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Review Article

# Understanding fibrous dysplasia & its variants from cause to care

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## Abstract

Fibro-osseous lesions represent a diverse group of conditions characterized by the replacement of normal bone with fibrous connective tissue and varying levels of mineralization. Fibrous dysplasia (FD) is a benign fibro-osseous condition where normal bone is replaced by fibrous tissue and irregular metaplastic bone. It often results from mutations in the GNAS1 gene, leading to hormonal imbalances and café-au-lait spots. FD commonly affects craniofacial bones, especially the posterior maxilla, and may cause bone weakening and fractures. It is classified as monostotic (single bone) or polyostotic (multiple bones), with variants like Jaffe's type, Albright syndrome, craniofacial form, and cherubism. Elevated alkaline phosphatase levels are common in affected individuals.

**Keywords:** Fibrous dysplasia, Cherubism, Bone disorder, GNAS mutation, Variant G protein, Rare disease, Pathogenesis, Trabeculae.

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## 1. Introduction

The term "fibro osseous lesions" is a collective term for a variety of diseases in which connective tissue takes the place of healthy bone, with variable degrees of mineralization manifesting as woven bone or cementum-like circular acellular basophilic formations.[1] Waldron defined fibro osseous lesions as a group of pathological changes in the jaw bones in which normal bone is replaced by fibrous tissue, with or without calcification, in 1970.[2]

Fibrous dysplasia (F.D) Fibrous dysplasia (FD) is a benign, non-neoplastic and non-encapsulated fibro-osseous situation concerning cranial and extra-cranial bones which display alternative of everyday bone by way of cell fibrous tissue containing islands of metaplastic bone.[3] Lichtenstein in 1938 brought the term Fibrous dysplasia.

It's far labeled by using W.H.O. in 1992 as developmental in starting place. Some declare that FD is a dysplastic or hyperplastic system with diffuse margins that mix into adjoining bone. In keeping with a few authors, FD is typically caused by mutation inside the GNAS1 gene (20q13.2). The GNAS1 (guanine nucleotide binding protein, α-stimulating activity polypeptide) gene encodes a G-protein that stimulates the production of cAMP. The mutation outcomes in a continuous activation of the G-protein main to overproduction of cAMP in affected tissues. This results in hyper feature of affected endocrine organs, often giving upward thrust to precocious puberty, hyperthyroidism, growth hormone and cortisol overproduction. Secondly, there's an extended proliferation of melanocytes resulting in big café-au-lait spots with abnormal margins instead of the everyday outlined café-au-lait spots in neurofibromatosis. Thirdly, cAMP is notion to have an effect on the differentiation of osteoblasts main to FD.[4]

FD maximum regularly happens in the metaphysial and diaphysial regions of the lengthy bones, the shoulder bones,

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the bones of the pelvic girdle and those of the jaws and cranium.

The posterior maxilla is the most common web page, especially within the area around the first molar. The anterior maxilla is hardly ever concerned. Mandibular lesions are generally determined between the mental foramen and perspective of the mandible.

It is a properly described mutation of osteogenic mesenchyme into an isomorphic spindle referred to as stroma containing thin dependent bizarre shaped trabeculae of metaplastic bone.[5]

In FD, the normal spongiosa is changed by using a fibrous and osseous tissue that's ill perfect for expanding and contracting pressure. The cortex will become narrower due to endosteal bone resorption that could result in pathological fracture.

In Laboratory investigations, l/3rd of FD patient's exhibit accelerated alkaline phosphatase ranges in blood chemistry opinions.[6]

FD has been traditionally classified as:

1. Monostotic: involving only one bone,
2. Polyostotic: involving multiple bone.[4]

Polyostotic FD is sub-labeled as:

a) Jaffe's type - Many bones are affected and light brown spots over skin I.e. cafe-au- lait spots.
b) Albright syndrome - numerous bones are involved with cafe-an-lait spots or lesions in the affected vicinity and alongside this endocrine disorder, like precocious puberty in ladies.
c) Craniofacial form - involving the craniofacial complex only.
d) Cherubism -An autosomal dominant version of FD
   - Involvement of maxilla and mandible
   - multiplied stages of serum alkaline phosphatase enzyme
   - Regression might also arise after formative years.[4]

## 2. Monostotic Fibrous Dysplasia

This time period is implemented to the ones styles of the sickness in which handiest one bone is affected. It happens in about 70 - 80 % of the cases of FD. It additionally does now not manifest extra-skeletal lesions along with those seen in polyostotic FD but may additionally end up polyostotic and affect a couple of bones. It is less extreme than polyostotic form. The clinical time period "leontiasis ossea" has been applied to instances of Monostotic FD which influences maxilla or facial bones and deliver the patient a leonine look.[4]

### *2.1. Etiology*

The etiology is unknown. Many viable factors have been counseled however none had been typically typical. In advance, it changed into counseled that it's far because of aberrant hobby in the bone forming mesenchymal tissue.

