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Fibro-Osseous Lesions: A Review

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Abstract:- Fibro-osseous lesions comprise a diverse group of pathological conditions including developmental lesions, reactive or dysplastic diseases and neoplasms. Fibrous dysplasia is a benign pathologic condition of bones,where normal bone is replaced by fibrous connective tissue. Osseous dysplasia is the second most common form of benign fibro-osseous lesion in the jawbones. These lesions only develop in tooth-bearing regions.

Keywords: *Fibrous Dysplasia, Cherubism, Ossifying fibroma, central giant cell granuloma.*

I. INTRODUCTION

Fibro-osseous lesions comprise a diverse group of pathological conditions including developmental lesions, reactive or dysplastic diseases and neoplasms. It is a group of jaw disorders characterized by the replacement of bone by benign connective tissue matrix.[1]

Clinically, fibro-osseous lesions may be associated with significant cosmetic and functional disturbances or they may be completely asymptomatic localized lesions that are identified only on routine radiograph.[13]

Radiographically, they may appear as solitary, multifocal, or multi quadrant lesion; they may have radiolucent, mixed radiolucent-radiopaque, predominantly radiopaque, or ground glass appearance[14]

The definitive diagnosis of a fibro-osseous lesion requires correlation of the histologic appearance of the lesion with the clinical, radiographic, and intraoperative findings.

II. WHO CLASSIFICATION OF FIBRO-OSSEOUS LESIONS (2005)

Diagnosis can be made by the correlation of clinical, radiological as well as by histological features:

- Ossifying fibroma
- Fibrous dysplasia
- Osseous dysplasia
- ✓ Periapical osseous dysplasia
- ✓ Focal osseous dysplasia
- ✓ Florid osseous dysplasia
- ✓ Familial gigantiform cementoma
- Central giant cell granuloma
- Cherubism
- Aneurysmal bone cyst
- Solitary bone cyst

III. FIBROUS DYSPLASIA

Fibrous dysplasia is a benign pathologic condition of bone, where the normal bone is replaced by fibrous tissues connective tissue.[2]

It is an uncommon, nonhereditary, developmental anomaly of the bone due to defective osteoblastic differentiation and maturation.

The exact cause is unknown, but recently the cause has been reported to be somatic mutation in the GNAS1 gene leading to overproduction of cAMP resulting in dysfunction of affected tissues.

- Monostotic fibrous dysplasia
- Polyostotic fibrous dysplasia

Initial manifestations are most commonly noted in persons aged between 3 and 15years. Men and women are affected equally. There isno racial predilection.

Monostotic fibrous dysplasia is the more common (70-80%) and less severe form than polyostotic fibrous dysplasia (20-30%). When only one bone is involved, it is called monostotic fibrous dysplasia. Most cases of monostotic fibrous dysplasia are discovered incidentally on radiographic examination, usually in the second decade of life. The most common site of monostotic fibrous dysplasia is the zygomatico-maxillary complex (ZMC).[11]

The characteristic presentation of monostotic fibrous dysplasia is a painless, slow-growing, unilateral swelling in the affected area.[11]

In monostotic fibrous dysplasia, patient gives typical leonine appearance (leontiasis ossea).

When multiple bones are involved, it is called **Polyostotic fibrous dysplasia.** In the polyostotic type, curvature of the femoral neck and proximal shaft leads to the characteristic **'shepherd crook deformity'**. (Fig 1) [10]

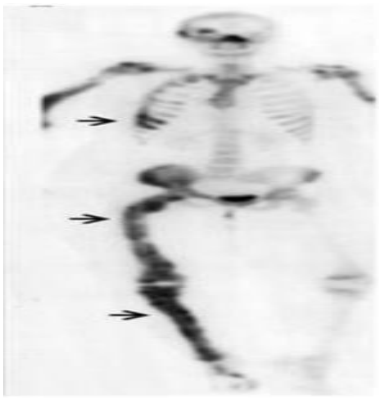

Fig. 1: Technetium bone scintigraphy view demonstrating patchy tracer uptake at affected sites.

