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| **Bisphosphonate Therapy** |
| **Bisphosphonates** | **Oral Bisphosphonate Theraphy:****Osteoporosis** |
|  - **Inhibit osteoclast differentiation & recruitment**- Induce osteoclast apoptosis- May inhibit angiogenesis- BON: First Case reports in 2003- Rare; Most cases are in the jaws- No clinical trial data- Can be PO or IV- Can be first or second generation drugs- **Currently only the 2nd gen drugs are assoc. w/ BON**2nd gen get incorporated into the skeleton, are very potent and have extended half-life (eg. Fosamax half life is 12 yrs) | - Fosamax (alendronate) po, 2nd, weekly- Actonel (risedronate) po, 2nd - Boniva (ibandronate) po or iv, 2nd, mo.- Didronel (etidronate) po 1st - Skelid (tiludronate) po 1st **- Reclast**(zoledronate) –once yearly iv, 2nd, FDA approved 2007, infusion takes 15 minutes- Incidence of BON in pts treated for osteoporosis is low esp. 1st gen & esp. po🡪 ~0.7 per 100,000 pt-years use of Fosamax est by drug industry-Increased risk of BON with PO drug with: - Dental surgery (extractions most cases) - Dental disease/infections - Oral trauma - Periodontitis - Concomitant chemotherapy or corticosteroids - Over 65 yo - Tori or bony exostoses - Diabetes - Smoking alcohol use - Poor OH - Duration of drug over 3 years |
| - Normally:- Osteoclasts repair microfractures and resorb foci with nonviable osteocytes- With Bisphosphantes:- Decrease in osteoclast fnct which allows microfractures to accumulate.- Bone turnover slows so osteocytes that have exceeded their 150 day lifespan (ie. Die) are allowed to remain |
| - Indications:- Osteoporosis- Treat Paget’s disease- Cancer: Slow metastases of several cancers (breast, prostate, multiple myeloma), and prevention of cancer treatment induced bone loss- On the horizon: prevent metastases - may have antitumor activity & may prevent bone metastasis - mechanisms may include induction of apoptosis, inhibition of tumor cell invasion and angiogenesis, and tumor growth reduction | **IV Bisphosphonate Therapy:****Cancer** |
| - Treatment for Myeloma, breast and prostate cancer- **Zometa**(zoledronic) iv, 2nd gen (same drug as Reclast)- Aredia (pamidronate) iv, 2nd gen- Bonefos (clodronate) po or iv, 1st gen**- 94% of cases of BON assoc. w/ Zometa and Aredia**- 85% of cases in myeloma pts- Incidence for pts taking 2nd gen for cancer: 6-10% |

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|  | **Bisphosphonate Associated Osteonecrosis (BON)** |
| **Definition** | Etio:- (60%)Exposed bone from invasive procedure - (40%)Spontaneous - (Small Number) After minor trauma to bony protuberances like torus |
| **Clinical Feat.** | - 2/3 in mandible, but maxilla or both jaws simultaneously- 2/3 painful- Exposed bone- +/- pain- +/- infection  - Infection can lead to fistula formation or fracture |
| **Radio** | - Early: before clinical, increased radiodensity of ALVEOLAR bone (crestal)- Late: “**moth-eaten**” (ill defined) radiolucency or mixed (opaque sequestra in center if any) |
| **Histology** | - Necrotic bone is sclerotic w/ loss of osteocytes from the lacunae and peripheral resorption w/ bacteria- Trabecular bone pattern can look like Paget’s disease- Ostoclasts big and vacuolated |
| **Tx/Prognosis** | **- Conservative management: the goal of therapy is to stop pain and infection**- ASx pts: Chlorhexidine rinse daily, smooth rough edges of exposed bone- Sx pts: Chlorhexidine and Antibiotics- Amoxicillin +/-metronidazole- Clindamycin, azithromycin- Iv antibiotics in hospital if the po fail-Analgesics-Minimal surgical debridement-No role for HBO-No real role for d/c drug in short term because of half life, but it is commonly done in practice |

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|  | **Osteogenesis Imperfecta****(OI)** | **Cleidocranial Dysplasia****(CD)** | **Osteopetrosis (Osteopetrosis)** |
