Benign Bone Tumors

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PLEASE CLICK ON THE FOLLOWING LINK TO WATCH THE LECTURE ONLINE:-

https://www.youtube.com/watch?v=wlyLG0amGkk&list=PLuBRb5B7fa eyzMA0u7jajzWugcmiRi5s&index=2

• I. Bone

- A. Osteoid osteoma
- B. Osteoblastoma

- C. Parosteal osteoma
- D. Bone island (enostosis)

• II. Cartilage

- A. Enchondroma
- B. Periosteal chondroma
- C. Osteochondroma

- D. Chondroblastoma
- E. Chondromyxoid fibroma (CMF)

• III. Fibrous/Histiocytic

- A. Nonossifying fibroma (N O F)
- B. Fibrous dysplasia

- C. Osteofibrous dysplasia
- D. Desmoplastic fibroma
- E. Langerhans cell histiocytosis (LCH)

- IV. Cystic
- A. Unicameral (simple) bone cyst (UBC)
- B. Aneurysmal bone cyst

V. Giant Cell Tumor of Bone

Osteoid osteoma

- A distinctive, painful, benign osteoblastic bone tumor
- Male- to- female ratio = 2:1
- Most patients are between(5 30) years of age
- Genetics/etiology
 - a. The etiology is **unclear**, but nerve fibers associated with blood vessels within the nidus likely play a role in producing pain
 - b. High prostaglandin and cyclooxygenase levels are present within the lesion

Clinical presentationOsteoid osteoma

- □Classic symptom is **night pain** relieved by aspirin or NSAIDs
- ☐ The pain is **progressive** in its severity
- □can be **referred** to an adjacent joint

may be present for months to years **before** diagnosis

 Most common locations include the femur, tibia, vertebral arch, humerus, and fingers

• The **proximal femur** is the most common site; the **hip** is the most common intra- articular location.

 Osteoid osteomas usually occur in the diaphyseal or metaphyseal regions of long bones.

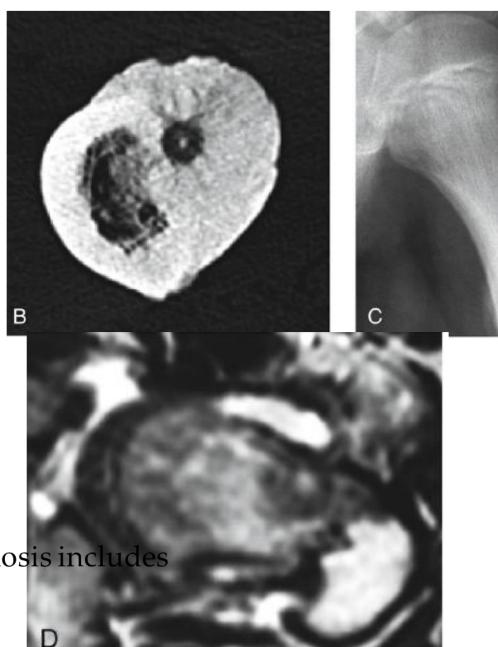
• Osteoid osteomas cause extensive inflammatory symptoms in the adjacent tissues (joint effusions, contractures, limp, muscle atrophy).

Imaging DDX





- Ewing sarcoma, stress fracture.



- Round, well-circumscribed intracortical lesion with radiolucent nidus
- Lesions usually less than 1 cm in diameter

- Extensive periosteal reaction that may obscure the nidus (Figure A).
- Lesions are occasionally intra-articular, subperiosteal, or medullary; these cause less surrounding periosteal reaction(Figure C).
- Intense and focal **increased** tracer uptake on technetium Tc- 99m bone **scans**.
- Thin-cut **CT** scan is often the **key** to diagnosis because it frequently identifies the small radiolucent **nidus** (Figure B).
- MRI often shows extensive surrounding edema (Figure D).

