Study Questions

Bone Pathologies:

* **Bisphosphonate-Associated Osteonecrosis:**
  + BON is associated with **2nd generation bisphosphonates** because 2nd gen bisphophonates have **very long half lives** (i.e. half life of fosamax is 12 years) and get incorporated into the skeleton
  + Factors that increase risk of BON in the mandible include
    - Dental surgery, dental infections/disease, oral trauama, periodontitis
    - Concomitant **chemotherapy** or **corticosteroids**
    - **Age >65 years**
    - Presence of bony tori/exotoses
    - Diabetes, smoking/alcohol use, poor oral hygiene
    - Duration of Rx use for **>3 years**
  + Etiology of BON:
    - Bisphosphonates decrease **osteoclast activity**, which normally includes the repair of microfractures and the resorption of foci with nonviable osteocytes, allowing the **accumulation of microfractures** and the persistence of **osteocytes past their 150 day lifespan**
  + Management of BON includes **conservative** management where the goal of therapy is to **stop pain** and **infection** (if present)
    - **Asymptomatic** patients:
      * **Chlorhexidine** rinse daily
      * Smooth edges of rough bone
    - **Symptomatic** patients:
      * Treat with **chlorhexidine** and **antibiotics**
        + Amoxicillin +/- metronidazole
        + Clindamycin, azithromycin
* **4 genetic diseases of bone**:
  + **Osteogenesis imperfect** – **AD** (**type I**) or **AR** mutation of **chromosome 7** and **17**
  + **Cleidocranial dysplasia – AD** mutation of **chromosome 6**
  + **Osteoporosis** (infantile and adult)
  + **Cherubism**
  + **Note**: **Gardner’s syndrome** is an **autosomal dominant** genetic mutation of **chromosome 5** that clinically produces **osteomas**
* **Osteogenesis imperfect**
  + OI is a disorder of **collagen maturation**
  + All 4 types of OI produce the following **craniofacial,** **teeth** and **ocular findings**
    - **Class III malocclusion** (due to **maxillary hypoplasia**)
    - (sometimes) **florid osseous dysplasia-like bone changes**
    - Clinical and radiographic evidence of **dentinogenesis imperfect** in primary and permanent teeth
      * Produces opalescent teeth
    - Classic ocular presentation of OI includes **blue sclera**
* **Cleidocranial dysplasia**
  + CCD is a condition that affects **osteoblastic differentiation**, affecting primarily membranous bones (clavicle and skull)
  + Clinical features of CCD include:
    - **Clavicles**:
      * **Hypoplastic** or **aplastic**
      * **Unilateral** or **bilateral**
      * Increased **shoulder mobility**
    - **Skull bones**:
      * **Sutures** and **fontanels** remain open
      * **Wormian** bones (extra sutural bones) develop in suture lines
      * **Frontal** and **parietal** bone bossing
      * Produces large heads
      * **Hypertelorism** (increased length between 2 organs), **high palate**, **depressed nasal bridge**, **absent maxillary sinus**
    - **Teeth**:
      * many **unerupted** permanent and supernumerary teeth
      * often **misshapen** teeth
      * **retention of primary teeth** into adulthood
      * **absent cementum**
* **Osteopetrosis:**
  + A condition that affects **osteoclasts**, preventing their function and producing **osteosclerosis**
* **Paget’s Disease of the Bone**
  + PDB is a condition characterized by increased bone turnover/metabolism, affecting **white men** (esp. England) **over age of 40**
  + Clinically characterized by:
    - **Pain**, especially near joints
    - **Bowing** of long bones
    - **Enlarged** skull
    - Usually **polystotic**
  + **17%** of patients experience **jaw involvement** (primarily the body and alveolus) where the **maxilla** becomes **larger** than the **mandible**, there can be **sinus obstruction,** and there can be **bone expansion**
  + Radiographic findings:
    - **Early** phase: often **osteoclastic**
      * **Decreased bone density** and **increased radiolucency**
    - **Late** phase: **osteoblastic** stages with **patchy sclerosis** becoming **confluent** (“**cotton wool**”)
  + Histologically, the **active phase** presents with **alternating** phases of **resorption/osteoclastic activity** and **formative/osteoblastic activity** on the same bone trabeculae and very **vascular fibrous CT** replacing bone marrow
  + Diagnosis involves:
    - **Blood** test: **increased alkaline phosphatase** (a marker of osteoblastic activity) with **normal** calcium and phosphorous
    - **Urine** test: **increased hydroxyproline** and other markers of **bone resorption**
  + **Osteosarcomas** can develop (1-13% of patients), usually in the lower extremities and pelvis
* **Idiopathic osteosclerosis**
  + A **common** condition (5% of population) that affects **teens and young adults** (20’s)
  + Usually an incidental finding; **asymptomatic** with **no bone expansion**
  + Radiographically presents as a **well-defined**, **rounded** or **triangular radiodensity** that is **uniformily opaque** with **no lucent portions**
  + **No treatment**
* **Focal osteoporotic bone marrow defect**:
  + A condition which affects **women (3:1)** and is often found in the **posterior mandible** (70%)
