CASE PRESENTATION

PATIENTS NAME: Mr. Amol Chandanshive

O.P.D NO : 91538

DATE: 10/ 7/ 2013

AGE: 26 years

SEX: Male

OCCUPATION : Farmer

ADDRESS: Latur

CHIEF COMPLAINT:

HISTORY OF PRESENTING ILLNESS:

- Alright 3years back.
- Then he experienced pain- continuous & sharp.
- Followed by intraoral swelling on the buccal side which was small in size & soft in consistency.
- Visited to the dentist for the same 3 years back.
- Pain relieved on medication.

- But the swelling did not subside & gradually it became firm as compared to previous episode.
- Enlarged to the present size.
- The patient overlooked it for a long time.
- No h/o fever, presence other swelling, pus discharge, impairment of function, loss of body weight, & trauma, referred pain, hot fomentation, balm application.

- **PAST MEDICAL HISTORY**: No relevant past medical history.
- **FAMILY HISTORY**: Not relevant.
- **PAST DENTAL HISTORY:** H/O extraction with 36, 15years back.

PERSONAL HISTORY:

- HABITS: Brushes his teeth once in the morning with hard bristle toothbrush in horizontal scrub method.

- DELETERIOUS HABITS: Not present.

- DIET: Mixed.
- APPETITE: Normal
- BOWEL/BLADDER: Regular.
- SLEEP PATTERN: Normal.

CLINICAL EXAMINATION:

✓ **General examination :**

- General appraisal:

1. Built : Asthenic.
2. Pallor : Absent.
3. Clubbing & cyanosis: Absent.

- Vital signs :

1. Temperature: 98.2 ˚F.
2. Pulse : 85 beats/min.
3. Blood pressure: 118/72 mm Hg.
4. Respiratory rate: 16 cycles/min.

✓ EXTRA ORAL EXAMINATION:

- Face :Asymmetrical.
- Lips : Compe
- TMJ : No ten heard.
- Lymph nodes : Submandibular lymph nodes are palpable on left sides, single, soft in consistency, ovoid in shape, mobile & are non tender.

LOCAL EXAMINATION:

- ✓ **Inspection**

- Solitary with no change in colour overlying the skin of swelling.
- Extending horizontally from corner of the mouth to 3cm in front of ala of the ear, vertically from infraorbital ridge to corner of the mouth.
- Surface of the swelling is smooth with diffuse margins.
- Pulsations were not visible.
- No signs of pus discharge.

✓ Palpation :

- Local temperature was not raised.
- All the inspectory findings were confirmed.
- Not tender on palpation.
- Ovoid in shape, & was 4X3 cm in dimension.
- Surface of the swelling was smooth with diffuse margin.
- Bony hard in consistency.
- Fluctuation not seen.
- Not reducible & not compressible.
- Not fixed to the overlying skin.
- Paresthesia of lip is not seen.

INTRAORAL EXAMINATION

- Inspection :

  Soft tissue

Palpation :

- All the inspectory findings were confirmed by palpation.
- Swelling was non tender on palpation.
- Swelling was ovoid, smooth, & extending from 23 to 27.
- Hard in consistency.
- Swelling showed no fluctuation.
- Swelling was not reducible & not compressible.

Hard tissue

- Teeth present:
- Missing teeth- 36
  - Root piece – 24, 25, 15,46
  - Stains +
  - Spacing is seen between 31 & 41
  - Midline is shifted towards left side.