There is additionally scientific proof which suggests that neighborhood infections or trauma to the bone effects on this bizarre reparative response by bone. However, a few instances have been stated to be congenital that constitute an autosomal recessive sickness.[7]

### *2.2. Clinical features*

It happens with same predilection in ladies and men with a mild predominance for girls. it's far more common in children and young adults. Mean age of incidence being 27-34 years.[4]

The first medical sign of the disorder is a painless swelling or bulging of the jaw. The swelling usually includes buccal and labial plate and infrequently the lingual plate.[8]

While it involves the mandible, it every so often causes a protuberant excrescence of the inferior border. The overlying mucosa is nearly forever intact over the lesion. Tenderness in the end develops. There can be some malalignment, tipping or displacement of the enamel because of the progressive expansile nature of the lesion.[4]

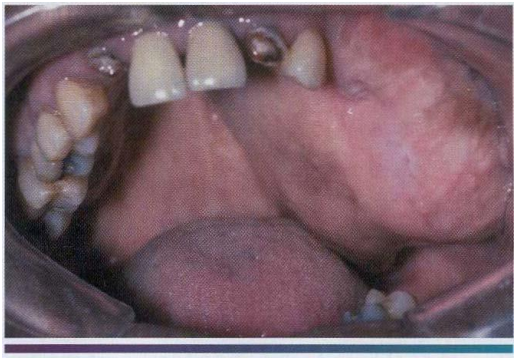

**Figure 1:** Expansile mass of the left maxilla in a 45 -year-old woman.

### *2.3. Radiographic features*

The roentgenographic appearance of F.D of the jaw is extremely variable. There are 3 basic patterns which may be seen.

1. In one type, the lesion is generally a rather small unilocular radiolucency or a somewhat larger multilocular radiolucency, due to marked fibrous proliferation and deposition of osteoid which will subsequently calcify, both with a rather well circumscribed border and containing a network of fine bony trabeculae.

2. In second type, the pattern is similar except that increased trabeculation renders the lesion more opaque and typically mottled in appearance.[4]

Third type is quite opaque, with many delicate trabeculae giving a ground glass or peau d' orange appearance to the lesion. This type is not well circumscribed but blends into the adjacent normal bone.[8]

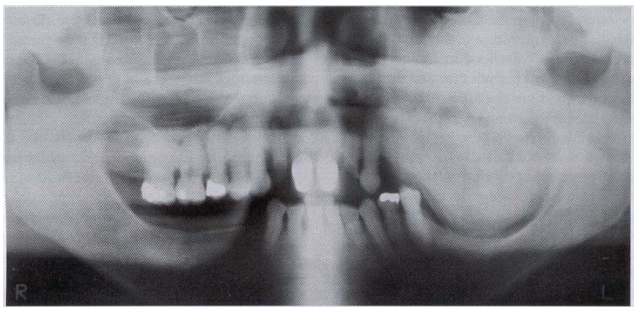

**Figure 2:** Panoramic radiograph of the patient showing diffuse "ground-glass" radiopacity is evident.

In all the three sorts, cortical bone turns into thinned due to the expansile nature of the boom but this bony plate is seldom perforated. These photos are pleasant visible in bone window C.T snap shots. The roots of tooth in the involved vicinity can be separated or moved out of everyday function however simplest once in a while show off extreme resorption. In a few instances, the bone seems so opaque that the roots of tooth can be indistinct or now not seen.[9]

*2.4. Histopathological features*

The lesion is essentially a fibrous one made of proliferating fibroblasts in a compact stroma of interlacing collagen fibers. The trabeculae are thin and placed at ordinary periods. Abnormal trabeculae of bone are scattered via out the lesion, without a particular sample of arrangement. Typically, some of those trabeculae are delicate C-fashioned or as Chinese language man or woman-fashioned hook or horse shoe formed and sometimes may additionally shape earrings.