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Shepherd crook deformity is the pathognomonic feature for the diagnosis of polyostotic fibrous dysplasia. **McCune-Albright syndrome** (MAS) is characterized by a triad of polyostotic fibrous dysplasia, **café au lait spots** (Fig 2) [10], and multiple endocrinopathies[3].Mazabraud syndrome, a rare, disease that is characterized by polyostotic fibrous dysplasia and intramuscular myxomas.[1]

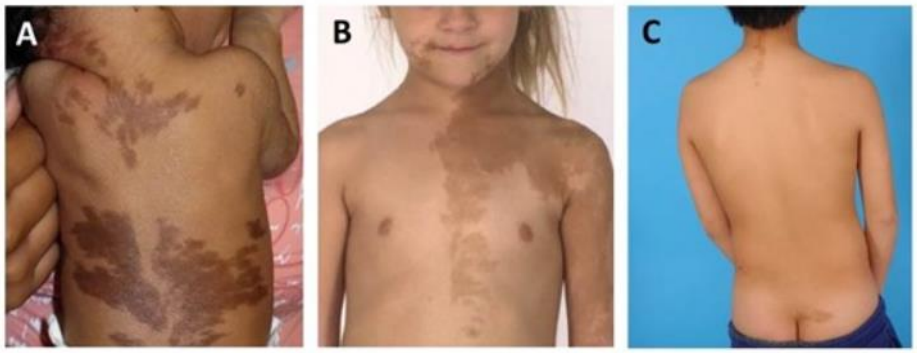

Fig. 2: Café au lait skin pigmentation

The oral manifestations may include expansion of jaws and their deformity and altered teeth eruption due to loss of normal bony support during development. Endocrine disturbances may be due to the latter.[4]

Radiographically they have a ground glass or orange peel appearance due to presence of radio-opaque spicules in the radiolucency.

Histopathologic appearance includes haphazard arrangement of fibroblasts. The trabeculae resemble Chinese script pattern. The treatment includes surgical correction of the skeletal deformities.

IV. OSSEOUS DYSPLASIA

Osseous dysplasia is the second most common form of benign fibro-osseous lesion in the jawbones.[1]These lesions only develop in tooth-bearing regions.[6]

There is predilection for females, which may be due to hormonal imbalance.

According to the 2005 WHO classification, it is classified into 4 types:

- Periapical osseous dysplasia
- Focal osseous dysplasia
- Florid osseous dysplasia
- Familial gigantiform cementoma

A. Periapical osseous dysplasia:

It is a reasonably well-defined entity, predominantly involving the apical region of vital mandibular anterior teeth, either as solitary or multiple small lesions. They rarely exceed 1cm in diameter. Most patients are asymptomatic and the lesions are self-limiting and rarely expand the bone.[6]

Radiographically they consist of multiple round to ovoid, radiolucent lesions at the apex of the vital teeth, mimicking periapical pathology of pulpal origin.

B. Focal osseous dysplasia:

It is an ambiguous lesion described previously by various terminologies. However, the recently used term focal cemento-osseous dysplasia was recommended by Summerlin and Tomich. It is common in middle-aged females, especially in African-Americans, with a mean age of mid-thirties.[6]

Mandibular molar region is the commonly affected site. Majority are asymptomatic and on average 1.5cm in size. The characteristic feature at the periapical region often leads to inaccurate diagnosis of periapical infection and therefore pulp vitality testing could be useful in ruling out pulpal pathology. Radiographically they appear as focal circumscribed apical lesions less than 2cm in diameter.

C. Florid osseous dysplasia:

It is a reactive, non-neoplastic process, most commonly seen in middle-aged African women. When patients are asymptomatic, conventional radiographs showing multiquadrant diffuse radiopaquelesions in the alveolar bonehas a major role in diagnosis. Radiographically they appear as multi quadrant opacities.