Provisional diagnosis

- Benign odontogenic tumour

DIFFERENTIAL DIAGNOSIS

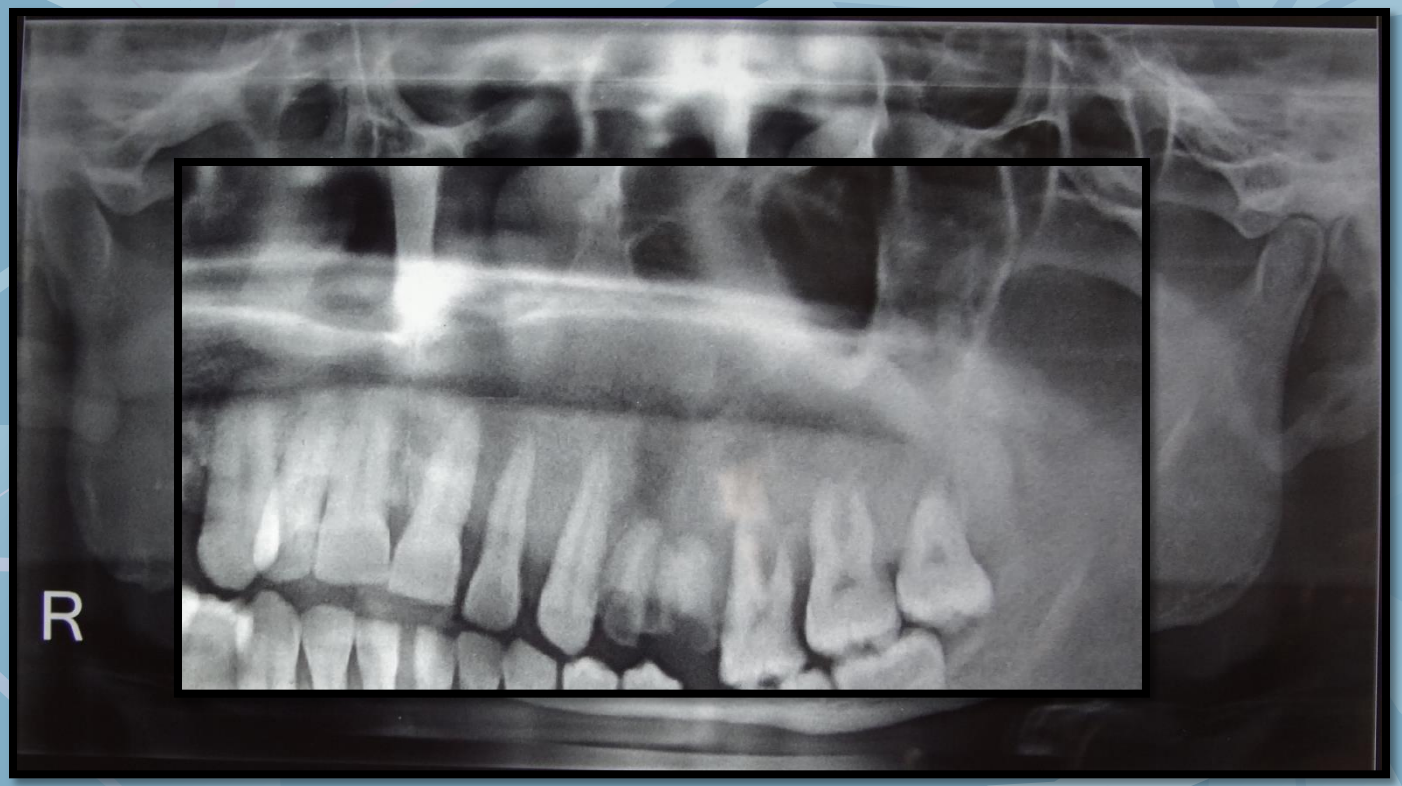
- CHRONIC DIFFUSE SCLEROSING OSTEOMYELITIS
- CEMENTO – OSSIFYING FIBROMA
- FIBROUS DYSPLASIA
- OSTEOSARCOMA
- PAGETS DISEASE

INVESTIGATIONS

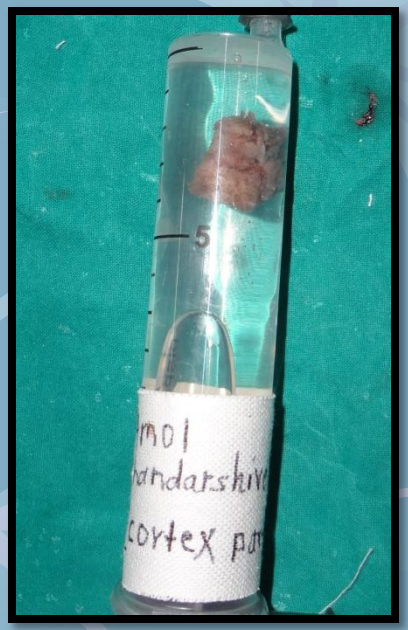
- IOPA
- Panaromic radiograph
- Vitality of 22,23,26,27.
- Complete Blood count
- Incisional biopsy