The trabeculae are usually coarse woven bone however may be lamellar bone. The connection of osteoblasts and osteoclasts to the trabeculae is just like that of polyostotic form. Large lesions may also display variation from location to place and every now and then gift a bony response greater on the outer edge than within the primary element.[4]

Areas of moth eaten spicules devoid of osteoblastic pastime can also be visible in some lesions which represent a burned out phase. The presence of lamellar bone does now not verify a analysis of FD. Other morphological sorts of calcification, such as small rounded bodies (cementum our bodies or globular calcifications) and minute basophilic and laminated calcification may be visible in FD.

The histopathology of FD is not pathognomonic and it must be showed with the aid of correlation of histopathology, radiography and scientific features. Massive cells may also be visible in a few instances.[4]

*2.5. Treatment*

The treatment consists of surgical elimination of the lesion. The general public of lesions go away an extreme facial deformity as they may be too big at the time of diagnosis. Lesions specifically with ground glass or orange peel appearance aren't properly circumscribed and consequently, would should be block resected. Majority of cases are dealt with by using a conservative removal of that portion of the lesion, contributing to facial deformity. A few sufferers with minimal cosmetic or useful deformity won't require surgical treatment.

However, deformity associated with psychological troubles or practical issues may also require surgical discount of the lesion to the desirable contour without attempting entire elimination. It’s far predicted that among 25% and 50% of patients will display a few regrowth after a conservative surgical procedure. Regrowth after surgical discount tactics can also occur suggesting that surgical intervention have to be delayed as long as viable.

Radiation remedy is contraindicated, because it incorporates the hazard of put up irradiation bone sarcoma. Some examples of spontaneous sarcomatous modifications have also been stated.[9]

## 3. Polyostotic Fibrous Dysplasia

Polyostotic fibrous dysplasia (McCUNE albright syndrome) Weil in 1922, diagnosed the case of polyostotic FD related to pores and skin lesions and endocrine disturbance. The circumstance has been specially described by Albright, from wherein the obvious syndrome derives its eponym. ‘Polyostotic’ were carried out to those lesions in which more than one bone is affected.

There are seemingly seperate forms of Polyostotic FD (PFD) which are

1. Jaffe's kind
2. Albright's syndrome[4]

Other endocrine disturbances like hyperthyroidism, Cushing’s syndrome, hyperparathyroidism, acromegaly, goiter, and gynacomastia might also arise. In addition to those, the occasional incidence of a couple of intramuscular soft tissue. Myomas as greater-skeletal manifestations of PFD has also been cited.[10]

*3.1. Etiology*

It is thought to be similar to that of Monostotic fibrous dysplasia. The difference is that it influences multiple bones like jaws, femur, tibia, pelvis, ribs, skull, clavicle and facial bones.

*3.2. Clinical features*

PFD takes place in approximately 20-30 % cases of FD.

The sickness commonly manifests early in existence with a glaring deformity of long bones, frequently unilateral in distribution. It has insidious onset. Recurrent bone pain is the maximum not unusual providing skeletal signs and symptoms.[11] Because of the intense bone changes, spontaneous fractures are a commonplace hardship of the sickness. The structural integrity of the bone is weakened and the burden bearing regions grow to be bowed.

The curvature of the femoral neck and proximal shaft of the femur markedly boom causing a 'shepherd crook deformity', which is a function signal of the disorder. Overgrowth of adjoining tender tissues can be present.

Two apparently separate kinds of Polyostotic FD are described as

1. Jaffe's type - FD involving a variable wide variety of bones, observed by means of pigmented lesions of the skin or "cafe-au-lait" spots of thin mild brown colour. It's far slight and non- revolutionary shape. This type occurs in about 50% of the cases.
2. Albright's syndrome - FD even more intense, regarding almost ail bones within the skeleton followed wih the aid of pigmented lesions of the skin in addition to endocrine disturbances of varying kinds. Female sufferers exhibit precocious puberty, every so often beginning on the age of two or three years. Vaginal bleeding is a commonplace manifestation.[4]

Cutaneous pigmentation in PFD is ipsilateral to the aspect of bony lesions, a characteristic that differentiates pigmentation of this disease from that during neurofibromatosis. The pigmented macules or cafe-au-lait spots are associated with multiplied quantities of melanin inside the basal cells of the epidermis.[10]

They're organized in a linear or segmental sample near the midline of the frame, normally overlying the lower lumber backbone, sacrum, buttocks, neck and shoulders. Comparable lesions may arise on lips and oral mucosa additionally.[12]

Some severely affected patients may additionally gift with associated hepatic, cardiac and gastrointestinal disorder (i.e. improved hepatic transaminases, gastrointestinal polyposis and cardiomyopathy.[9]