| **Definition** | - Disorder of **collagen** maturation - **Most common inherited bone disease but still rare** **(1/8000 live births)** - Spontaneous/Sporadic possible**- A group of diseases (4 types)** - Abnormal maturation of type I collagen (chromosome 7 or 17) | - **Osteoblastic** differentiation effected - Genetic defect on chromosome 6- Autosomal dominant- 40% spontaneous- Membranous bones predominately effected (ie. clavicles and skull), but also long bones (endochondral) | - **Osteoclasts** not function- Rare, genetic - group of diseases - 2 major forms (infantile & adult) vary by severity & genetic origin |
| **Clinical**  | - Bone fragility, blue sclera, altered teeth, hearing loss, long bone&spine deformities, joint hyperextensibility- Craniofacial findings: - Class III malocclusion due to maxillary hypoplasia , sometimes florid osseous dysplasia-like bone changes- Teeth:- dentinogenesis imperfecta primary & permanent teeth (opalescent teeth)- Type I:- Most common form and mildest- Autosomal dominant- Preschool fractures, adult hearing loss, joint hypermobility, bruising, blue sclera, - +/-opalescent dentin- Type II: most severe form, often stillborn- Type III: most severe form in live births surviving beyond perinatal period- Type IV: mild –moderate form | - Clavicles: - hypoplastic or aplastic, uni or bilaterally, increased shoulder mobility- Skull bones:  - sutures and fontanels remain open, wormian bones develop in the suture lines, frontal and parietal bossing, large heads- Hypertelorism- High palate, depressed nasal bridge- Absent maxillary sinus- Teeth: - Many unerupted perm & supernumerary teeth (often misshapen)- Primary teeth may be retained into adulthood- Absent cementum |  - InfantileType: - auto recessive - marrow failure - facial deformity (frontal bossing, broad face, hypertelorism, delayed tooth eruption, deafness, facial paralysis) - no distinction btwn cortical & medullary bone on radiograph- Adult Type: - auto dominant - axial and craniofacial skeleton - may discover on dental radio - bone pain - common  |
| **Radio** | - thin cortex- fine trabeculation- osteopenia- bowing of long bones- multiple fractures- wormian bones of skull |  | - infantile: Dense bone, sclerotic marrow spaces replaced w/ bone or fibrous tissue- Adult: generalized increase in bone density, reduced sinus cavities in size, unerupted teeth are common |
| **Histo** | - Immature bone architecture ie. Woven bone does not mature to lamellar bone |  | - sclerotic bone – NO marrow spaces |
| **Tx/Prog** | - Tx: None- Manage fractures- tooth attrition and tooth loss- Prognosis: variable w/ type of OI | - Tx: none- Function well- Extrusion of perm. teeth orthodontically to increase fnct, encourage growth of the alveolus, and correct vertical growth of the jaws to prevent class III occlusion | - Tx: none- Tx complications of osteomyelitis and fracture, bone marrow obliteration, etc. |

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|  | **Paget’s Disease of Bone** | **Idiopathic Osteosclerosis** | **Focal Osteoporotic** **Bone Marrow Defect** |
| **Def** | - Common (1% of men over 45 yo), esp in England- Acquired bone metabolism problem of **increased bone turnover** (increased resorption and deposition)- Etiol: ? | - common (5% of population)- Focal radiodensity of the jaw which is **not** inflammatory, dysplastic, neoplastic, nor manifestation of a systemic disease- Etiol: ? | - Area of normal bone marrow in the jaws which is big enough to produce a radiolucency on plain film- Importance: can confuse w/ pathology- Etiology: ?? Marrow hyperplasia secondary to healing/bone fill |
| **Clinical Feat.** | - Over 40 yo, white men- Sx vary from subclinical to pain esp. near joints, bowing of long bones, enlarging skull, some monostotic (malignant), most polyostotic- 17% of Paget’s pts have  - jaw (body and alveolus) expansion - max enlarges>mand (lion-like) - sinus obstruction - chronic and slow progression- Dx: - Blood tests: inc alkaline phosphatase (marker of osteoblast activity), normal calcium, normal phosphorous - Urine: inc hydroxyproline, and other markers of bone resorption - Biopsy usually unnecessary | - Teens and 20s- Asx: Incidental finding on radio- No jaw expansion- Dx:  - radio, hx - biopsy **rarely** needed - follow with radio periodically –should have minimal growth and sometime even regression | - 70% posterior mand- Women 3:1- Asx: Incidental finding on radio- No jaw expansion- Freq at healed exo site- Dx: radio, +/-biopsy |
