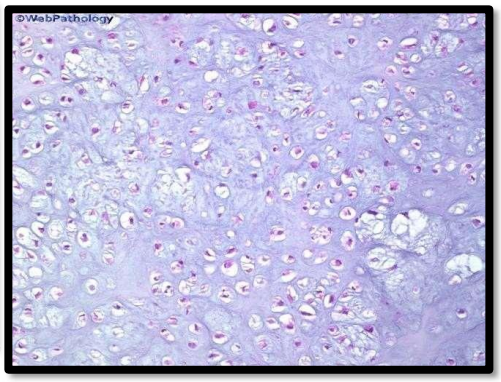
This is a case of MHE

CHONDROMA (ENCHONDROMA):

- Benign hyaline cartilage tumors in bones with endochondral origin; medullary enchondroma or cortical chondroma
- Most time it's solitary,occurring in the metaphyseal lesions, most common location: the hands and the feet; 20-50 years
- ***Multiple enchondromas***: Ollier disease
- ***Maffucci syndrome***: multiple enchondromas + skin hemangiomatosis
- IDH1 & IDH2 gene mutations







The radiologist will tell the surgeon, that is chondroma, it has the cartilaginous appearance on x-ray, no destruction, no elevation in the periosteum, and it's in the medulla.

Histologically: benign cartilage

No atypia

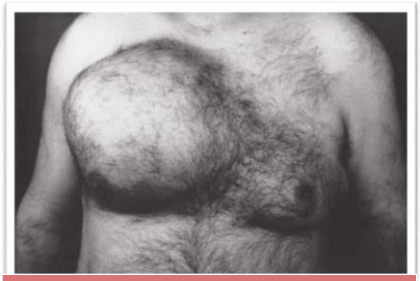
CHONDROSARCOMA:

- Malignant tumors producing malignant cartilage
- 50% incidence of osteosarcoma, chondrosarcoma is less common, if you treat 20 cases of osteosarcoma, you will maybe treat 10 in chondrosarcoma.
- 40-50 years of age; M:F (2:1)
- Characterized with large masses around the shoulder, the pelvis, the ribs
- Genes: *EXT, IDH1, IDH2, COL2A1, CDKN2A*
- Prognosis depends on grade (grade 1 excellent px)
- Trx: surgical +/- chemotherapy, and there is also progress in the methods, we use to treat these lesions.

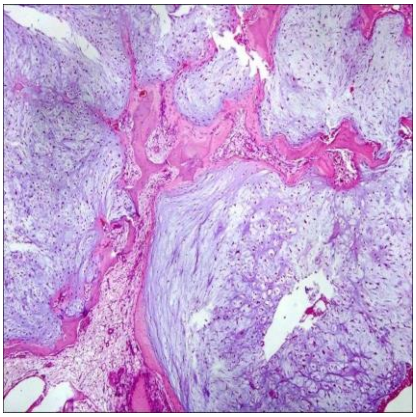
CHONDROSARCOMA FEATURES:

Huge chondrosarcoma in the diaphysis of the humerus, there's Codman triangle, the tumor is infiltrating into the bone marrow and outside of the soft tissue elevating the periosteum.

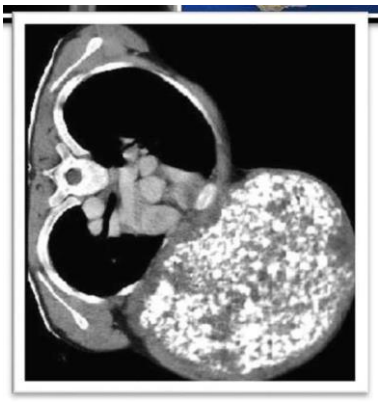
The second half of the pic, the gross specimen which when it was removed, characteristic cut surface of cartilaginous, it's large and infiltrating the soft tissue and the medulla bone.



Large huge chondrosarcoma of the ribs.



Histologically, it’s malignant lobulated cartilage, this is probably grade 1 to 2, because I can still see the cartilaginous differentiation is obvious → low grade tumor.



Probably CT scan, where huge mass with a cartilaginous morphology on imaging, this is called bubble soap appearance.

من الجمال..

في المواصلات، أهديت طفل حبّة "بسكوت" وشَكَرني وقال لي "الله يسعدك" استغربت بدايةً من جمالية الكلمة منه وعدم خجله، مع أني أردّدها دائمًا! سألته؛ شو يعني الله يسعدك؟ قال لي؛ يعني الله يعطيك شاورما 😂

خلي صاحبك يعزمك على سعادة قبل الميد >_<

بالتوفيق يا رب