Mazabraud's syndrome is a rare disorder induced because of association of FD and intramuscular myxoma that occur in the same anatomical location. Patients with gentle tissue myxomas need to be very well examined for FD as extra hazard of sarcomatous transformation in FD with Mazabraud's syndrome has been pronounced. Malignant transformation can also include OsteosarcoMcCune Albright Syndrome (most not unusual), ChondrosarcoMcCune Albright Syndrome, FibrosarcoMcCune Albright Syndrome and LiposarcoMcCune Albright Syndrome. Those malignancies arise most normally within the placing of healing irradiation exposure. Girls may additionally have an extra hazard for breast cancer, probable due to their extended exposure to improved estrogen ranges. The underlying GS alpha gene mutation may also play a function in this.[4]

*3.3. Oral manifestation*

The oral manifestations of PFD are associated with intense disturbance of bony tissue. One 0.33 of the polyostotic sufferers have lesions in the mandible. There might also be enlargement and deformity of the jaws and the eruption pattern of the tooth is disturbed because of lack of support of the developing tooth. The endocrine disturbance additionally, can also regulate the time of eruption of the teeth.[4]

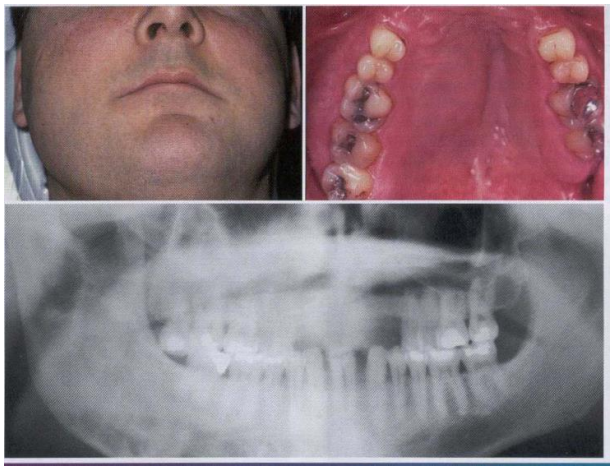

**Figure 3:** Young man exhibiting enlargement of the right maxilla and mandible; B intraoral photograph showing unilateral maxillary expansion; C. panoramic radiograph showing ill-define d lesionsof the right side of both jaws

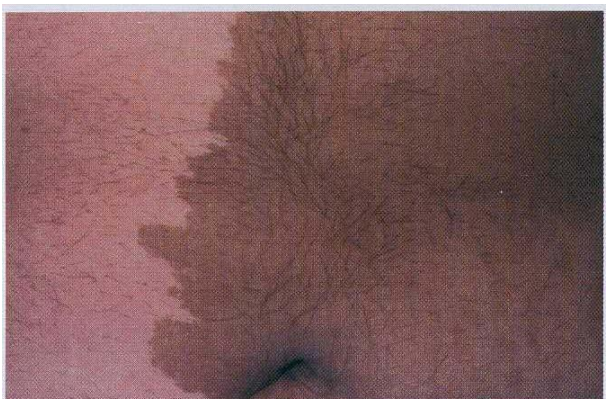

**Figure 4:** Cafe au lait pigmentation of the abdomen.

*3.4. Radiographic features*

The lesions include a radiolucent lesion in the diaphysis or metaphysis with endosteal scalloping. It is able to be present without or with cortical expansion. The radiolucent lesion has a thick sclerotic border and is known as ‘rind sign’.[4]

In fashionable, the medullary part of bone is in the main involved and present abnormal trabeculations that give multilocular cystic appearance. A number of the skull and facial bones, the frontal bone is maximum often worried after which the sphenoid bone, with obliteration of frontal and sphenoid sinuses. Most commonly, maxillary and mandibular involvement has a blended, radiolucent and radiopaque pattern with displacement of teeth and distortion of nasal cavities.[13]

### *3.5. Histological features*

Histological capabilities are much like that of Monostotic FD. The lesion consists of fibrillar connective tissue within which numerous trabeculae, woven immature bone, irregular in form, can be visible.[9]

The osteocytes are quite large and collagen fibers of these trabeculae can frequently be found extending into fibrous tissue. Bone formation by stellate osteoblasts may be seen but rows of cuboidal osteoblasts last on the surface of the trabeculae (osteoblastic rimming) are absent. Those trabeculae usually have wide osteoid seams. Osteoclastic hobby will also be visible in a few areas.[4]