Treatment/outcome

 Long-term medical management with aspirin or NSAIDs is useful to relieve symptoms because these lesions are

- ✓ self- limiting and
- ✓burn out after an average of 3 years

Treatment/outcome

• Standard of care is outpatient percutaneous radiofrequency ablation (RFA) of the lesion

• A CT-guided probe is inserted into the lesion with the temperature raised to 90°C for 4 to 6 minutes to produce a 1- cm zone of necrosis

• Recurrence rates after RFA are **less** than 10%

• **Contraindications** include lesions **close** to the (spinal cord or nerve roots)

Treatment/outcome.....CONT

• Surgical treatments such as surgical resection or burring are rarely used if the lesion is accessible to perform RFA

• In lesions around the **hip**, patients often require **internal fixation**, sometimes with bone **grafting**, if a large portion of cortex is surgically removed with the lesion

Osteoblastoma

• A rare, aggressive, benign osteoblastic tumor

- Male- to- female ratio = 2:1
- Osteoblastomas are much less common than osteoid osteomas
- Most patients are between (10 and 30)years of age

Genetics/etiology—

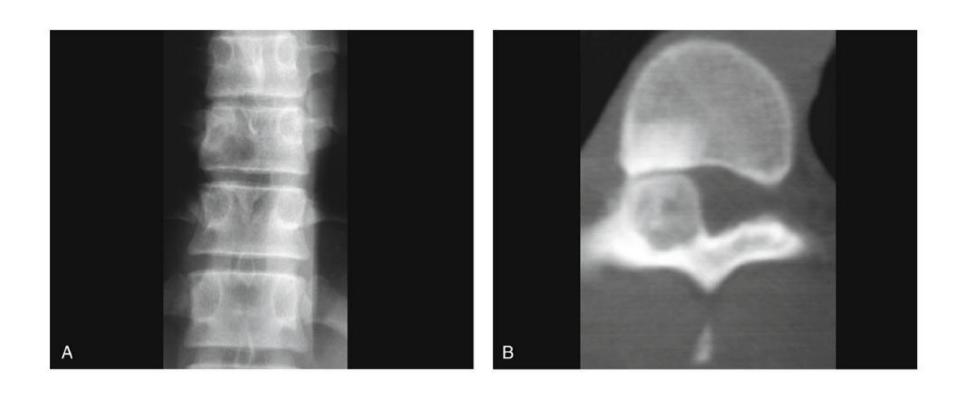
Recurrent rearrangements of FO S and FO S B are Reported

Clinical presentation...... Osteoblastoma

- Slowly progressive, dull, aching pain of long duration
- Less severe than pain from an osteoid osteoma
- Night pain is **not** typical, and aspirin does **not** classically relieve the symptoms
- N eurologic symptoms can occur because the **spine (posterior elements**) is the most common location for osteoblastoma
- Other locations include the **diaphysis** or **metaphysis** of long bones (tibia and femur) and the mandible
- Related swelling, muscle atrophy, and a limp may occur because the lesions are large and present for a prolonged period

Factors Differentiating Osteoid Osteoma From Osteoblastoma

Factor	Osteoid Osteoma	Osteoblastoma
Site	Diaphysis of long bone	Posterior elements of spine,
		metaphysis of long bone
Size	5-15 mm	>1.5 cm
Growth characteristic	Self-limited	Progressive
Symptoms	Exquisite pain, worse at night,	Dull ache
	relieved by aspirin	



17 year-old male, pedicle of T10

Imaging appearance

Radiolucent lesion 2 to 10 cm in size with occasional intralesional densities

Two-thirds of osteoblastomas are **cortically** based; one-third are medullary

- **Expansile** with extension into the surrounding soft tissues and a rim of reactive bone around the lesion
- 25% of osteoblastomas have an extremely **aggressive** appearance and are mistaken for malignancies
- Three-dimensional imaging (CT, MRI) is necessary to fully evaluate the extent of the lesion before surgical treatment