| **Radiographic** | - Early phases “osteoclastic”(osteopenia): decreased density and lucencies esp. of skull (osteoporosis circumscripta)- Later “osteoblastic” stages: patchy sclerosis becomes confluent (“**cotton wool**”), teeth hypercementosis | - Well defined- Rounded or triangular- Uniformly Radiopaque (no lucent)- mm-cms- Rt apex or inter-radicular area 80%- **Rare** rt resorption/tooth movement (but documented) | - ill-defined borders- Lucency- mm to cm(s)- Fine trabeculations (decreased bone density, not “lesion”) - Biopsy proved it to be filled w/ normal bone marrow |
| **Histology** | - Very vascular fibrous CT replaces marrow- Basophilic reversal lines=junction btwn alternating resorptive and formative phases of bone (A.K.A. “jigsaw” or “mosaic” pattern)- Active phase: Alternating resorption (osteoclastic) & formation (osteoblastic) activity on same bone trabeculae- Less active phases (final stage): more sclerotic , dense bone w/ prominent reversal lines | - Lamellar bone and marrow | - normal hematopoietic bone marrow |
| **Tx/Prognosis** | - None - If severe course, parathyroid hormone antagonists (blocks osteoclastic activity)- Bisphosphonates (eg.Skelid, 7 Bisph. drugs currently indicated)- Osteosarcoma may develop (1-13%)—usually lower extremity or pelvis- Benign & malignant giant cell tumors – rare but jaws most commonly involved | - none | * None (no assoc w/ systemic disorders)
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|  | **Langerhan’s Cell Histiocytosis****= Histiocytosis X****= “Eosinophilic granuloma”** | **Central Giant Cell Granuloma (CGCG)** | **Cherubism** |
| **Def** | - Monoclonal proliferation of histiocyte-like cells (Langerhans cells w/ a polyclonal infiltrate of multinucleated giant cells, eosinophils, and other chronic inflammatory cells)- Langerhans cells are antigen presenting mononuclear cells- Spectrum of diseases | - Tumor? “Reparative”? - Osteoclast 🡪 tumor 🡪 over-resorption of bone | - Genetic disease; **syndrome**- autosomal dominant- chromosome 4 - variable expressivity- Spontaneous/Sporadic possible |
| **Clinical** | - Children (50% < 10 y.o)- Bone lesions most common manifest.- Jaw lesions (10 -20%) in pts over 20 y.o - **Eosinophilic granuloma of bone** (mono- or polyostotic): radiolucency in bone- **Chronic disseminated histiocytosis:** bone, skin, viscera (“**Hand-Schuller-Christian disease”-** triad of bone lesions, exophthalmos, diabetes insipidus)- **Acute disseminated histiocytosis**: prominent visceral, bone marrow, skin involvement—usually **infants**(“**Letterer-Siwe disease**”); Leukemia-like clinical feature | - Mand anterior to 1st perm M - Frequently cross the midline- 60% < 30 y.o but, wide age range- Female often- Asx, slow growth- Aggressive lesions: - rapid growth, expand bone, pain, rt resorption, paresthesia, perforate cortical plate, and produce an associated soft tissue lesion (ulcer or mass), high recurrence rate | - Posterior body and ramus - Begin to see changes 2-5 y.o and then stabilize at puberty- **Bilaterally** develops bone lesions - Painless expansion- Expanded mandible & orbital bone produced cherubic like facial appearance (angelic chubby cheeks, eyes turned twd heaven)- Severe cases can expand alveolus and displace teeth or block eruption- maxilla involved in severe cases as other craniofacial, long bones- DX: radio, family hx, biopsy, rule out hyperparathyroidism and other syndromes with CGCG |
| **Radiographic** | - Punched out radiolucencies  - well defined w/o corticated rim- Teeth may appear "floating in space" - absolutely no bone around a tooth- sometimes ill-defined esp. in the jaws  - alveolar bone destroyed from crest 🡪mimicking severe perio +/-gingival ulcers or masses, **or**  **-** body of mandible/maxilla 🡪 mimicking periapical inflammation | - Unilocular or multilocular radiolucencies- Well defined, corticated or non-corticated - If multifocal: rule out cherubism and hyperparathyroidism | - **bilateral**- multifocal, multilocular radiolcencies (unilocular possible) |