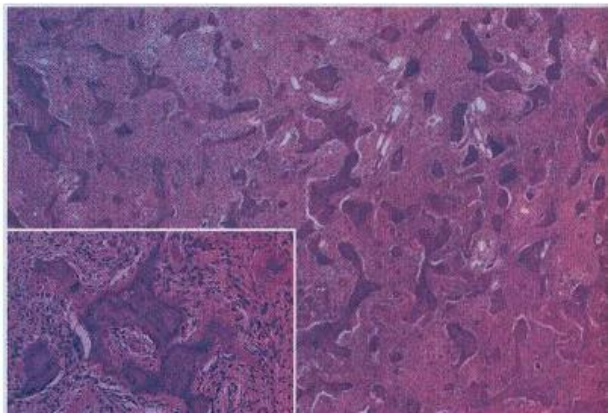

**Figure 5:** Irregularly shaped trabeculae of woven bone in a fibrous stroma.

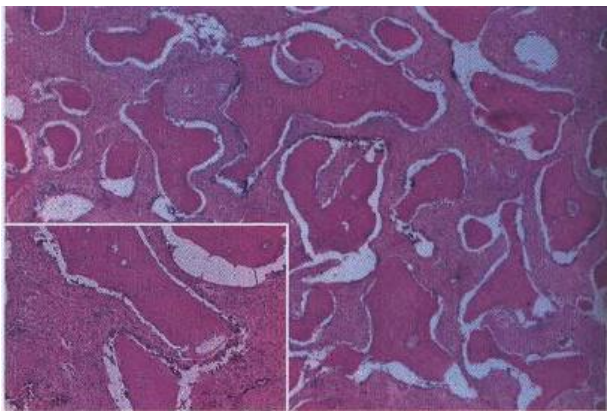

**Figure 6:** Long-standing lesion showing scattered trabeculae of bone within fibrous connective tissue.

### *3.6. Differential diagnosis*

The number one differential consideration for FD of the jaws is Ossifying fibroma (OF). The well circumscribed OF as compared with the diffuse FD frequently serves because the differentiating aspect. Other differentiating capabilities are

| **Fibrous dysplasia** | **Ossifying fibroma** |
|---|---|
| 1st and 2nd decade | 3rd and 4th decade |
| Maxilla > mandible | Mandible > maxilla |
| Diffuse opacity | Circumscribed |
| Self-limited | Continuous growth |
| One or more bones | One bone |
| Woven bone trabeculae | Bony islands and trabeculae |
| Hormone related | Not hormone related |
| Recontouring for cosmetics | Excision |

Chronic Osteomyelitis may every now and then mimic the radiographic appearance of FD. Inflammation, regularly mild, is found in osteomyelitis and can be accompanied with the aid of signs that consist of tenderness, pain and drainage which might be generally not present in FD. The slowly revolutionary and asymptomatic nature of FD typically allows differentiation from malignant tumors of bone.[10]

### *3.7. Laboratory findings*

No constant and vast modifications in serum calcium or phosphorous may be seen, even though, serum alkaline phosphatase degree is occasionally improved. Premature secretion of pituitary follicle stimulating hormone is discovered. There can be an expanded basal metabolic price.[13]

### *3.8. Treatment*

Slight cases may be treated surgically. Severe forms are impossible to deal with like this considering that they tend to be progressive. Because of this, the X-ray radiation turned into used with a few fulfillment however it is hazardous due to the possibility of improvement of brought about osteosarcomas.[4]

### *3.9. Prognosis*

It depends on degree of involvement of skeleton. Uncomplicated lesions are compatible with life. The polyostotic form undergoes spontaneous malignant transformation greater frequently than the monostotic form.[13]

## 4. Craniofacial Fibrous Dysplasia

FD of maxilla is an in particular severe form of the ailment because it has a marked predilection for occurrence in children and is nearly not possible to eliminate without radical, mutilating surgical operation. Those lesions aren't properly circumscribed normally extend regionally to involve the maxillary sinus, the zygomatic technique and the floor of

the orbit and can even expand lower back closer to the base of the skull.[13]

Extreme malocclusion and bulging of the dog fossa or severe prominence of the zygomatic technique generating a marked facial deformity are standard sequelae of this ailment within the maxilla.[9]

*4.1. Clinical features*

This pattern of FD occurs in 10-25% of patients with the Monostotic form and in 50% with the polyostotic form. It additionally happens in an isolated craniofacial form. In an remoted variety, no more cranial lesions are present.

