LECTURE # : 14

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Lecture Outline:

- **Inherited and developmental disorders:**
  1. Osteogenesis imperfecta
  2. Osteopetrosis
  3. Cleidocranial dysplasia
  4. Achondroplasia

- **Fibro-osseous lesions:**
  1. Osseous dysplasia
     A. Fibrous dysplasia
        ~ Monostotic
        ~ Polyostotic
        B. Cemento-osseous dysplasia
        ~ Periapical cemental dysplasia
        ~ Focal cemento-osseous dysplasia
        ~ Florid cemento-osseous dysplasia
  2. Benign neoplasia
     ~ Cemento-ossifying fibroma

- **Inflammatory diseases of bone:**
  1. Osteitis:
     A. Alveolar osteitis (Dry socket):
     B. Focal sclerosing (condensing) osteitis:
  2. Osteomyelitis:
     A. Suppurative osteomyelitis
     B. Sclerosing osteomyelitis
     C. Chronic osteomyelitis w productive periostitis (Garre’s osteomyelitis)
  3. Periostitis:
     Chronic periostitis associated w hyaline bodies
  4. Osteoradionecrosis

- **Metabolic and endocrine disorders of bone:**
  1. Osteoporosis
  2. Hyperparathyroidism
  3. Ricket’s and osteomalacia
Inherited and developmental:

1. **Osteogenesis imperfecta**: is defined as Excessive fragility of bone due to a Defect in synthesis of type I collagen. This leading to an Inadequate or less formation of bone → generalized osteoporosis. Pt susceptible to fracture due to mild trauma

Features:

Slender, weakness, deformity of bone and fracture tendency, Osteoporosis, Dwarfism

Thin skull and bulge over ears
Thin sclera and may be colored (sometimes blue)
± deafness of deformity in the middle ear
joint hypermobility, shortening of pt similar to dwarfism
heart valves defects (for our dental concern)

4 main types that are variable in severity:
Type I: AD, Blue sclera, ± Dentinogenesis imperfecta → mild
Type II: AD → lethal; pts die directly after birth
Type III: AD or AR, Dentinogenesis imperfecta → pts live but with progressive deformity that increases throughout life
Type IV: AD, White sclera, ± Dentinogenesis imperfecta

** not all Pt have blue sclera or OI, have DI, and some may be seen in your daily clinic

**Dental aspects:**
1. Dentinogenesis imperfecta; dental malformation grayish, bluish, translucent primary and secondary teeth all surfaces, severe attrition, no pulp, no endo TTx, short root, mobility, loss of teeth.

NOTE: you need to revise it

2. Extractions → complications: fracture of bone

3. Class 3 malocclusion; deformity in maxilla

4. Impaction of 6s and 7s

5. Difficulty in intubation for GA due to deformity

2. **Osteopetrosis**: (marble bone disease)
Features:
• Solid dense but brittle bones
• Inactivity of osteoclasts (no bone resorption) excessive bone formation on expense of bone marrow spaces causing anemia
• Excessive bone formation but woven
• Fracture
• Anaemia and hepatosplenomegaly

RGx:
_ Generalized \( \uparrow \) in bone density
_ Cortical = medullary ; no medullary spaces only dense bone
_ Marked radio-opacity of skull base
_ Greatly reduced sinuses, for example reduction in maxillary sinus space.
_ greatly reduced skull foramina which causes pressure on the cranial nerves that pass these foramina leading to neuropathy and facial palsy.

Dental aspects:
1. Invisible roots because of density of bone
2. Unerupted teeth
3. spread of infection because of the dense bone(reduce vascularity) leading to Osteomyelitis
4. fracture tendency

3. Cleidocranial dysplasia:
Features:
• mostly AD/ some Sporadic cases
• affecting Face, skull & clavicles
• Frontal & occipital bossing, open fontanells & sutures \( \rightarrow \) bulging deformity
• Underdeveloped midface, depressed maxilla & nasal bridge
• Normal size mandible however class 3 malocclusion
• Partial or complete absence of clavicles

Chest radiographs:
Partial or complete absence of clavicles \( \rightarrow \) ability to depress shoulders closer to midline

Dental aspects:
1. Retention of primary teeth
2. Multiple unerupted teeth
3. Multiple supernumerary teeth & dentigerous cysts
4. Thin roots(why???) and hypocementosis ( in cellular cementum)

Q: is there a relationship between hypocementosis and delayed eruption?

4. Achondroplasia:
Features:
• Most common genetic skeletal disorder
• Short-limbed dwarfs
• Cartilage proliferation in epiphyses & skull base
• Head & trunk of normal size however Defective middle 1/3 of face (nonproportional unlike the dwarfism that is caused by defect in growth hormone secretion)
Malocclusion class 3

**Fibro-osseous lesions:**

- **Definition:** fibrous tissue replacing bone then gradual calcification occurs.
A variety of diseases which, histologically, are characterized by the replacement of normal bone by cellular fibrous tissue within which varying amounts of predominantly woven bone and acellular islands of mineralized tissue develop. **increase in size of bone but the content is fibrous tissue then calcify to bone at the end.**