| **Hist** | - Langerhans cells (CD1a+, S100 +, peanut agglutinin +, Birbeck granules) plus eosinophils, MNGC, plasma cells and lymphocytes | - MNGC (probably osteoclasts)- Spindle cell background- Hemorrhage | - **Central giant cell granulomas (CGCG)** |
| **Tx/Prognosis** | - Bone lesions:  - surgically removed (curetted) if accessible - if not RT or steroid injections 🡪 very good prognosis- Chronic disseminated: chemo- Acute disseminated: chemo w/ more guarded prognosis | - Surgery (curettage 🡪20% recur and respond well to curettage)- Aggressive lesions: intralesional or systemic chemotherapy w/ corticosteroids or calcitonin respectively- Very good prognosis overall | - ?- Often remission and involution after puberty, sometime grotesque deformities remain- Surgical curettage good for some/aggravating to others |

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|  | **Traumatic Bone Cyst****= simple bone cyst****= solitary bone cyst** **= “Idiopathic bone cavity”** | **Aneurysmal Bone Cyst (ABC)** | **Hemangioma of Bone** **(Intrabony vascular malformation)** |
| **Def** | - Empty or fluid filled cavity in bone- Misnomer: not true cyst- Etiology? Trauma-hemorrhage theory - trauma without fracture - hemorrhage that does not reorganize into bone- Reported in almost all bones of body | - Intrabony accumulation of blood. - Reactive fibrous CT and bone- Misnomer: not true cyst- Etiol: ? Primary lesion or disrupted vascular dynamics in a preexisting bony lesion? |  |
| **Clinical** | - Mandible>>>>mx - Pm/molar body of mand - 10-20 y.o, rare over 35 y.o- slight male > female- painless, no expansion usually- Rarely, pain, paresthesia, cortical expansion - DX: radio, cl, and “biopsy” – empty bone cavity found at surgery | - Posterior mandible- Children and young adults- More common in long bones, vertebrae- Pain, rapid swelling, ballooning or “blow out” of bone- Rarely paresthesia, teeth mobility and root resorption, maxilla involving sinuses | - Mandible- Usually 10-20 y.o, (HOWEVER, **adults can manifest vascular Malformations!!**)- Females- Sx variable (asx, pain, swelling)- Tooth mobility- Can expand jaw - Spontaneous bleeding from sulcus, venous or a/v malformation- if high flow, may feel pulse  |
| **Radiographic** | - Unilocular Radiolucency- variable definition (well to poorly defined)- 1-10cm- **scallops** btwn rts of teeth w/o devitalizing or resorbing or moving- Very very rare multilocular | - Uni or multilocular - well defined to poorly defined- Non-corticated Radiolucency- Cortical expansion and thinning- +/-small calcifications | - Multilocular or less often unilocular radiolucency- **Sunburst** periosteal reaction (cross-section) |
| **Hist** |  | - Blood filled spaces lined by fibrous CT and MNGC- Calcifications in wall |  |
| **Tx/Prognosis** | - Surgical exploration of cavity (curettage; bleeding into cavity) often causes repair within 6 months- Rare recurrence or persistence | - Surgery -“blood soaked sponge” curetted out and dark venous blood fills the cavity- Surgical defect heals within 6 months- Cryosurgery may be needed | - MAKE THE DX!!!! Pose tremendous surgical risk!!- Reason to aspirate all intrabony lesions prior to surgery- No extractions in the area- No extractions without a radiograph- Angiography delineated flow and feeder vessels- May need embolization prior to resection |

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|  | **(Benign) Fibro-osseous Lesions of the Jaws (BFOL)** - Def: group of pathologic processes in which fibrous CT and mineralized product replaces bone- Clinical and radiographic features +/-histology is required to make a specific diagnosis |
|  | **Fibrous Dysplasia (FD)** | **Cemento-osseous Dysplasias (COD)** | **Ossifying Fibroma (OF)** |
| **Def** | - **Sporadic postzygotic gene mutation** causes tumorous swellings of bone(s) and possible skin & endocrine abnormalities- Severity of disease likely depends on the point in fetal life that the mutation occurs (undifferentiated stem cells vs. skeletal progenitor cells, etc.) | - All types occur **only** on the jaws and in the tooth bearing areas (alveolus, not body of jaws, etc.)- **3 Types**: Periapical, Focal, Florid COD- Represent variants of the same process, and probably a progression of disease | - Similar in radiographic and histologic appearance to CODs but OF is a **true neoplasm** - Historically reported w/ focal COD |