It happens at some stage in 1st and 2nd many years. The common sites of involvement are frontal, sphenoid, maxillary and ethmoid bones.

The fundamental characteristics of FD within the cranial bones are headache, specifically when orbits are affected, sporadic bouts of partial or general lack of awareness and impaired sight and hearing. Hypertelorism, cranial asymmetry, facial deformity, visual impairment, and exophthalmos may additionally occur because of involvement of orbital and periorbital bones.[4]

While ethmoid bone or the horizontal desk of the frontal bone is affected, the lesion normally occurs on one facet most effective and reasons a narrowing and displacement of the orbital hollow space. This process can spread to nasal and paranasal cavities and result in breathing impediments.[9]

*4.2. Radiological features*

There may be function roentgenographic thickening of the base of the skull. The most not unusual appearance is an ‘orange peel pattern’ which includes areas of alternating granular density and radiolucency. This radiographic pattern corresponds to the ground-glass or 'cotton wool' appearance described less commonly as 'whorled plaque like', 'diffuse sclerotic’, 'cyst like', ‘pagetoid' and 'chalky' patterns in worried bone. Tooth displacement and lack of lamina dura is cited in patients with lesions involving the enamel.[9]

*4.3. Treatment*

The treatment includes surgical elimination of the lesion. Most people of lesions leave a severe facial deformity as they are too big at the time of analysis. Lesions specifically with floor glass or orange peel appearance aren't nicely circumscribed and subsequently would should be block resected. Majority of cases are dealt with by a conservative elimination of that part of the lesion contributing to facial deformity.[9]

*4.4. Cherubism*

(Familial fibrous dysplasia of the jaws, Disseminated juvenile fibrous dysplasia, Familial multilocular cystic sickness of the jaws, Familial fibrous swelling of the jaws)

Cherubism is an uncommon, benign fibroosseous lesion which reasons a revolutionary, painless, symmetrical growth of the jaws. it is in general determined in the mandible.[8] It is a non-neoplastic hereditary bone lesion that is histologically much like relevant large mobile granuloma, influences the jaws of youngsters bilaterally and symmetrically, typically producing the so-known as cherubic look. The disorder turned into first defined in 1933 via Jones, who known as it ‘familial multilocular disease of the jaws’.[4]

According to the WHO classification 1992, Cherubism belongs to a collection of non- neoplastic bone lesions affecting handiest the jaws. It is an extraordinary, benign circumstance with autosomal dominant inheritance, and it is one of the only a few genetically decided osteoclastic lesions in the human body. It appears to have 100 percentage penetrance in males and handiest 50-70 percentage penetrance in women with 2:1 male predominance.[14] There is top notch version in the medical expression. although the situation is known to be hereditary, in a few instances there was no detectable own family history, and even though it typically takes place bilaterally there have also been cases of unilateral involvement, perhaps because of incomplete penetrance or new mutations.[4]

Some investigators consider that cherubism arises from the mutation of a nonsex-connected gene chargeable for the improvement of the jaw bones. Usually, the jaw lesions of Cherubism remit spontaneously when affected youngsters attain puberty, however the purpose for this remission is unknown.[4]

The discount in osteoclast formation resulting from intercourse steroids and the growth in plasma concentrations of estradiol and testosterone at puberty each advocate that the genetic disorder responsible for the localized increase in osteoclasts in Cherubism is overridden and normalized through the multiplied synthesis of sex steroids.[4] The gene associated with Cherubism turned into positioned on chromosome 4p l6.3, which encodes the SH3 binding protein, SH3 BP2.[14]

*4.5. Clinical features*

Affected children are normal at beginning and are with out clinically or radiographically obvious sickness till 14 months to 3 years of age. At that point, symmetric growth of the jaw begins. Typically, the earlier the lesion appears, the more swiftly it progresses. The self- restrained bone boom typically starts offevolved to sluggish down whilst the patient reaches five years of age, and stop with the aid of the age of 12-15 years. At puberty, the lesions begin to regress. Jaw remodeling continues via the 0.33 decade of life, on the stop of which the scientific abnormality can be diffused. The signs and symptoms and symptoms depend on the severity of the circumstance and variety from clinically or radiologically undetectable capabilities i.e. mandibular and maxillary overgrowth with respiratory obstruction to impairment of

vision and hearing, trouble in speech and swallowing.[14] Arnott (1978) advised the subsequent grading device for the lesions of cherubism:

