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Classifications of Fibroosseous lesion with brief review on Fibrous Dysplasia

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ABSTRACT

Fibro-osseous lesions (FOLs) are a group of lesions which are recognized to affect the jaws and the craniofacial bones which are known for their confusing area in diagnostic pathology. The disease involves the normal architecture of bone is replaced by fibrous tissue containing varying amount of foci of mineralization. The article pitches a bright on the various classification systems are given for FOLs and also highlights the role of radiographic & Histopathological features in the diagnosis of FOLs, which will qualify us to adopt a uniform vocabulary and to aid the Diagnostic and surgical pathologist in the diagnosis of this diverse group of maxillofacial lesions.

Keywords— *Fibroosseous Lesions, Bony Lesion, Fibrous Dysplasia, Albright syndrome, Café u late spots.*

1. INTRODUCTION

The fibro-osseous lesions of the jaw are defined as the replacement of normal bone with a variably collagenous, connective tissue matrix containing trabeculae of new bone and in some lesions cementum-like material. It is also defined as lesions in which normal bone is replaced by fibrous connective tissue showing the varying amount of mineralization. The definitive diagnosis of FOLs is not possible only alone by examination of incisional /excisional biopsy material and it mainly relies on close clinical as well as radiological correlation. [1, 2, 3,4, 5]

2. CLASSIFICATIONS

The various classifications of FOL are listed below. [1-10]

2.1 WHO Classification – 1971

Neoplasms and other tumors related to the odontogenic apparatus Cementomas:

- Benign Cementoblastoma (true cementoma)
- Cementing fibroma
- Periapical cemental dysplasia (periapical fibrous dysplasia)
- Gigantiform Cementoma (familial multiple Cementomas)

Neoplasms and other tumors related to bone

Osteogenic neoplasm:

- Ossifying fibroma (fibro-Osteoma)

Non-neoplastic bone lesions:

- Fibrous dysplasia
- Cherubism
- Central giant cell granuloma
- Aneurysmal bone cyst
- Simple bone cyst

2.2 Edward and Cario classification-1984

- Benign cementoblastoma
- Ossifying fibroma
- cemetifying fibroma
- cemento-ossifying fibroma
- Periapical cemental dysplasia

2.3 Waldron classification – 1985

Fibrous dysplasia:

- Polyostotic
- Monostotic

Fibro-osseous (cemental) lesions presumably arising in the periodontal ligament:

- Periapical cemental dysplasia
- Localised fibro-osseous-cemental lesions
- Florid cement-osseous dysplasia (gigantiform cementoma)
- Ossifying and cementifying fibroma

Fibro-osseous neoplasms of uncertain or debatable relationship to those arising in the periodontal ligament:

- Cementoblastoma
- osteoblastoma
- osteoid osteoma
- Juvenile active ossifying fibroma
- aggressive active ossifying / cementifying fibromas

2.4 Classification of fibro-osseous lesions of the head and neck by Pecaro B.C. (1986)

Fibrous dysplasia:

- Polyostotic
- Monostotic

Fibro-osseous lesions from dental structures:

- Periapical fibrous dysplasia
- Cemento-osseous dysplasia
- Cemento-ossifying fibroma

Fibro-osseous neoplasm:

- Cementoblastoma/Osteoblastoma(osteoid osteoma)
- Aggressive active ossifying fibroma

2.5 WHO Classification – 1992

Osteogenic neoplasms:

- Cemento-ossifying fibroma (cementifying fibroma, ossifying fibroma)

Non-neoplastic bone lesions:

- Fibrous dysplasia of jaw
- Cemento-osseous dysplasia
    i. Periapical cemental dysplasia
    ii. Florid cemento-osseous dysplasia
    iii. Other comments-osseous dysplasia
- Cherubism (familial multilocular cystic disease of the jaw)
- Central giant cell granuloma
- Aneurysmal bone cyst
- Solitary bone cyst

2.6 Yoon JH, et.al classification—1989

Periodontal ligament origin:

- Neoplastic
    i. Cemetifying fibroma
    ii. Benign cementoblastoma
    iii. Gigantiform cementoma
    iv. Cemento-ossifying fibroma
    v. Ossifying fibroma
- Non-neoplastic
    i. Periapical Cemental dysplasia
- Medullary bone origin
  Neoplastic:
    i. Osteoma
    ii. Osteoblastoma
  Non-neoplastic:
    i. Chronic sclerosing osteomyelitis
    ii. Fibrous dysplasia

2.7 Classification of fibro-osseous lesions by Neville B.W. et al (1995)

Fibrous dysplasia

Cemento-osseous dysplasia

- Periapical cemento-osseous dysplasia
- Focal cemento-osseous dysplasia
- Florid cemento-osseous dysplasia
- Cemento-ossifying fibroma

2.8 Eversole classification, 2008

In 2008, Eversole et al. gave a comprehensive classification including developmental lesions, neoplastic lesions, and inflammatory /reactive processes. This classification emphasized that final diagnosis can be attained by correlation of microscopic, imaging and clinical features together but not on the basis of histopathological features alone.