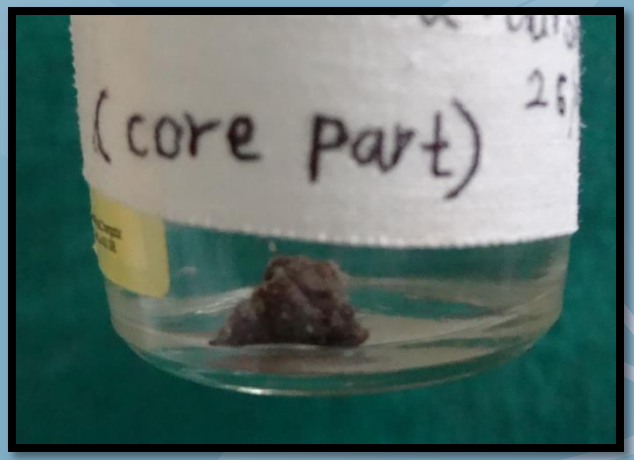
IOPA

OPG



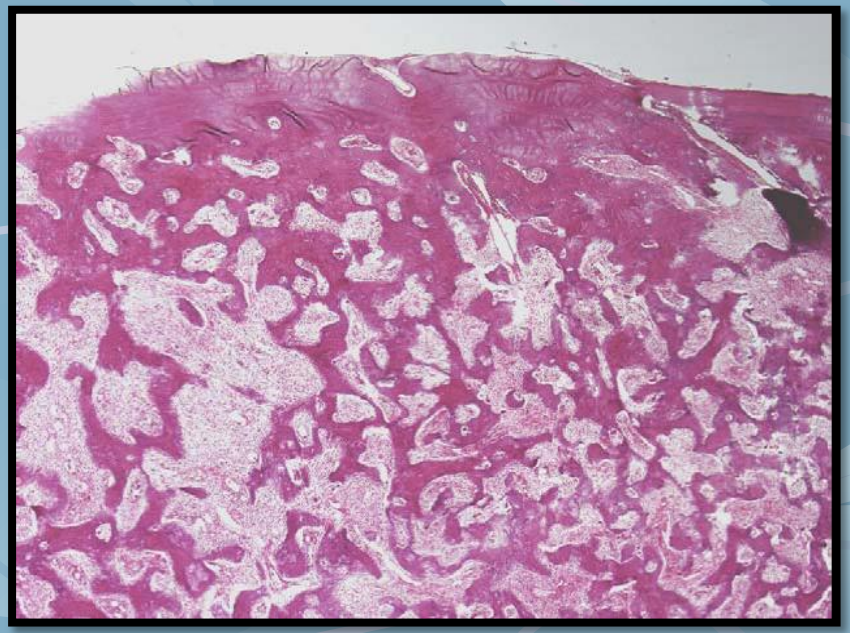
Incisional biopsy





HISTOPATHOLOGIC DIAGNOSIS

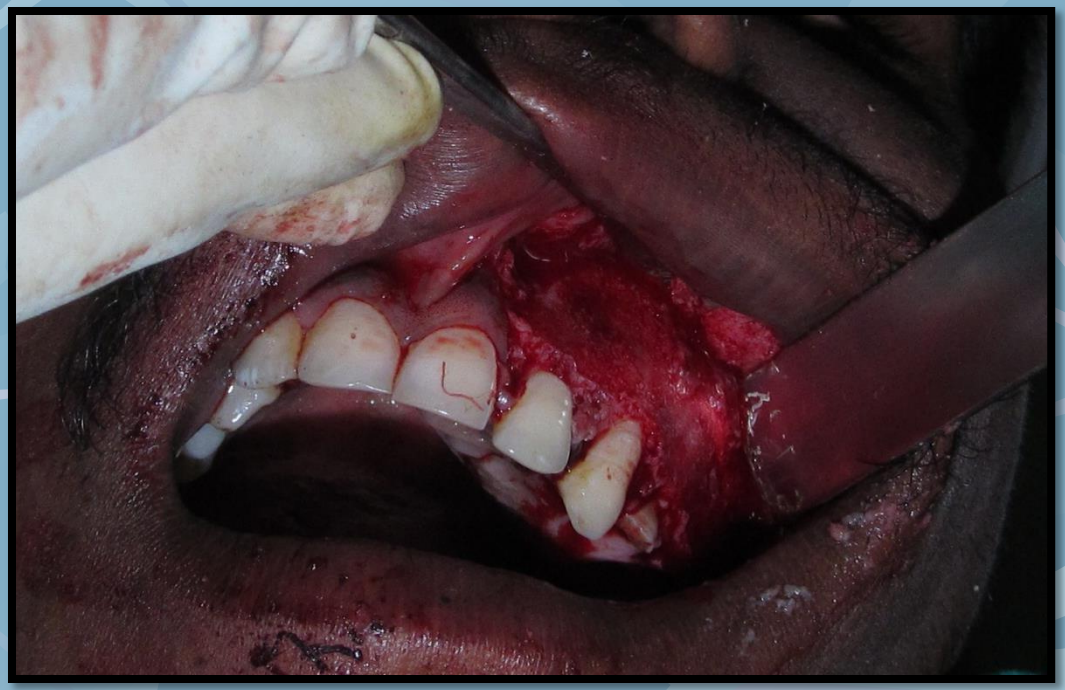
FIBROUS DYSPLASIA



Final diagnosis

- FIBROUS DYSPLASIA

Management



FIBROUS DYSPLASIA

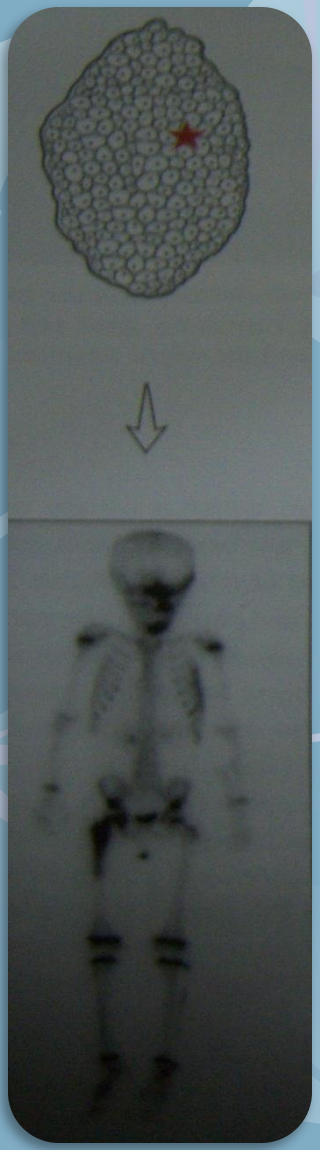
DISCUSSION

DEFINITION

Fibrous dysplasia is a disease of bone maturation and remodelling in which the normal medullary bone and cortices are replaced by a disorganized fibrous woven bone.

PATHOGENESIS:

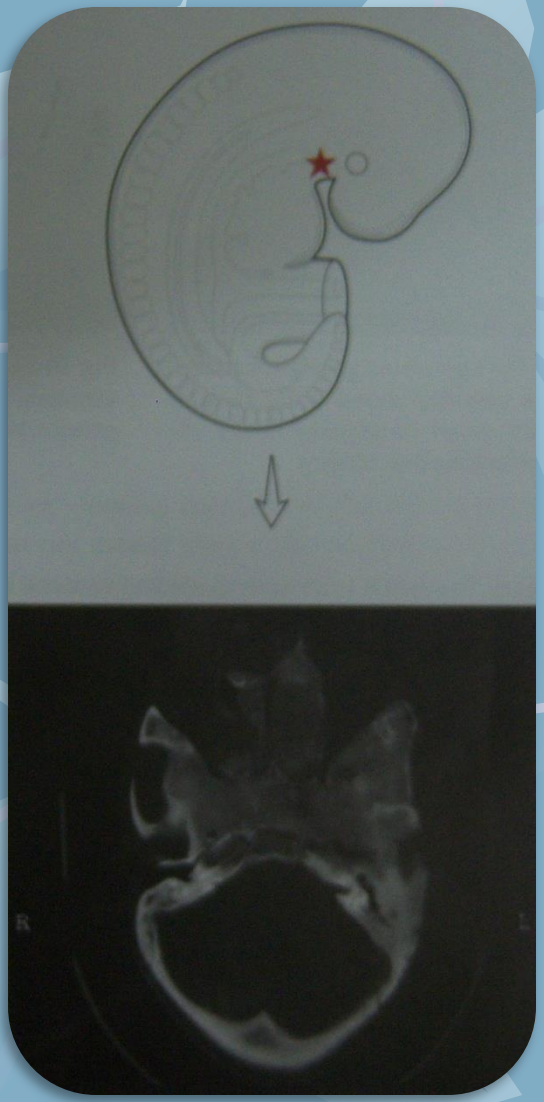
- Defects in bone maturation that begins in the embyo.
- Histodifferentiation phase of embryo, genetic mutation or deletion of gene GNAS-1(guanine nucleotide binding protein α- stimulating polypeptide-1) that encodes for an intracytoplasmic transducer protein required for bone maturation.
- Daughter cells of this aberrant cell will lack this transducer & produce only immature bone.