| **Clinical Feat.** | - **Monostotic** FD (85%): - single bone or craniofacial bones - jaws most common bone - mx>md  - Dx in teens - slow growing, painless swelling  - mx often involves zygoma, sphenoid, etc=craniofacial FD - teeth displaced but **not** mobile- **Polyostotic** FD: - 2 or more bones - long bones subject to fracture and resultant deformity - **Jaffe-Lichtenstein syndrome**:  PolyFD +café au lait (skin lesion) - **McCune-Albright syndrome**:  PolyFD +café au lait + multiple endocrinopathies | - Asx, no expansion- **Periapical COD(PACD)**- **Anterior mand only**  - **ave 40 yo AA women (30-50 y.o)**- **Periapical area** - **Focal COD**- Posterior mand often - ave 40 y.o AA (+ some Caucasian) women - Any area of the jaw- **Florid COD**- Multifocal involvement - **ave 50 yo AA female (40-60 y.o)** - often all 4 quadrants  - Bilateral and symmetric - **VERY** radiopaque-Dx: hx, radio findings, and pulp testing- **AVOID biopsy**! (b/c infection of bone) | - Posterior mandible (PM-M region)- Females in 20s - 30s- jaws and craniofacial bones- expand bone- grow slowly in a centrifugal way- Rare, pain and paresthesia |
| **Radiographic** |  - Poorly defined uniformly radiopaque  - “Ground glass” trabecular pattern (poorly calcified & haphazardly arranged)- Pdl narrowed, lamina dura blends- Mx: floor of sinus displaced superiorly or sinus obliterated, extension into adj bones (occiput, sphenoid, orbit, frontal, etc.)- Md: inf alv canal displaced superiorly, lower border of md bowed | - Early lesions: radiolucency at the apex of vital teeth (radiographically identical to pa granuloma/cyst)- Later lesions: adj teeth develop radiolucencies and the lucencies fuse to form a continuous lucency at apex of several teeth- Mature lesions: mixed w/ radiolucent rim and a radiopaque center, w/ PDL intact & no fusion to the tooth 🡪 **“cotton wool”** | - Early: well defined unilocular radiolucency or mixed lesion, varying amounts of radiopacity, some have sclerotic border- Later: become large, cause rt divergence and bow the inferior border of mandible, increasing radiopacity in center? |
| **Histo** | - Haphazard mix of immature (woven) bone in fibrous ct stroma.- **Metaplastic bone**—arises directly from stroma w/o osteoblastic rimming- **“Chinese character-like” trabeculae**: irregularly shaped trabeculae that do not connect w/ each other | - All three CODs have same histology- “Fibro” fibrous connective tissue- “osseous” metaplastic immature woven bone (**not** rimmed by osteoblasts)- ”cemento” cementum-like droplets (parvicellular)- Proportion of each element relates to the degree of maturity of the lesion | - very similar if not identical to cods- “Fibro” fibrous connective tissue- “osseous” immature woven bone  (**may be** rimmed by osteoblasts)- ”cemento” cementum-like droplets (parvicellular)-Usually submitted to lab in one piece b/c at surgery it shells out (potato-like) mass |
| **Tx/Prognosis** | - Cosmetic shave down; best left until skeletal maturity when lesions usually stop enlarging- Contraindication: Radiation due to risk for osteosarcoma | - Tx: none - Contraindication: surgery of all kinds due to risk of osteomyelitis, esp. Florid COD- Dental care aim to avoid tooth loss & surgical needs- May develop traumatic bone cysts in lesions | - Enucleate, rare recurrence- **Must treat** because it is a neoplasm with capacity to destroy large segments of bone |

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|  | **Osteoma** | **Gardner Syndrome** | **Osteoblastoma** | **Cementoblastoma** |
| **Def** | - True neoplasm?- **Restricted to craniofacial skeleton** | - Genetic- Auto Dom- chromosome 5- 33% spontaneous, rare- Multiple osteomas- Part of “**familial colorectal polyposis”** | - Rare benign bone neoplasm | - Neoplasm of cementum (really an odontogenic tumor) |