Grade I - is characterized by involvement of both mandibular ascending rami
Grade II - by way of involvement of each maxillary tuberosities as well as the mandibular ascending rami
Grade III -through McCune-Albright syndrome involvement of the complete maxilla and mandible except the coronoid manner and condyles.[4]

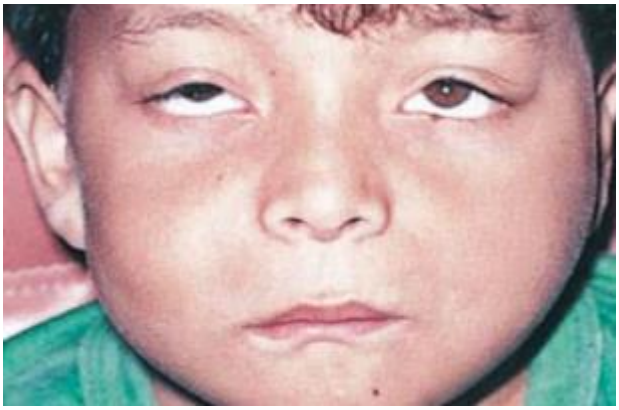

**Figure 7:** Charachterstic facies of a young boy with cherubism. Extensive maxillary involvement has stretched the skin to expose the sclera resulting in the eyes upturned to heaven appearance every other grading machine has been proposed with the aid of Marck PA (1992) this is

Grade 1 – involvement of each mandibular ascending rami.
Grade 2 - involvement of both mandibular ascending rami and both maxillary tuberosities.
Grade 3 – massive involvement of the entire maxilla and mandible except the condylar tactics.
Grade 4 – equal as Grade three with involvement of the orbits inflicting orbital compression.

The jaw lesions are typically painless and symmetric and have florid maxillary contain¬ment. The lesions, which are firm to palpation and nontender, bilaterally symmetrical, maximum generally involve the molar to coronoid areas, the condyles always being spared, and are often associated with cervical lymphadenopathy.[16]

Enlargement of the cervical lymph nodes contributes to the affected person's full-faced look and is stated to be as a result of lymphoid hyperplasia with fibrosis. The lymph nodes turn out to be enlarged earlier than the patient reaches six years of age, lower in length after the age of eight years and are rarely enlarged after the age of 12 years. Intraoral swelling of the alveolar ridges might also arise. When the maxillary ridge is concerned, the palate assumes a V shape. A rim of sclera can be seen below the iris, giving the conventional 'eye to heaven' look.[17]

In Laboratory investigations for Cherubism, serum calcium and phosphorous levels are within ordinary limits however alkaline phosphatase stages may be improved.[14]

*4.6. Oral manifestations*

Severa dental abnormalities, such as agenesis of the second one and 1/3 molars of the mandible, displacement of the teeth, premature exfoliation of the primary tooth, behind schedule eruption of the everlasting enamel, and transpositions and rotation of the tooth were mentioned. In intense cases, teeth resorption may additionally arise. The deciduous dentition can be shed prematurely, starting as early as three years of age. The permanent dentition is frequently defective, with absence of numerous enamel and displacement and lack of eruption of these present. The oral mucosa is usually intact and of regular color.[4]Although Cherubism turned into first of all described as a familial disease affecting the jaws, instances with none apparent hereditary foundation were suggested. In a few instances, Cherubism has been defined as being connected with other illnesses and conditions which includes Noonan's syndrome, a lesion inside the humerus, gingival fibromatosis, psychomotor retardation, orbital involvement and obstructive sleep apnea. The association between those two uncommon inherited diseases indicates that these are independent sicknesses that may be transmitted by means of genes intently related on the several chromosomes.

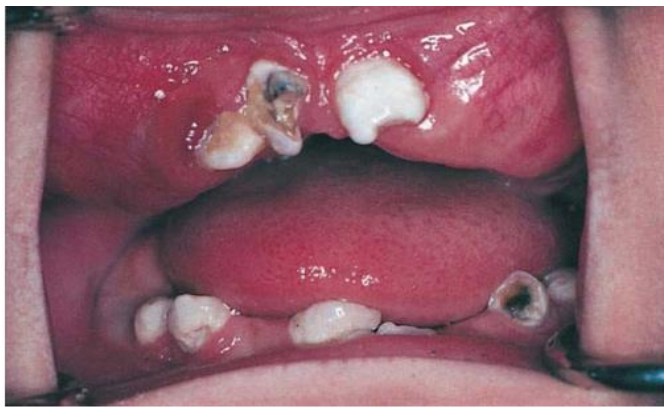

**Figure 8:** Intaoral view of patient showing enlargment of right maxilla and multiple missing teeth

*4.7. Radiographic features*

Radiographically, Cherubism is characterised by means of bilateral multilocular, radiolucent, cystic enlargement of the jaws. Early lesions occur inside the posterior frame of the mandible and the ascending rami. Maxillary lesions may also occur at the same time but break out early radiographic detection due to overlap of the sinus and nasal cavities.[8] Displacement of the inferior alveolar canal has been stated.