  + It is **asymptomatic** and does **not expand bone**
  + Radiographically presents as a **radiolucency** with **ill-defined borders** and **fine trabeculations**
  + **No treatment**
* **Langerhan’s Cell Histiocytosis**
  + A condition with the monoclonal proliferation of **histiocyte-like cells (Langerhan’s Cells)** with polyclonal infiltration of **MNGC**, **eosinophils,** and other **chronic inflammatory cells**
  + 3 forms of LCH:
    - **Eosinophilic granuloma of bone**: can be monostotic or polystotic
    - **Chronic disseminated histiocytosis/Hand-Schuller-Christian disease**:
      * Affects **bone, skin and viscera**
      * Presents as a **triad** of symptoms:
        + **Bone lesions**
        + **Exophtalamos**
        + **Diabetes insipidus**
    - **Acute disseminated histiocytosis/Letterer-Siwe disease**
      * Prominent **visceral, bone marrow, and skin** involvement
      * Usually in **infants**
  + LCH is a **childhood** disease, with **50%** of patients being **<10 years old**
  + **Jaw** lesions develop in **10-20%** of cases, usually in patients **>20 years old**
  + Radiographic presentations:
    - **Well-defined, non-corticated, punched out radiolucencies**
    - In jaws, is sometimes **ill-defined** due to **destruction of alveolar bone** from the crest
      * Mimics severe periodontal disease
  + Histologically, LCH is diagnosed when samples present as:
    - **CD1a** - positive
    - **S100** - positive
    - **Peanut agglutinin** – positive
    - **Birbeck granules** – positive
* **Central Giant Cell Granuloma**
  + Clinically presents in:
    - **Wide** age range, though **60%** of patients are **<30 years old**
    - Presents more often in **women**
    - Usually located in the **mandible**, **anterior to 1st permanent molar**
    - **Asymptomatic** and grows **slowly**, though the more aggressive course of the disease causes rapid growth, bone expansion, pain, root resorption, parasthesia, and perforation of the cortical plate
  + Radiographically presents as a **unilocular or multilocular radiolucency** with **well-defined**, **possibly corticated** margins
  + Histologically, CGCG presents with **MNGC**’s, which are mostly likely **osteoclasts**
  + **Normal** lesions are treated with surgery
    - **Curettage** leads to **20% recurrence rate**, but responds well
  + **Aggressive** lesions are treated with **intralesional** or **systemic chemotherapy** with **corticosteroids or calcitonin** (respectively)
* **Cherubism**:
  + A condition that begins to present in patients between **ages 2-5** and **stabilizes at puberty**
  + Clinically present with an **expanded mandible** and **orbital bone**, which produces a “cherubic” facial appearance
  + Radiographically, the lesion presents as **bilateral, multifocal, multilocular radiolucencies**
  + Diagnosis is made utilizing information gained from **radiography**, **family history**, and **biopsy**
    - need to **rule out** **hyperparathyroidism** and  **CGCG** (which are found in this disease’s histology)
* **Traumatic Bone Cyst**:
  + AKA:
    - **simple bone cyst**
    - **solitary bone cyst**
    - **idiopathic bone cyst**
  + Not a true cyst, but is rather an **empty or fluid-filled bone cavity**
  + TBC’s are reported in **almost all bones** of the body
  + **Jaw** lesions develop in **10-20 year olds** (**rare** to find in patients **>35 years old**) with a slight **male predilection**
    - Radiographically presents as a **unilocular radiolucency** with **variable definition** and **scallops between roots** (without devitalizing or resorbing them)
  + **Rare** for TBC’s to produce **pain, parasthesia or bone expansion**
  + **Surgical exploration** of the cavity often causes repair of the lesion
* **Aneurysmal Bone Cyst:**
  + An **intrabony accumulation of blood** with growth of **reactive fibrous CT and bone**
  + ABC’s are more common in **long bones** and **vertebrate**, but presents in the **posterior mandible** of **children and young adults**
  + ABC’s cause **pain, rapid swelling, ballooning expansion of bone**, but **rarely parasthesis, tooth mobility or root resorption**
  + Treat by **surgically curetting** out the “**blood-soaked sponge**” lesion
    - The defect usually heals within **6 months**
    - **Cryosurgery** may be needed
* **Benign Fibro-osseous Lesions of the Jaw**:
  + Contains 3 categories of diagnoses:
    - **Fibrous dysplasia**
    - **Cemento-osseous dysplasia**
    - **Ossifying fibroma**
  + **Fibrous dysplasia**:
    - Clinically appears as **tumorous swellings of bone(s) and skin with endocrine abnormalities**
    - Usually diagnosed in **teenage years**
  + **Cemento-osseous dysplasia** (in jaws only)
    - 3 types:
      * **Periapical** COD
      * **Focal** COD
      * **Florid** COD
  + Radiographic presentations:
    - **FD**:
      * **Poorly defined uniform radioppacity with “ground glass” trabeculae appearance**
    - **CODs:**
      * **PA-COD**: **radiolucency** at the apex of **vital** teeth; identical looking to a periapical granuloma/cyst
      * **Florid COD**: adjacent **radiolucencies** fuse to form a **continuous lucency at the apex of several teeth**