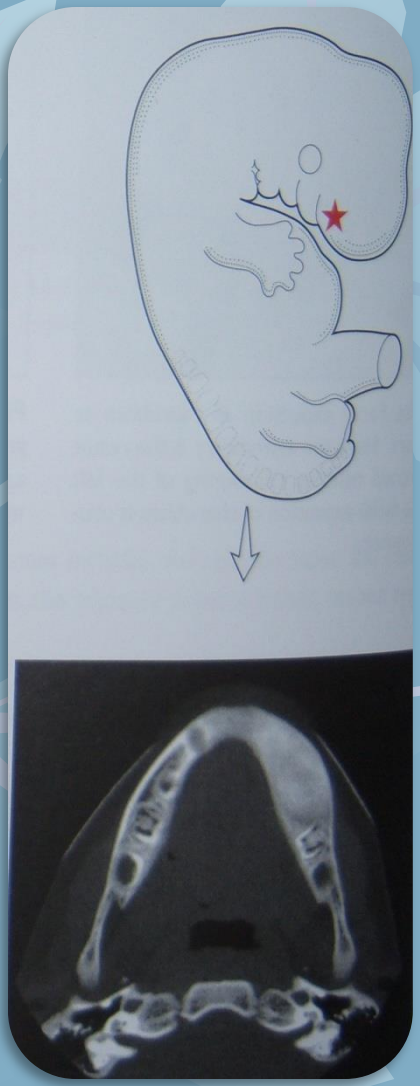
- If the genetic defect occurs in even earlier phase-
  Produce daughter cells of divergent differentiation
  i.e, some will migrate into bone primordia
  some into skin primordia &
  some into endocrine gland primordia

- Thus produce Mc-Cune- Albright syndrome or
  Jaffe-Lichtenstein type of PFD

Genetic alterations are thought to occur before 6th week of fetal life



- If genetic defect occur at 6th week-
- Daughter cells will be localized to one region producing craniofacial type of FD



- If genetic defect occurs slightly later-
- Daughter cells will be even more localized & produce MFD

Classification

- ***Sub clinical fibrous dysplasia***

  Unsuspected lesion of fibrous dysplasia comes to light accidentally on routine radiographic examination, without any clinical evidence of the suspected disease.