Osteoblastoma

• Radiographic differential diagnosis includes

- osteosarcoma
- aneurysmal bone cyst (A BC)
- osteomyelitis
- osteoid osteoma

Treatment/outcome

 Osteoblastoma is not self-limiting, and it requires surgical treatment

• In most cases, **curettage** and bone **grafting** is adequate to achieve local control

- Nerve roots should be maintained when treating spinal lesions
- Occasionally, en bloc resection is required for lesions in the spine

Enchondroma

- benign tumor composed of mature hyaline cartilage and located in the medullary cavity
- Enchondromas can occur at any age, but they are most common in patients 20 to 50 years of age
- The incidence is unclear because most lesions are found incidentally
- Genetics/etiology
- a. Thought to be related to incomplete endochondral ossification,
- in which fragments of epiphyseal cartilage displace into the
- metaphysis during skeletal growth.
- b. IDH1 and IDH2 somatic mutations are reported in most
- enchondromas.

Enchondroma

Genetics/etiology

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Clinical presentation..... Enchondroma

• Most enchondromas are **asymptomatic** and are noted **incidentally** on radiographs

• Lesions in the **small bones** of the hands and feet can be **painful**, especially after a pathologic fracture

• In a patient with an enchondroma and pain localized to the adjacent joint, the pain often has a cause that is unrelated to the tumor

• If a patient has **pain** and the radiographic appearance is **concerning**, **low-grade chondrosarcoma** must be considered

Clinical presentation..... Enchondroma

- One-half of all enchondromas occur in the **small tubular bones**, with most in the **hands**
- Enchondroma is the most common bone tumor in the hand
- Other common locations include the **metaphysis** or **diaphysis** of long bones (proximal humerus, **distal femur, proximal tibia**)
- Enchondromas are classified by Enneking as inactive or latent bone lesions
- The incidence of high-grade malignant transformation is less than 1%. Rarely, a dedifferentiated chondrosarcoma develops from an enchondroma





Imaging appearance

- Enchondromas begin as well-defined, lucent, **central** medullary lesions that **calcify** over time
- The classic radiographic appearance involves rings and stippled calcifications within the lesion

• Lesions can be 1 to 10 cm in size

- Minimal endosteal erosion (<50% of the width of the cortex) or cortical expansion may be present
- In hand **enchondromas**, the cortices may be **thinned** and expanded

- Cortical **thickening** or moderate to extensive cortical **destruction** suggests a **chondrosarcoma**
- The radiographic differential diagnosis includes
 - ✓a bone infarct and
 - ✓low- grade chondrosarcoma
- The radiographic appearance is more important than the pathologic appearance in differentiating an enchondroma from a low- grade chondrosarcoma

• Enchondromas frequently have mild increased uptake on bone scans due to **continual remodeling** of the endochondral bone within the lesion

Treatment/outcome

- Asymptomatic lesions require no treatment and can be followed with serial radiographs to ensure inactivity for a period of time
- Rarely, when pain due to other causes is excluded, symptomatic enchondromas
 can be treated with curettage and bone grafting
- Pathologic fractures through enchondromas in small, tubular bones can be allowed to heal before consideration of curettage and bone grafting
- Surgery is necessary when clinical presentation and radiographic findings are suspicious for a chondrosarcoma
- A needle biopsy is not reliable to differentiate enchondroma from low-grade chondrosarcoma and should be performed only if confirmation of cartilage tissue type is needed

Related conditions: Ollier disease; Maffucci syndrome

- Ollier disease is characterized by multiple enchondromas with a tendency toward unilateral involvement of the skeleton (sporadic inheritance)
- Multiple enchondromas are thought to indicate a skeletal dysplasia with failure of normal endochondral ossification throughout the metaphyses of the affected bones
- *IDH1* and *IDH2* mutations are present in patients with O llier disease and Maffucci syndrome
- Patients with multiple enchondromas have growth abnormalities causing **shortening** and **bowing** deformities

• **Maffucci syndrome** involves multiple enchondromas and soft- tissue **angiomas**