- Bone dysplasias:
  i. Fibrous dysplasia:
     Monostotic
     Polyostotic
     Polyostotic with endocrinopathy (McCuneAlbright)
     Osteofibrous dysplasia
  ii. Osteitis deformans or Pagets disease
  iii. Pagetoid heritable bone dysplasias of childhood
  iv. Segmental odontomaxillary dysplasia
- Cemento-osseous dysplasias:
  i. Focal cemento-osseous dysplasia
  ii. Florid cemento-osseous dysplasia
- Inflammatory/reactive processes:
  i. Focal sclerosing osteomyelitis
  ii. Diffuse sclerosing osteomyelitis
  iii. Proliferative periostitis
- Metabolic Disease: hyperparathyroidism
- Neoplastic lesions (ossifying fibromas):
  i. Ossifying fibroma
  ii. Hyperparathyroidism-jaw lesion syndrome
  iii. Juvenile ossifying fibroma:
     Trabecular type
     Psammomatoid type
  iv. Gigantiform Cementoma.

3. FIBROUS DYSPLASIA

It is a developmental condition that is characterized by replacement of normal bone by an excessive proliferation of cellular fibrous connective tissue intermixed with irregular bony trabeculae." It was first described by Von Recklinghausen in 1891.

Other names: Fibro-osseous dysplasia, Fibrosis of bone (Elmslie, 1931), Chronic hyperplasia (Davis, 1941), Fibrocystic Disease, Osteitis fibrosa localisata, Focal osteitis fibrosa, Fibro-osteodystrophy. [1,4,6,8,9,11]

3.1 Classification

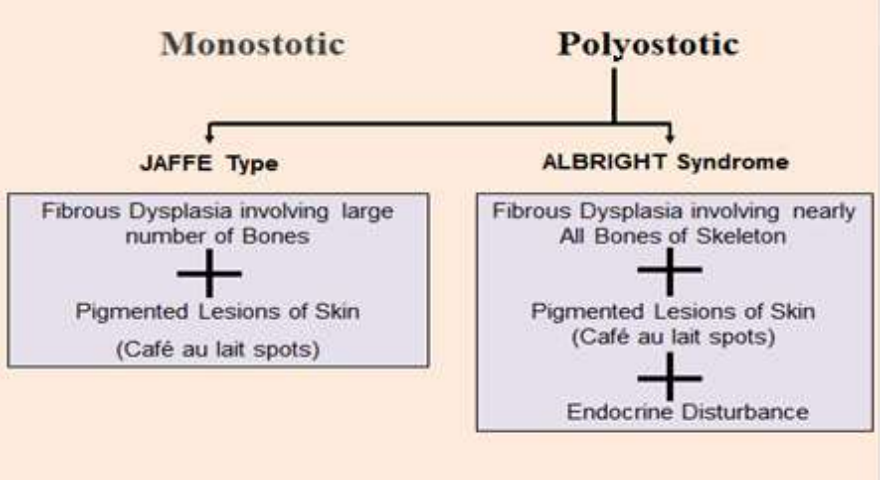


Fig. 1: Classification flow chart

3.2 Types of Fibrous Dysplasia with clinical Presentation

1. **Monostotic fibrous Dysplasia:** it involves Single Bone. 70-80% of all type of fibrous dysplasia are Monostotic form. Both the sex is equally affected. Most commonly seen in 1st and 2nd decade of life. Maxilla most commonly involved than

mandible. Unilateral variety more common than bilateral variety. Most common clinical features are Painless swelling & bulging. Expansion of the buccal cortex rarely lingual cortical plate expansion seen. [9,10,11]

Intra-Orally – Swelling of the affected bone. A protuberant excrescence of inferior Border. Mucosa is intact over the lesion. Teeth mayor may not displace. Delayed / Disturbed eruption of teeth. Displacement of Sinus floor. Obliteration in the Maxillary sinus. The most common intraoral region is premolar-molar region. [7,8,9]

2. **Polyostotic Fibrous dysplasia:** it involves two or more bones. 20-30% of all fibrous dysplasia are Polyostotic form. Unilateral involvement most common than bilateral involvement. Syndromes associated with it are Jaffe-Lichtenstein syndrome, McCune-Albright syndrome. Most commonly seen in long bones compared to craniofacial bones. The most common symptom is Recurrent Aching Bone Pain. The most common complication is Spontaneous Fracture of bone. Most commonly involved bone are Skull, Clavicle, Pelvic bones, Scapula, Long bones. bowing of long bones, Shepherd crook deformity, Lateral bowing of proximal part of the thigh, Widening of hip region.[12,13,14]
   Skin lesions show 1. Café au Lait spots 2. Increased melanin pigmentation in basal cell
   Endocrine problems: Commonly in females. Sexual precocity. Menstrual bleeding, Breast development, Pubic hair at an early age (3–4 year). [4,5]