Clinical Features

Monostotic fibrous dysplasia

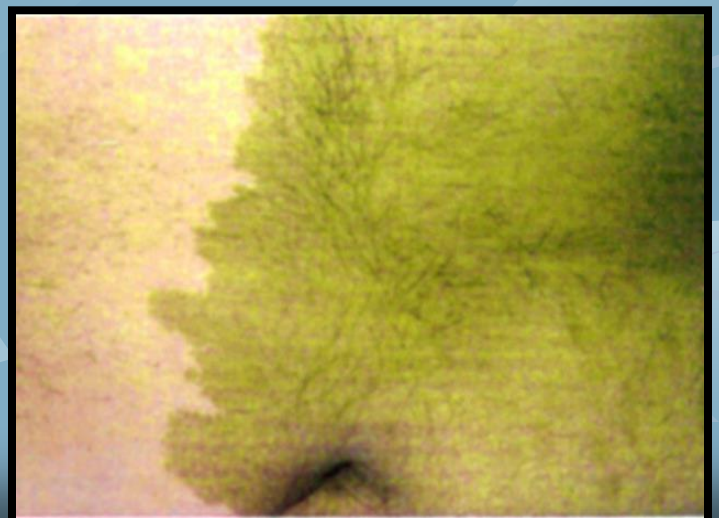
- Most frequent sites are ribs, femur, maxilla and mandible.

Fibrous dysplasia discovered in young patients, usually in children younger than 10 years affecting both the sexes equally.

- **<u>Polyostotic fibrous dysplasia</u>**
- Involvement of multiple bones
- female to male ratio of 3:1.
- Bowing or thickening of long bones.
  Symptoms—recurrent bone pain.
- ***Skeletal lesions—they*** may be unilateral in distribution or may involve nearly all bones of the body.
- Complication—spontaneous fracture.

Polyostotic fibrous dysplasia (Jaffe's type)

- ***Skin lesions-*** *the* skin lesions consist of irregularly pigmented, light brown melanotic spots, described as 'cafe-au-lait spot'.
  • Sites- back, buttocks, thighs, shoulders, chest, neck and face in the mentioned order.



Coastline of Maine

Albright's syndrome

- Endocrinal disturbances like precocious puberty, goiter, hyperthyroidism, hyperparathyroidism, Cushing's syndrome and acromegaly.

- Sex- exclusively found in females.

- **Albright's syndrome**

- ***Symptoms***- *vaginal* bleeding has been noted. Signs—secondary sexual characteristics such as pubic and axillary hair and development of breasts are evident by the age of 5 years.

- It may result in crippling deformities or fracture.

Oral Manifestations

Monostotic

- Sites- Maxilla, Premolar-molar area
- *Appearance*- Unilateral facial swelling, which is slow growing with intact overlying mucosa.
- Symptoms—swelling is usually painless but patients may feel discomfort.

- *Cortical plates*- enlarging deformities of alveolar process mainly buccal and labial cortical plates.
- *Mandible- in* mandible, it causes protuberant excrescence of the inferior border of mandible.
- Malalignment, tipping or displacement of teeth.

Craniofacial fibrous dysplasia

- Sites- if fibrous dysplasia extends to involve the maxillary sinus, the zygomatic process, floor of orbit and sometimes toward the base of the skull, it is known as craniofacial fibrous dysplasia.

- It results in severe malocclusion and marked facial deformity.
- Craniofacial lesions may lead to anosmia (loss of sense of smell), deafness and blindness.
- Signs—there may be proptosis of the affected eye.