• Radiographically, the enchondromas in Ollier disease and Maffucci syndrome have **variable** mineralization and often **expand** the bone markedly

• The **angiomas** in Maffucci syndrome can be identified on radiographs because of the presence of **phleboliths** (small, round, calcified bodies)

• The histologic appearance of lesions in a patient with multiple enchondromas is similar to solitary lesions in small tubular bones (hypercellular with mild chondrocytic atypia)

- Patients with multiple enchondromas may require surgical correction of skeletal deformities at a young age
- Patients with **Ollier** disease have an increased risk of malignant transformation of an enchondroma to a low-grade chondrosarcoma (25% to 30%)

- Patients with Maffucci syndrome have an increased risk of
 - ✓ malignant transformation of an enchondroma to a low-grade chondrosarcoma (23% to 100%),
 - ✓ as well as a high risk of developing a fatal visceral malignancy

• Patients with Ollier disease or Maffucci syndrome should be followed long-term because of the increased chance of malignancy

Osteochondroma

• benign **osteocartilaginous** tumor arising from the **surface** of the bone

- Demographics
 - ✓Osteochondromas are the most common benign bone tumor
 - ✓The true incidence of osteochondromas is unknown because most lesions are asymptomatic
 - ✓ Most lesions are identified in the **first 2 decades** of life

Genetics/etiology

• Osteochondromas are **hamartomatous** proliferations of both bone and cartilage

• They are thought to arise from trapped growth-plate **cartilage** that **herniates** through the **cortex** and grows via endochondral ossification beneath the periosteum

- A defect in the perichondrial node of Ranvier may allow the physeal growth to extend from the surface;
- as the cartilage ossifies, it forms cortical and cancellous bone that comprises the stalk of the lesion

Clinical presentation

- Most lesions are **solitary** and **asymptomatic**
- Most are **less than 3 cm** in size, but they can be as large as 15 cm
- Depending on size and location, patients can have **pain** from an inflamed overlying **bursa**, fracture of the **stalk**, or **nerve** compression
- When close to the skin surface, osteochondromas can be palpated as firm, immobile masses
- Osteochondromas continue to grow until the patient reaches skeletal maturity

• The lesions most commonly occur **around the knee** (distal femur, proximal tibia), **proximal humerus**, and pelvis;

• spinal lesions (posterior elements) are rare

• A **subungual exostosis** that arises from beneath the nail in the distal phalanx is a **pottraumatic** lesion and not a true osteochondroma

• When **multiple** lesions are present, the condition is called multiple **hereditary** exostoses

• The risk of malignant degeneration of a **solitary osteochondroma** to a chondrosarcoma is **less** than 1%

• Rarely, a **dedifferentiated chondrosarcoma** can develop from a solitary osteochondroma

Imaging appearance

- Osteochondromas can be **sessile** or **pedunculated** on the bone surface
- **Sessile** lesions are associated with a higher risk of **malignant** degeneration
- Lesions arise near the epiphyseal plate and appear to become more diaphyseal with time
- Pedunculated lesions grow away from the adjacent joint
- The medullary cavity of the bone is **continuous** with the stalk of the lesion
- The cortex of the underlying bone is continuous with the cortex of the stalk
- The affected bony metaphysis is often flared or widened



- The cartilage **cap** is usually **radiolucent** and involutes at skeletal maturity
- Metaplastic cartilage nodules can occur within a bursa over the cartilage cap
- The radiographic differential diagnosis includes parosteal osteosarcoma and myositis ossificans.
- CT and MRI can evaluate the cartilage cap and surrounding soft tissues better than plain radiographs and are useful when malignant degeneration is a concern
- A **thick** cartilage cap implies **growth** but is not a reliable indicator of malignant degeneration

Treatment/outcome

- **Nonsurgical** treatment is preferred in **asymptomatic** or minimally symptomatic patients who are **still** growing
- Relative indications for surgical excision of an osteochondroma (performed by excision at the base of the stalk)
 - ✓ S ymptoms secondary to **inflammation** of surrounding soft tissues (bursae, muscles, joint capsule, tendons) not controlled by NSAIDs or activity modification
 - ✓ Symptoms secondary to frequent **traumatic** injury
 - ✓ Significant **aesthetic** deformity
 - ✓ Symptoms secondary to **nerve** or **vascular** compression
 - ✓ Concern for **malignant** transformation

• The **perichondrium** over the cartilage cap must be **removed** to decrease the likelihood of local recurrence

• **Delaying** surgical excision until skeletal maturity **increases** the chance of local control.