3. **Craniofacial form: it is** 10-25 % of patients with monostotic fibrous dysplasia. 50% in polyostotic fibrous dysplasia. It can occur as an isolated lesion. Most common sites are Frontal, Sphenoidal, Maxillary, Ethmoidal bones. Most common features are Hypertelorism, Cranial asymmetry, Facial deformity, Visual impairment, Exophthalmos, Blindness, Hearing loss, Tinnitus, Hyposmia, Anosmia. [10-16]

3.3 Radiographic features: [10,11,13,16]

- **Location:**
  i. Maxilla : mandible = 2:1
  ii. Posterior aspect
  iii. Unilateral, rarely bilateral
- **Periphery:**
  i. Ill-defined
  ii. Occasionally can appear sharp and even corticated especially in young lesions
- **Internal structure:** The density and trabecular pattern vary considerably. The variation is more pronounced in the mandible and more homogeneous in the maxilla. The abnormal trabeculae usually are:
  i. Shorter
  ii. Thinner
  iii. Irregularly shaped
  iv. Numerous than normal trabeculae.

  The internal aspect of bone may be:
  i. Radiolucent
  ii. Radiopaque
  iii. Mixed

Radiolucent: Round, oval, irregularly shaped radiolucency with a sclerotic margin. May resemble a cyst or with a wider band of increased density giving it a granular appearance in adults suggestive of Fibrous dysplasia. Sharply defined and punch out lesion without any certification. There is Multilocular radiolucent lesion with septa ranging from few in number, wispy & of poor density resembling central giant cell granuloma to coarse & thick resembling Ameloblastoma.

Radiopaque: Orange peel or fingerprint appearance – younger age group, Stippled (fingerprint pattern) more common in the mandible, Inferior cortex – not thinned with little or no resorption, Inferior cortex with thumbprint – confirm the diagnosis, Ground glass appearance. Dense structure less homogenous density referred to as hyperostosis.

Mixed: Small area with ground glass or Orange peel appearance**.** The proportion of radiolucent & radiopacity varies**-** Worm like radiolucency with coiled up pattern within an area of slightly increase radiolucency.
Loss of lamina dura**,** Resorption of roots. The displace the inferior alveolar nerve canal in a superior direction.

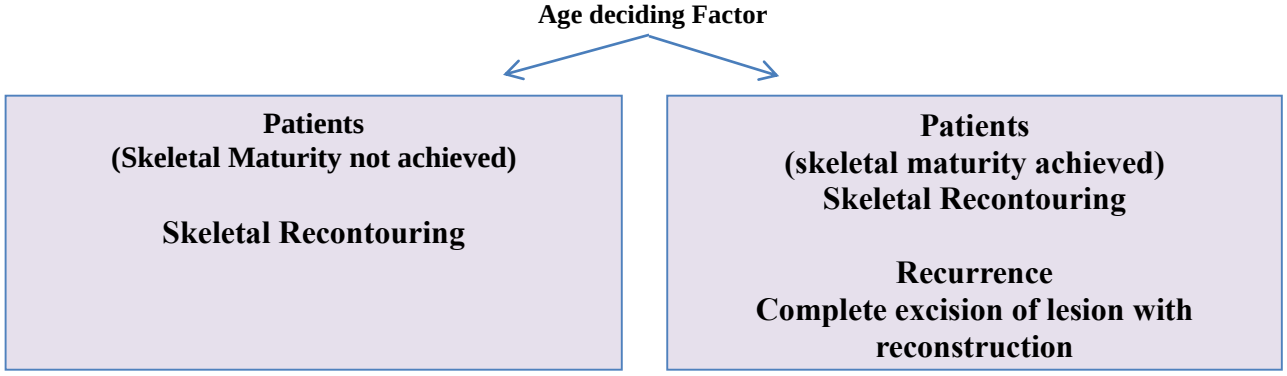
Histopathology: 1. Fibrous stroma – proliferating fibroblasts 2. Interlacing collagen fiber 3. Delicate trabeculae of woven bone enmeshed in the fibrous stroma 4. Trabeculae – Chinese letter pattern [13,-16]

Differential diagnosis: Paget's disease, Chronic osteomyelitis, Chronic ostitis, Periapical cemental dysplasia, Central giant cell granuloma, Traumatic bone cyst, Aneurysmal bone cyst. [10,14,16]

Investigation: In 50% of cases- serum alkaline phosphatase level – elevated**.** Pituitary follicle stimulating hormone – premature secretion. Basal metabolic rate - Moderately elevated. [11,13,16]

Management: Medical Management**:** Bisphosphonates (BPNs)Include Etidronate, Pamidronate, Alendronate [12,13,14]

Surgical Management:
Small lesions are under observation with 3 months follow up.
Large Lesions are treated according to the age of the patient:



4. CONCLUSION

The maxillofacial bones make up a diverse collection of disorders that include neoplastic and non-neoplastic and hereditary and non-hereditary conditions. FOLs of the jaw has been under frequent renaming and reclassification due to their varied features. This review will give clue to diagnose Fibrous dysplasia.

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