- **Polyostotic (Jaffe's type)**

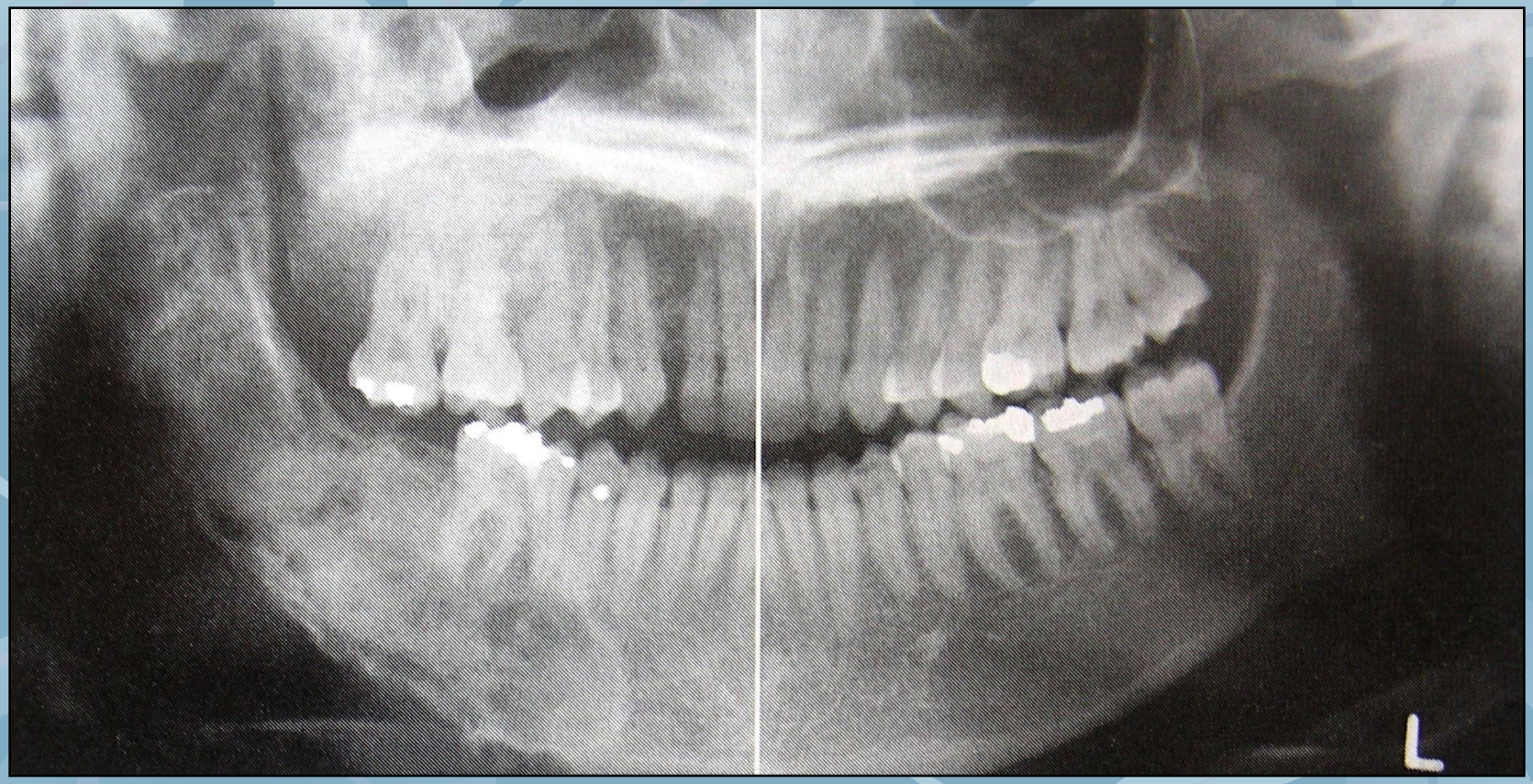
  • Expansion and deformities of jaws.

  • The eruption pattern of teeth is disturbed because of loss of support of the developing teeth.

  • Asymmetry of facial bones. There is ballooning of jaws, so there is gross enlargement and deformity.

  • In some cases, intraoral pigmentation can be seen.