1. **Fibrous dysplasia:** (most common; as in 1 or 2 cases may be encountered all through your career life)
   - **Monostotic FD:** unilateral affecting one bone
     - 80% of cases
     - One bone
     - Max > Mand however not exclusive to facial bones and can also be found in lower and upper limbs and femur
     - could be Craniofacial FD affecting more than one bone and the adjacent
     - diagnosed during the growth period in Childhood, arrest in adulthood
   - **Clinically:**
     - Clinical presentation: Facial asymmetry; best inspected from above the head
     - Not well defined in early stages, hard, smooth, round, painless persistent swelling, progressive enlargement of bone (so the DDx maybe bone tumor, odontogenic tumor or cyst, giant cell lesion, fibro osseous lesion)
     - Normal skin, don’t confuse it with infection,
     - Disturbs function & causes malocclusion
     - Extension to max sinus if in maxilla
     - can affect mandible mostly in posterior Mandible
   - **Rx:** depending on the stage
     - Radiolucent → radio-opaque

     Early → radiolucent (fibrous tissue)
     Mid → mix of radiolucency and radioopacity (calcify)
     Late → ground glass or orange peel or thumb print pattern appearance. Not a normal trabecular appearance
     - should take a biopsy
     - **Border:** gradual transition into adjacent
     - Displacement of roots, obscured lamina dura; the more the maturity of the disease the more the loss of lamina dura & thin PL space in the affected area
     - Superior displacement of mandibular canal

   **DD:**
   Cementoblastoma; radioopacity attached to the resorbed roots
   Condensing ostitis;

   - **Polyostotic FD:**
     - Expansion in multiple bones
     - Segmental or one side
• F>M
• Albright syndrome: FD affecting multiple bones + café-au-lait spots on skin + endocrine defects; hyperthyroidism, hyperparathyroidism, precocious puberty

Histology for two type:
• **Initially**: cellular CT replacing normal bone
• **Gradually**: deposition irregular, immature trabeculae (osteoclast like giant cell trimming bony trabeculae), (not arrange for function) delicate woven bone = *Chinese characters*
• Spherical calcification
• calcified material like cementum
• Border: ill defined, no capsule gradual change into normal bone.
• **Late stage**: change of woven to lamellar bone

**DD:**
Giant cell lesions of bone; central giant cell granuloma, brown tumor (hyperparathyroidism), Cystic lesions (Aneurysmal bone cyst).

**Blood chemistry:**
± increase in Alkalinephosphatase (ALP) in PFD bc of the increase in bone activity

**Aetiology:**
• unknown, developmental occurring in growth period (childhood),
**Prognosis:**
• Fibrosarcoma 1% when treated by radiotherapy
• Cosmetic surgery in adult stage
• MFD does not spread to other bones or transform into PFD (reassurance)
• Monostotic doesn’t convert to polystotic

**2. Periapical cemental dysplasia:**
• Asymptomatic, diagnosed radiographically by chance (incidental), no deformity present, no expansion, no pain, no swelling, no root resorption
• Middle-age F, African-Americans
• Below apices of mandibular incisors
• Multiple radiolucencies
• Vital → this point cuts out the diagnosis if nonvital we would have thought of other DDs such as: chronic alveolar abscess, periapical granuloma, cystic lesion.
• Very rare and not found in our region

**Rx:**
_ Well-defined radiolucency below apices separated by normal PL
_ Increasing radio-opacity
_ Thin radiolucent margin like a fibrous capsule
_ root resorption is rare not like the cementoblastoma which is also not multiple like this periapical cemental dysplasia
**Histology**
_ Cellular CT & then gradual calcification

****focal cemental dysplasia:**
A localized variant of periapical cemental dysplasia that is not multiple, white people, posterior mandible, well defined, starts as radiolucency then mix then radiopaque, normal PL space
3. Florid cemento-osseous dysplasia

- Extensive Periapical cemental dysplasia ≥ 3 quadrants
- More severe

Clinically:
- Same group of pts → African American
- Asymptomatic unless infected → osteomyelitis
- Expansion is present (recall; no expansion in PCD)

Rx:
- Radio-opaque, irregular masses
- Frequently symmetrical
- May involve 4 quadrants

Histology:
- = PCD → Cellular CT & then gradual calcification; irregular abnormal bone trabeculae

4. Cemento-ossifying fibroma or aka ossifying fibroma: (benign tumour)

Clinically:
- Similar to Fibrous dysplasia histologically & clinically
- Bony enlargement

Similarities:
- Slowly growing, painless swelling, gradual calcification on Rx.

Differences:
1. Age group: 20-40ys (FD → childhood)
2. Well defined radiolucency
3. Radiolucent rim (easily resected) \ spherical calcification more than FD
4. Numerous cementicles histologically (chinese letters in FD)
5. Mandibular molar & premolar region (FD → maxilla more)

Rx:
- Well defined radiolucency
- Gradual calcification
- Radiolucent rim

Histology:
- Cellular FCT
- Trabeculae of bone & numerous cementicles
- Outer zone of FCT

One type of cemento-ossifying fibroma is: Juvenile ossifying fibroma(tumor).
- Rapidly growing
- < 15 ys
- Richly cellular & mitotically active FCT w trabeculae of woven bone
- ↑ LRR (high recurrence rate).