• The surgeon should be aware that a patient with an osteochondroma extending into the **popliteus fossa** can have a *pseudoaneurysm* and is at risk for *vascular* injury during excision

Related condition: multiple hereditary exostoses

• a skeletal dysplasia that is inherited with an **autosomal dominant** pattern

• Patients may have up to **30** osteochondromas throughout the skeleton

• EXT1 and EXT2 are genetic loci associated with this disorder

multiple hereditary exostoses

 Clinically, patients with the disorder have skeletal deformities and short stature

The lesions are similar radiographically and histologically to solitary osteochondromas

 Radiographs reveal primarily sessile lesions that may grow to be very large

Metaphyseal widening is present in affected patients

• **Deformities** occur as a result of **disorganized** endochondral ossification in the epiphyseal plate and may require surgical correction, especially in the paired bones (radius/ulna, tibia/fibula)

- The risk of malignant transformation is higher (\sim 5% to 10%) in patients with this condition than in patients with solitary lesions
- The most common location of a **secondary** chondrosarcoma is the **pelvis**
- The malignant tumors are usually **low grade** and grow **slowly**

Chondroblastoma vs Chondromyxoid fibromas





well- circumscribed round lesion

in the proximal tibial **epiphysis** extending slightly into the metaphysis





A periosteal shell that is not easily seen

eccentric lytic lesion with a well- defined intramedullary border

eccentric lesion expanding the cortex With a visible rim

Nonossifying fibroma (N O F)

• A developmental abnormality related to **faulty ossification**; not a true neoplasm

Very common skeletal lesions

• Occur in children and adolescents (age 5 to 15 years)

• NOFs are found in 30% of children with open physes

Also frequently called

- ✓ fibrous cortical defect or
- ✓ metaphyseal fibrous defect

• Genetics/etiology—Possibly caused by abnormal **subperiosteal osteoclastic resorption** during remodeling of the **metaphysis**

Clinical presentation.....NOF

- Usually an incidental finding
- May be multifocal, Types include
 - ✓ Familial multifocal
 - ✓ Neurofibromatosis
 - ✓ J affe-Campanacci syndrome (congenital, with cafe-au-lait pigmentation, mental retardation, and nonskeletal abnormalities involving the heart, eyes, and gonads)
- Most common in long bones of lower extremity (80%)
- Patients occasionally present with a **pathologic fracture** (more common in the distal tibia)

Radiographic appearance,,,,,,,,,,,, NOF

- Eccentric, lytic, cortically based lesions with a sclerotic rim
- Occur in the **metaphysis** and appear to migrate to the diaphysis as bone **grows**
- May thin the overlying cortex with expansion of the bone
- Lesions enlarge (1 to 7 cm) as the patient grows
- As the patient reaches skeletal maturity, the lesions **ossify** and become **sclerotic**

- Occasionally associated with secondary ABC
- Plain radiographs are diagnostic
- An avulsive cortical irregularity
 - is the result of an avulsion injury at the insertion of the **adductor** magnus muscle on the posteromedial aspect of the distal femur and can be similar in appearance to an NOF

Treatment/outcome

- Most are managed with **observation**; spontaneous regression usually occurs
- Large lesions can be monitored along with skeletal growth
- Curettage and bone grafting may be indicated for **symptomatic** and **large** lesions
- Pathologic fractures are often **allowed to heal** and then are observed or treated with curettage and grafting
- Internal fixation is rarely needed; depends on anatomic location





an 11- year- old boy reveal an NOF that has **healed** after a minimally displaced **pathologic** fracture.