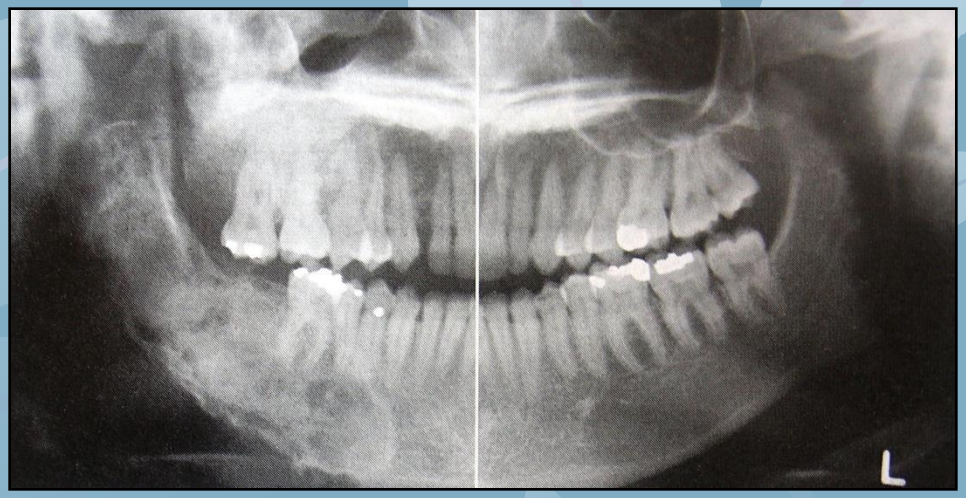
Radiographic Features



Lesions showing predominance of fibrous tissue

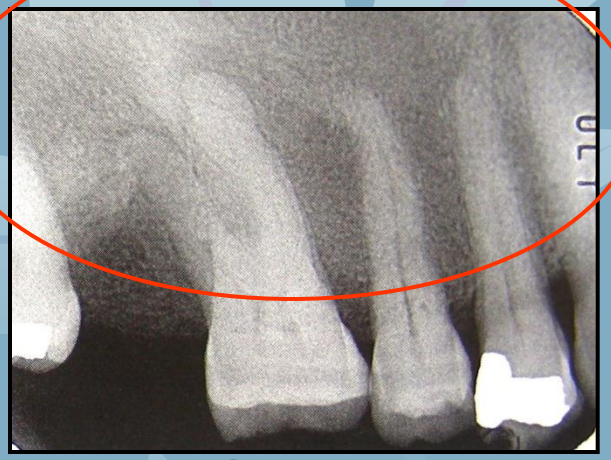
- Early—radiolucent with ill defined borders. The bony defect may be often unilocular but occasionally bony septa may be apparent creating an impression of multilocular cavity.
- Margins—margins may be well defined with a tendency to blend imperceptibly with surrounding normal bone.

- ***Lamina dura-*** *when* the lesion involves the apices of teeth there is loss of lamina dura or if retained, it has less density than normal.
- Teeth—resorption of roots and destruction of developing teeth



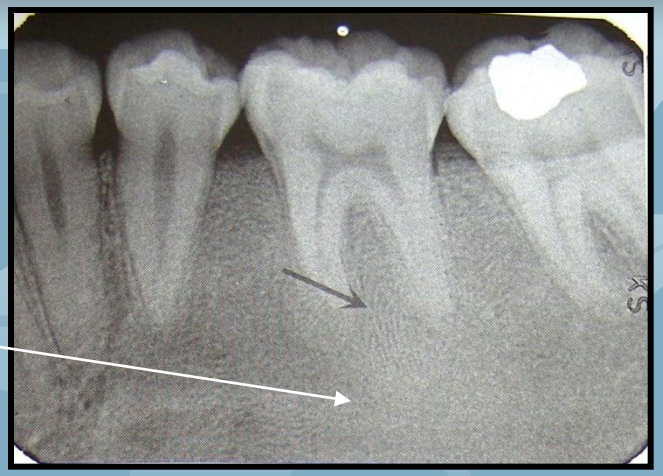
- ***Lesions showing mixed radiolucent and radiopaque appearance***
- Appearance—radiographic appearance of lesions with heterogeneous distribution of fibrous and osseous tissue show a mixed radiolucent and radiopaque appearance, depending on the maturity of the lesions.

Mature radiopaque lesions where bone is predominant.



- **Orange peel**—the radiograph shows bone of increased density. The normal structure of bone is replaced by a stippled appearance which resembles the ring of orange which is called as called as orange peel.

Fingerprint pattern



- ***Thumb print appearance-*** *when* mandible is affected, the vertical depth of mandible is increased. The inferior border of mandible appears as a ribbon like cortex. In some cases, the localized area over the cortex is lost and instead, there is a smooth curved downward projection of the inferior margins of the bone. The appearance resembles a ‘thumb print’, as if the bone had been soft and pressed upon by the thumb.

- ***Smoky mottled appearance***—*as* the lesions mature, dysplastic bony trabeculae increase in size and number and appear like smoky mottled radioopacities.
- ***Ground glass appearance***—*another* characteristic appearance of fibrous dysplasia is ground glass appearance, also termed as granular.

- It may demonstrate areas of whorled amorphous partially calcified materials that are well circum scribed.

According to Obisesan & others, radiographic appearance of FD of the skull can be classified into various types as

- ***Whorled plaque like type***—*in* this type, the matrix of the well circumscribed lesion is composed of plaques of amorphous material of intermediate radiodensity, which on close examination are seen to be arranged in whorled onion peel appearance.

- ***Diffuse sclerotic type***—*the* lesions of this show as homogeneous dense area, which gradually merges with the normal bone.

- ***Cyst like type***—*in* this type the lesions is radiolucent. It is unilocular or multilocular, more often multilocular with well defined margins.

- ***Pagetoid type***—*in* this type of lesions, the affected area of bone markedly expands and shows alternating areas of radiopacities and lucency, as those seen in Paget's disease of bone.

- ***Chalky type***—*it* manifests itself as a well circumscribed lesion consisting of an amorphous dense radiopaque material.

- **“Orange-peel”**

  alternating areas of granular density & lucency giving a radiographic appearance resembling the rind of orange.