D. Familial gigantiform cementoma:

Most often the diagnosis is made on the basis of typical clinical, radiographic and histological features. Swelling of the jaw bones is the primary complaint by most patients. It presents as a slow growing, multifocal swelling involving multiple quadrants of the jaw bones. Radiographs of the jaw bones reveal multiple, irregular, often lobular radio-opacities in the maxilla and mandible, often crossing the midline.[6]

Histological features of osseous dysplasia is similar irrespective of clinical type. It constitutes of a highly cellular connective tissue background with plenty of spindle shaped fibroblasts and collagen fibres. The connective tissue also shows numerous islands of woven or lamellar bone and globular calcification resembling cementum. Inflammatory cells are also present.[8]Most of the cemento-osseous dysplasia are asymptomatic and self-limiting and therefore do not require any treatment or surgical intervention except periodic follow-ups. Any surgical procedures such as

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extraction or diagnostic biopsy, are often contraindicated as they may result in infection and osteomyelitis.

V. OSSIFYING FIBROMA

Ossifying fibromais a benign, rare, fibro-osseous lesion of jawbones characterised by replacement of normal bone with fibrous tissue. The fibrous tissue will show varying amounts of calcified structures that resemble bone and/or cementum.[12]

It is differentiated from Fibrous dysplasia as it is well demarcated. It can occur in any facial bone. Occurs due to mutation in GNAS 1 gene. May occur at any age, but common in young adults with a predilection for the mandible. Females are more commonly affected.

It is usually asymptomatic, slow growing resulting in mild deformity. Teeth may be displaced. Skin and overlying mucosa are intact.

Radiographically it is well delineated from the adjacent unaffected sites. It mainly causes expansion of jaws.

Histologically the osteoid trabeculae show aggressive growth. There are many delicate interlacing collagen fibres interspersed by large number of proliferating fibroblasts.[12]

Complete surgical removal of the lesion is the recommended treatment. Recurrence is rare.

VI. CENTRAL GIANT CELL GRANULOMA

They are an uncommon, histologically benign but locally aggressive and destructive lesion occurring in the jaw bones. The aetiology is unknown.[7]

Most common in people less than 30 years of age with a female predilection. Either jaw may be involved, more common in mandible. Usually asymptomatic and are discovered accidentally. It may sometimes cause expansion of cortex, perforation, mobility, displacement and root resorption of involved teeth.

Based on clinical presentation and radiographic features it is classified into two types: nonaggressive and aggressive. Nonaggressive type is slow growing without causing root resorption and cortical perforation. It often shows new bone formation. The aggressive type has quick growth associated with pain, cortical perforation and root resorption.[7].

Radiographically shows a radiolucent area with a smooth or ragged border with faint trabeculae.

Histologically there are loose fibrillar connective tissue stroma with proliferating fibroblasts and small capillaries. Multinucleated giant cells are seen throughout the connective tissue.

Treatment includes curettage or surgical excision. Radiotherapy is contraindicated.

VII. CHERUBISM

Also known as Familial Fibrous Dysplasia of Jaws/ Disseminated Juvenile Fibrous Dysplasia is a peculiar genetic disease of the jaw bones of children. It was first reported by Jones in 1933. It is a distinct entity from fibrous dysplasia with an autosomal dominant trait with mutations in the gene coding SH3-domain binding protein-2(SH3BP2) located on chromosome 4p16.3. The term Cherubism is because of the facial similarity of affected children to the plump cheekedangel “Cherubs”.[7]

It is a disease of children, manifesting from 14 months to 3 years as painless bilateral symmetrical enlargement of maxilla and mandible. There is a characteristic upward gaze, expansion of both maxilla and mandible and cervical lymphadenopathy. A rim of sclera is visible beneath the iris giving “eye to heaven” appearance. Sometimes it may lead to hearing and visionimpairment, dental anomalies such as missing teeth, crowding, malocclusion, premature loss of deciduous teeth and delayed eruption of permanent teeth.