It is an **eccentric**, **scalloped** lesion with a **sclerotic** rim. Anteriorly, the lesion is filling in with bone

Fibrous dysplasia

• A common developmental abnormality characterized by **hamartomatous** proliferation of fibro-osseous tissue within the bone

• Can be seen in patients of any age, but approximately 75% are seen in patients **younger** than 30 years

• Females affected more commonly than males

Fibrous dysplasia

- Genetics/etiology
- Solitary focal or generalized multifocal inability to produce mature lamellar bone
- Areas of the skeleton remain indefinitely as immature, poorlymineralized trabeculae
- Not inherited
- Monostotic and polyostotic forms are caused by the dominant activating mutations of GS α on chromosome 20q13, which produce a sustained adenylate cyclase–cyclic adenosine monophosphate activation
- Fibrous dysplasia tissue has high expression of fibroblast growth factor-23, thought to be the cause of hypophosphatemiaNin patients with McCune-A lbright syndrome or oncogenic osteomalacia

Clinical presentation

- Usually asymptomatic and found incidentally
- Can be monostotic or polyostotic
- Can affect any bone but has a predilection for the proximal femur, rib, maxilla, and tibia
- Fatigue fractures through the lesion can cause pain
- Swelling may be present around the lesion
- Severe cranial deformities and blindness with craniofacial involvement may be present

- Patients occasionally present with **pathologic** fractures
- McCune-A lbright syndrome—**Triad** of polyostotic fibrous dysplasia, precocious puberty, and pigmented skin lesions
 - ✓ Unilateral bone lesions
 - ✓ Skin lesions usually on the same side as bone lesions
 - ✓The syndrome is present in 3% of patients with polyostotic fibrous dysplasia

Radiographic appearance

- Central lytic lesions within the medullary canal, usually diaphysis/metaphysis
- Sclerotic rim
- May be expansile with cortical thinning
- Ground glass or shower- door glass appearance
- Bowing deformity in proximal femur (shepherd's crook) or tibia
- Vertebral collapse and kyphoscoliosis may be seen
- Long lesion in a long bone
- Plain radiographs usually diagnostic



central, lytic bone lesion with a ground glass appearance fills the femoral neck, consistent with fibrous dysplasia.



Long lesion in a long bone

Treatment/outcome

- Asymptomatic patients may be observed
- Surgical indications include painful lesions, impending/actual pathologic fracture, severe deformity, and neurologic compromise (spine)
- Surgical treatment: curettage and bone grafting of the lesion (I t is important to use cortical allograft, not cancellous autograft, because cancellous autograft is replaced by dysplastic bone.)

 Internal fixation (intramedullary device more effective than plate) usually required to achieve pain control in the lower extremity Osteotomies for deformity

- Beneficial long-term outcomes of long-term diphosphonate therapy in most patients with fibrous dysplasia.
- In approximately 1% of lesions, malignant transformation to osteosarcoma, fibrosarcoma, or undifferentiated pleomorphic sarcoma occurs, with extremely poor prognosis

Unicameral (simple) bone cyst (UBC)

A common, serous fluid-filled bone lesion

- Most cases occur in patients younger than **20** years
- Thought to result from a **temporary** failure of medullary bone formation near the epiphyseal plate during skeletal growth

Unicameral (simple) bone cyst (UBC)

- The cyst is active initially when adjacent to the epiphyseal plate
- When medullary bone formation resumes, the cyst appears to move into the diaphysis

- Possible causes and precursor lesions include
 - ✓ lymphatic/venous obstruction,
 - ✓intraosseous hematoma, and
 - ✓intraosseous synovial rest

Clinical presentation

• The most common presentation is a **pathologic fracture** after minor trauma

Painful symptoms resolve when the fracture heals

 The most common locations include the proximal humerus and proximal femur, but UBCs can also occur in the ilium and calcaneus