      * **Mature** lesions: **mixed** **radiolucent rim with radioopaque center**; no fusion of teeth
    - **OF:**
      * **Early** lesions: **well-defined unilocular radiolucency** or a **mixed lesion with varying radioopacity**; some have **sclerotic borders**
      * **Later** lesions: can become **large**, causing **root divergence** and **bowing of inferior border of mandible**
  + **Fibrous dysplasias** and **ossifying fibromas** can cause **bone expansion**, but **all COD’s** do **not**
  + Locations of lesions:
    - **FD: mandible** is the most common location
      * Usually **below** the **inferior alveolar canal**, thus displacing it superiorly during bone expansion
      * If found on the **maxilla**, it is found **below** the **maxillary sinus**, thus displacing it upward to possibly obliterate it
    - **CODs:** only occur on the **jaws**, in **tooth-bearing areas**
    - **OF:** most common in the **posterior mandible**
  + Histologically:
    - FD:
      * A haphazard mix of **immature/woven bone** in a **fibrous CT stroma**
      * “**Chinese-character**” trabeculae
    - COD:
      * **Fibrous CT**, **metaplastic immature/woven bone** with **cementum-like droplets**
    - OF:
      * Very similar, if not identical to CODs
  + Etiology:
    - FD: Found in **teens**
    - COD:
      * PA-COD: found in **40 year old African-American women**
      * Florid COD: found in **50 year old AA females**
      * Focal COD:found in **40 year old African-American women**
    - OF: Found in **females** in their **20’s-30’s**
  + Treatments:
    - FD: **Comestic shave down** once lesion is **mature**
    - COD: no treatment; surgery can cause **osteomyelitis**
    - OF: **enucleation**
* **Osteomas:**
  + Clinically presents as a **periosteal (on bone)** or an **endosteal (in bone)** lesion
  + Radiographically presents as **well-defined radiolucencies** either **in** or **on bone (endosteal, periosteal,** respectively)
* **Gardner Syndrome**:
  + Oral presentation includes:
    - **osteomas** in any part of the **mandible,** but most often at the **angle of the mandible**
    - **odontomas**
    - **supernumerary teeth**
    - **impacted teeth**
  + Oral manifestations of GS is **not** the most important determinant in the patient’s overall health because GS is primarily a disease of **familial colorectal polyposis**
* **Osteoblastoma**:
  + A **rare benign bone neoplasm** that is **very painful**
  + Radiographically presents as a **mixed** lesion with **varying amounts of patchy opacity**
    - >**2cm** large
* **Cementoblastoma**:
  + A **neoplasm of cementum** that causes **pain and swelling** (66% of patients) in the **posterior mandible** of patients **<30 years old**
  + Radiographically presents with the lesion **fused to the roots** with a **radiolucent rim** at the periphery
* **Hemangioma of the Bone**
  + Usually found in patients that are **10-20 years old**, though it can occur in adults as well
  + If the lesion is symptomatic, it will present as **pain, swelling and spontaneous bleeding from the sulcus**, which can mimic severe periodontal disease/abscess
* **Osteosarcoma**:
  + The most common **primary malignancy of bone** in patients **<40 years old**
  + Radiographic appearance, unique to osteosarcomas:
    - **Spiking root resorption**
    - **Sunburst periosteal reaction** (to cortical infiltration)
    - **Symmetric widening of PDL**
  + Histologic appearance:
    - **Pleomorphic round or spindle cells** that are producing **bone, fibrous CT, or chondroid**
  + Treatment includes **radical resection** and possible pre- and post-op chemotherapy
    - **30-50%** survival rate
* **Chondrosarcoma**:
  + **Half** as common as an osteosarcoma
  + Radiographic appearance:
    - Much like an osteosarcoma:
      * **Spiking root resorption**
      * **Sunburst periosteal reaction**
      * **Symmetric widening of PDL**
    - However, the lesions **vary in density** and have **poorly-defined borders**
  + Histologically, lesions have varying degrees of **cartilage** and **cellular atypia/pleomorphism**
  + Treatment includes **radical resection**, with chemotherapy as a last resort for unresectable tumors
    - Survival data is unreliable due to possible **late recurrences** (**>15 years** after surgery)
* **Metastatic Malignancies of the Jaws**:
  + The most common carcinomas to metastasize to the mandible are:
    - **Breast**
    - **Lung**
    - **Thyroid**
    - **Prostate**
    - **Kidney**
  + Most malignancies are most often **ill-defined radiolucencies**, except **breast and prostate malignancies**, which can present with **mixed lucent/opaque** lesions
* **Ewings Sarcoma**:
  + ES most often occurs in **white** **teenagers**
  + Clinically, ES can occur in **any bone**, including the **mandible** (which is **uncommon)**
    - Symptoms include **parasthesia, loose teeth, pain, swelling, soft tissue breakthrough, fever, elevated WBC count**
  + Radiographically, ES presents with **ill-defined radiolucencies**, with rare **onion skin periosteal reactions**
  + Treatment for ES includes **surgery and chemotheraphy** with possible radiation therapy
  + **40-80% survival rate**