The disease is self-limiting and stops at puberty and then begins to regress spontaneously.

Radiographically it is characterized by bilateral, symmetrical occurrence of multilocular radiolucencies with expansion of both jaws. Multiple unerupted teeth with destruction of alveolar bone leading to typical “floating tooth syndrome”. (Fig 3 and Fig 4) [9]

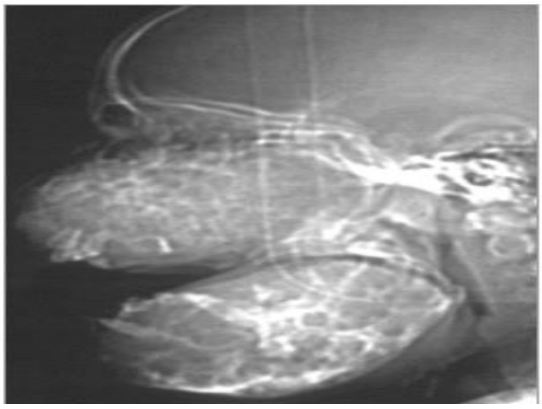

Fig. 3: Cherubism: Lateral CT showing extensive involvement of the jaws

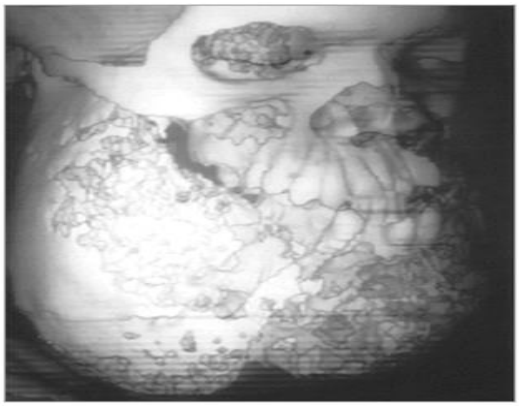

Fig. 4: Cherubism: CT showing enlargement of mandible

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Histologically there are numerous large, spindle fibroblasts and multinucleated giant cells in the highly vascular connective tissue. Capillaries are often lined by large endothelial cells and surrounded by thick, eosinophilic perivascular cuffing, characteristic of Cherubism.

No treatment is indicated except observation. Rarely surgery may be required for cosmetic reasons.

VIII. ANEURYSMAL BONE CYST

It is a pseudocyst which was separated as a distinct entity in 1942 by Jaffe and Lichtenstein. It may arise de novo in bone. Usually affects persons under 20 years of age, lesions were observed in every part of skeleton. They are usually tender or painful, limiting movement of affected bone.[7]

Radiographically bone is expanded, with honeycomb or soap-bubble appearance.

Histologically it consists of a fibrous connective tissue stroma containing many sinusoidal blood-filled spaces, young fibroblasts, multinucleated giant cell.

Treatment consists of surgical curettement or excision. No recurrence has been reported in jaws.

IX. SOLITARY BONE CYST

Also known as traumatic bone cyst, haemorrhagic cyst or extravasation cyst. It is a pseudocyst. The aetiology is unknown. According to traumatic theory, trauma forms a clot and this clot breaks down which leaves an empty cavity within the bone.[7]

Usually occurs in second decade of life with no sex predilection. In most cases where jaw bones are affected, pulps of the involved teeth are vital. Patients present with swelling and rarely pain.

Radiographically there is a smooth radiolucent area of variable size, sometimes with a thin sclerotic border.

Histologically it has a thin connective tissue membrane lining the cavity but no other significant features.

Treatment: Enucleation is done and the cavity is closed. Healing occurs in 6-12 months.

X. CONCLUSION

Fibro-osseous lesions are not only classified based on histologic features but also based on the clinical, Radiological corelations. When a differential diagnosis is not possible on the basis of clinical and radiographic features, molecular analysis are done. Proper final diagnosis will help in appropriate therapeutic action. Unless there is considerable destruction of bone, lesions may be treated conservatively.

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