Craniofacial fibrous dysplasia

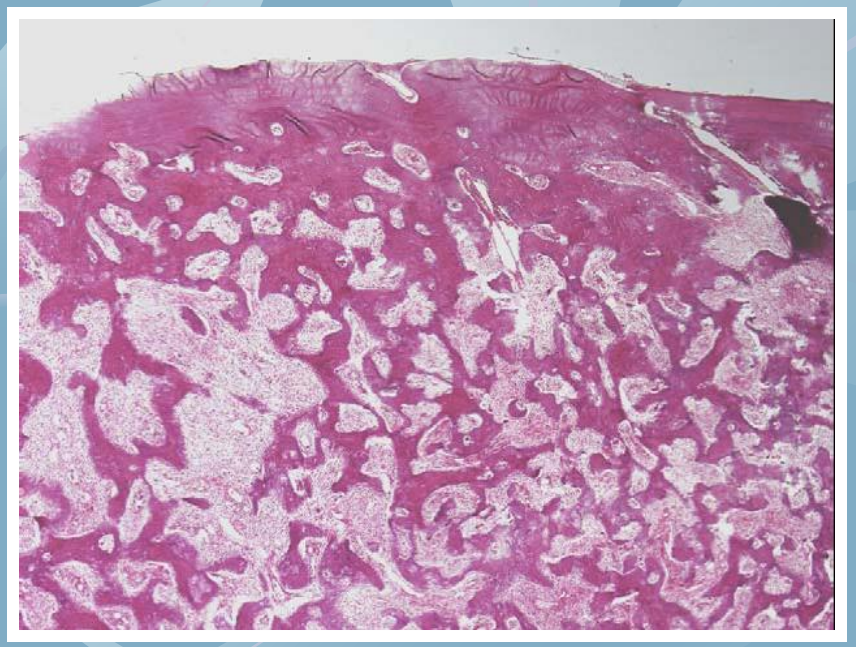
- Radiograph reveals the presence of marked density trabeculae which encroaches to a variable degree upon the orbit and antral cavity.
- The frontal bone is also thickened with homogeneous or variegated type of density.
- The nasal septum is grossly thickened, dense and curved.

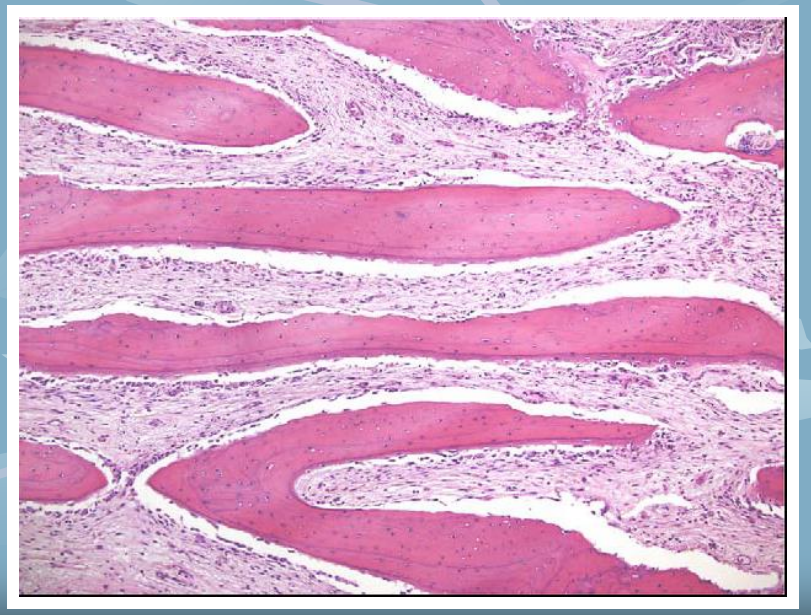
INVESTIGATIONS

LAB INVESTIGATIONS

- Increased alkaline phosphatase level
- Normal serum calcium and phosphate level
- Thyroid function tests
- Conventional radiographs
- Computed Tomography
- Magnetic resonance imaging
- Bone scan

HISTOPATHOLOGY





- Areas of fibrous tissue & newly formed bone like tissue
- Proliferating fibroblasts in a compact stroma of interlacing collagen fibres
- Irregular trabeculae of bone are scattered with no definite pattern of arrangement
- Bony trabeculae vary in shape- slender , “C” shaped & more variable giving a Chinese letter pattern.
- Trabeculae – usually coarse woven bone/ lamellar
- Lack of osteoblastic rimming- important criterion for recognition of FD
- Occasionally may encounter trabeculae rimmed by osteoblasts

MANAGEMENT

- Surgical intervention or shave-down of small lesions.
- Autologous graft/acrylic splint.
- Cosmetic deformity with associated psychological problems/functional deformity may dictate surgical intervention in the younger patients.

Medical management :

- Bisphosphonates anolog of pyrophosphates

- Recurrence - 25% to 50%
- Malignant transformation – 0.5% especially when treated with radiotherapy.


THANK
YOU
Http://maomao520.yeah.net