Imaging appearance

• Purely lytic lesion located **centrally** in the medullary canal

• UBCs start **metaphyseal**, adjacent to the epiphyseal plate, and appear to progress toward the **diaphysis** with bone growth

Narrow zone of transition between cyst and normal bone

Cortical thinning but no soft- tissue extension

- Bone expansion does not exceed the width of the physis
- Trabeculations occur after multiple fractures
- "Fallen leaf" sign is pathognomonic (cortical fragment that has fallen into base of empty lesion)

• Plain radiographs are usually diagnostic, but T2-weighted MRI shows a well-defined zone of bright, uniform signal intensity (when not fractured).



lytic lesion **centrally** located in the **medullary** canal of the metaphysis

The lesion does not expand the bone wider than the epiphyseal plate

Treatment/outcome

- Natural history: fills in with bone as the patient reaches skeletal maturity
- After acute fractures, lesions are occasionally stimulated to fill in with native bone
- Lesions continue to grow in young children and can be difficult to avoid fracture
- Evidence-based treatment: intralesional injection of methylprednisolone acetate
- Multiple injections may be required, especially in very young children

• No evidence to suggest improved outcomes with injection of bone marrow or graft substitutes; however, they remain in use

• Large or growing proximal femoral lesions with or without a pathologic fracture are often **treated** with curettage/bone grafting/internal fixation given the high-stress area and need to preserve ambulation.

Aneurysmal bone cyst

- A neoplastic process causing a destructive, **expansile** bone lesion filled with multiple blood- filled **cavities**
- 75% of patients are younger than 20 years
- Seventy percent of A BCs express recurrent translocations of *USP6*
- Can arise de novo or be associated with an underlying lesion that is identifiable in 30% of cases (most commonly **chondroblastoma**, **giant** cell tumor, chondromyxoid fibroma, nonossifying fibroma, osteoblastoma, or fibrous dysplasia)
- Secondary ABCs do not express USP6.

Clinical presentation

• Pain and swelling are the most common symptoms

• Pathologic fracture as a presenting symptom is rare

Neurologic symptoms are possible with lesions in the spine

• Most common locations are the **distal femur**, **proximal tibia**, pelvis, and spine (posterior elements)

Imaging appearance

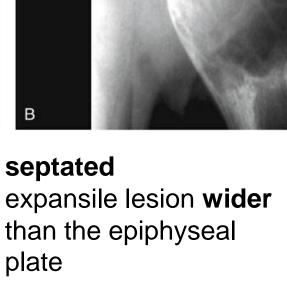
- Eccentric, lytic lesions located in the metaphysis
- Aggressive **destruction** of or **expansion** into the cortex and extension into the soft tissues may be seen
- Lesion can expand to greater than the width of the epiphyseal plate
- Usually a periosteal rim is maintained around the lesion
- Can grow contiguously across adjacent spinal segments or extend through the epiphyseal plate

No matrix mineralization

• T2-weighted MRI shows **fluid-fluid levels** (separation of serum and blood products)

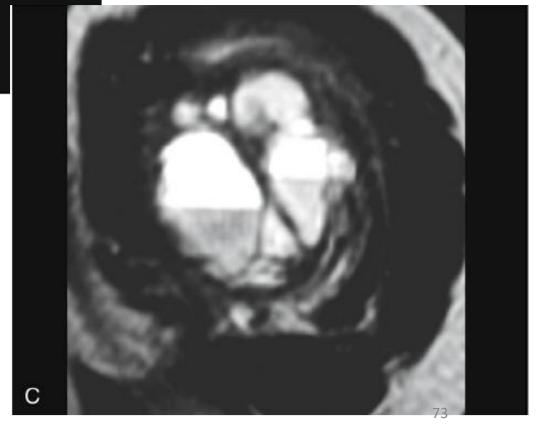
- Radiographic differential diagnosis includes
 - >UBC
 - ➤ telangiectatic osteosarcoma





eccentric lytic lesion located in the metaphysis that expands into the soft tissues with a periosteal rim