| **Clinical Feat.** | - Body of mandible or condyle or paranasal sinus - Young adults- Asx until large- Periosteal (on bone) or endosteal (in bone)(can cause bone expansion) | - GI polyps: colon mostly- Osteomas: At puberty craniofacial begin, any part of jaws but often **angle of mandible**- Dental: odontomas, supernumerary teeth, & impacted teeth- Skin: epidermoid cysts | - Post. Md- often < 30y.o- Slight male > female- **VERY painful**, pain usually is not relieved by aspirin- Extragnathic and jaws  | - Most perm posterior MD - < 30 yo - **pain and swelling (66% pts)** |
| **Radiographic** | - Well defined radiopacity - Small lesions indistinguishable from idiopathic bone sclerosis or sclerosis secondary to inflammation |  | - Mixed w/ varying amount of patchy opacity- Well ill-defined- **> 2cm** | - Radiopacity fused to root(s)- some **root resorption**- thin radiolucent rim at periphery- loss of normal PDL |
| **Histology** | -Normal bone with variable amount of fibro-fatty marrow |  | - Bone formation from large osteoblasts- Very vascular | - Very similar to osteoblastoma, but cells are cementoblasts and lesion is fused to cementum of tooth |
| **Tx/Prognosis** | - Small: watch. **Slow** continued growth is normal - Large: cosmetic or functional problems are surgically removed | - Prophylactic colectomy b/c colonic polyps are pre-malignant | - Local excision- Rare recurrence | - Removal of lesion and root(s) or tooth- Does not recur if completely removed |

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|  | **Osteosarcoma (OS)** | **Chondrosarcoma (CS)** | **Metastatic****Malignancies** **to the Jaws** | **Ewings Sarcoma** |
| **Def** | - **The most common primary of malignancy of bone under 40 yo****(vs. >40 y.o. multiple myeloma)** | - Half as common as osteosarcoma | - The **most common cancer** found **in bone** is **metastatic carcinoma**- Jaws: most common carcinomas mets are breast, lung, thyroid, prostate and kidney | - Relatively common primary malignancy of bone (1=os, 2=cs, 3=Ewings)  |
| **Clinical Feat.** | - md=mx- long bones: 10-20 y.o- jaws ave 33 y.o.- Sx of swelling and pain (also loose teeth, paresthesia) | - mx and maxillary sinus>>md- long bones: over 50 yo- jaws: wide age range- Sx of swelling- Pain is **unusual**! (also loose teeth, nasal, sinus or ocular symptoms) | - MD>>>MX - elderly- sx vary from Asx to pain, swelling, paresthesia, loose teeth | - md>mx - teens- Whites>>>>Blacks - **VITAL teeth!!!**- any bone, jaws are uncommon- paresthesia, loose teeth, pain, swelling, sometimes soft tissue breakthrough, fever, elevated white cell count and ESR (mimicking infection, osteomyelitis on radio) |
| **Radiographic** | - **poorly** **defined borders** - vary in density (radiolucency, radiodensity or mixed – “**Moth eaten**”)- Spiking root resorption- **sunburst** periosteal rxn to cortical infiltration- Symmetric **widening of pdl** around involved teeth | - same as Osteosarcoma | - Most often ill defined radiolucency- Sometimes resembles perio dx or cyst or widens pdl - Some present with mixed lucent/opaque (breast and prostate)  | - Ill defined radiolucency- **onion skin** periosteal reaction |
| **Hist** | - pleomorphic round or spindle cells producing bone- May also produce fibrous ct or chondroid | - cartilage (condyle only, if somewhere else, malignant)- amount of cellular atypia and pleomorphism varies w/ grade of tumor |  | - undifferentiated round cells- dx usually requires special studies for immunophenotype (e.g.CD99) or genetic alteration (t 11,22) |
| **Tx/Prognosis** | - Improved survival w/ pre and post op chemotherapy - Radical surgical resection- 30-50% survival - Local disease control is usually the problem (local recurrence) more often than distant mets (lungs and brain) | - Radical surgical resection- Chemo **only used as last resort** in unresectable tumors- Prognosis depends on size, location and tumor grade - Survival data poor – but sure of late recurrence (15 yrs or more), so the pt must be followed for lifetime- local disease control is usually the problem (local recurrence) much more often than distant mets | - poor prognosis- stage IV disease- <1 year survival unless solitary met (tx with surgery or RT) | - surgery and chemo, +/-RT- The more distal the tumor(s) the better the prognosis- Overall prognosis much improved w/ multimodality tx to 40-80% survival |