fluid-fluid levels



Factors	Unicameral Bone Cyst	Aneurysmal Bone Cyst
Presentation	Pathologic fracture	Pain, swelling
Common locations	Proximal humerus	Distal femur, proximal tibia
	Proximal femur	Pelvis
		Posterior elements of spine
Radiographic characteristics	Central, lytic lesion	Eccentric, lytic lesion
	Metaphyseal	Metaphyseal
	Symmetric expansion less than	Can expand wider than
	width of epiphyseal plate	epiphyseal plate
		Extends into soft tissues with a
		thin periosteal rim
Treatment	Intralesional steroid injection	Curettage and bone grafting
	Curettage/grafting/internal	Embolization (spine, pelvis, and
	fixation (proximal femur)	so forth)

Treatment/outcome

- Surgical treatment is curettage and bone grafting of the lesion
- Local adjuvants (argon beam coagulation, phenol) can be used after curettage
- Highest local recurrence is in young patients with an open physeal plate
- For local recurrence, repeat **curettage** and grafting is indicated; **sclerotherapy** for cases not amenable to repeat curettage
- Expendable bones (proximal fibula) may be resected
- Embolization or sclerotherapy can be useful for pelvic or spinal lesions alone or in combination with surgical treatment

Giant Cell Tumor of Bone

• a benign, aggressive bone tumor consisting of distinct, undifferentiated mononuclear cells

- Most occur in patients 30 to 50 years of age (90% older than 20 years)
- Affects **females** more commonly than males
- Etiology is unknown
- Stromal cells have alterations in the *c- myc, c- Fos,* and *N- myc* oncogenes

Clinical presentation

• Main symptoms: pain and swelling for 2 to 3 months

Decreased range of motion around a joint

• Some patients (10%) present with a **pathologic fracture**

 Located most commonly in the distal femur, proximal tibia, distal radius, proximal humerus, proximal femur, sacrum, and pelvis

Imaging appearance

- Eccentric, lytic lesions located in the epiphysis/metaphysis of long bones
- May arise in an apophysis
- Lesions extend to the subchondral surface with no sclerotic rim
- Can **destroy** the cortex and extend into the surrounding tissues
- Located in the anterior vertebral body when the spine is involved
- Commonly have a secondary ABC component
- 7.

• Associated soft- tissue calcifications may be present

Bone scan shows increased uptake in the lesion

• MRI is helpful only to define the extent of soft-tissue and marrow involvement; plain radiographs are usually diagnostic



Treatment/outcome

- Most lesions can be treated with thorough curettage and a high-speed burr
- Thorough intralesional treatment requires making a large cortical window
- Local surgical adjuvants (phenol, cryotherapy, argon beam) are commonly used to try to decrease local recurrence
- Defect can be filled with either bone graft or methyl methacrylate (equivalent recurrence rate), with or without internal fixation, depending on the defect size

• Local recurrence with intralesional treatment is 10% to 15%

• Local recurrence can be in the local bone or can manifest as adjacent soft-tissue masses

Aggressive lesions may require resection and reconstruction

• Embolization should be used for large pelvic or spinal lesions alone or in combination with surgical treatment

• Denosumab is FDA approved for the treatment of unresectable giant cell tumor of bone

• Studies have shown disease and symptom control for advanced or refractory disease

 Denosumab causes the giant cell tumor to ossify, potentially allowing easier surgical treatment

• Treatment with denosumab postoperatively can prevent recurrence but residual tumor cells will grow after cessation of treatment

 Radiation is occasionally used in multiply recurrent or surgically inaccessible lesions

• The tumor metastasizes to the lungs in 2% of patients (benign metastasizing giant cell tumor)

- ✓a. Treatment includes thoracotomy, radiation, chemotherapy, or observation
- ✓b. Of patients with metastatic disease, 10% to 15% die of the
- **✓** Disease

• Rarely, giant cell tumor is malignant (\sim 1%).

Thank you