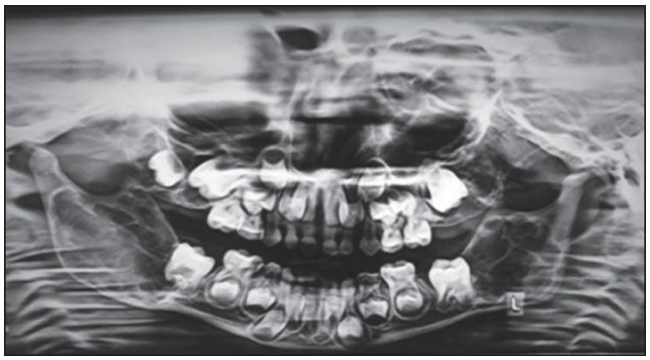

**Figure 9:** OPG showing variably expansile, multiloculated osteolytic lesions involving angle and bilateral rami of the mandible with sparing of condyles

The presence of numerous unerupted teeth and the destruction of the alveolar bone may displace the teeth, producing a radiographic appearance referred to as 'floating tooth syndrome'. With adulthood, the cystic areas in the jaws become reossified, which results in irregular patchy sclerosis. There is a classic (but nonspecific) ground glass appearance because of the small, tightly compressed trabecular pattern.[4]

*4.8. Histological features*

Histologic examination of the lesions normally exhibits severa multinucleated large cells. Those multinucleated cells display strong positivity for tartarate resistant acid phosphatase, which is feature of osteoclasts. The collagenous stroma, which contains a big wide variety of spindle-fashioned fibroblasts, is taken into consideration particular because of its water- logged, granular nature.[4] Numerous small vessels are gift, and the capillaries exhibit big endothelial cells and perivascular cuffing. The eosinophilic cuffing seems to be unique to Cherubism.[14] But, these deposits aren't found in many cases, and their absence does no longer exclude the prognosis of Cherubism. Older, resolving lesions of Cherubism show a growth in fibrous tissue, a lower within the range of large cells and formation of latest bone. The microscopic findings seldom allow a specific diagnosis of Cherubism inside the absence of medical and radiological data.

The differential prognosis of Cherubism consists of massive cellular granuloma of the jaws, Osteoclastoma, Aneurysmal bone cyst, Fibrous dysplasia and Hyperparathyroidism.[4] The stroma in cherubism is greater loosely arranged than that seen in giant cell granuloma. Serum calcium stage is raised in Hyperparathyroidism but does now not alternate in Cherubism. In Osteoclastoma, there may be upward thrust in acid phosphatase enzyme but it does not modify in Cherubism.[4]

*4.9. Treatment*

As Laskin (1985) stated, "the treatment of Cherubism ought to be based on the acknowledged herbal path of the ailment and the scientific behavior of the person case." consequently, surgery to correct the jaw deformities of Cherubism is rarely indicated. If necessary, surgical operation is normally undertaken after puberty, whilst the remission segment of the lesions has been reached, except esthetic concerns or excessive functional issues justify earlier remedy.

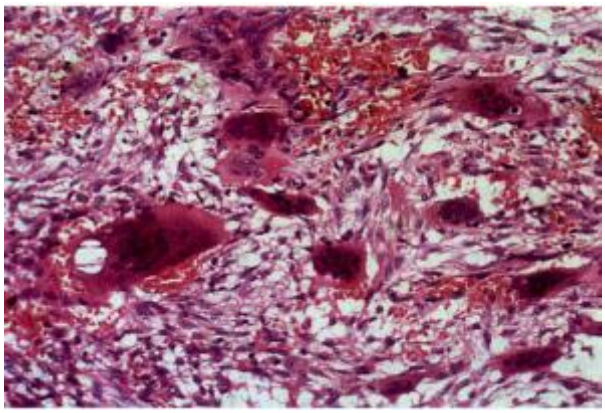

**Figure 10:** An early lesion showing multinucleate giant cells lying in haemorrhagic oedematous fibrous tissue. The appearances are indistinguishable histologically from giant cell granuloma.

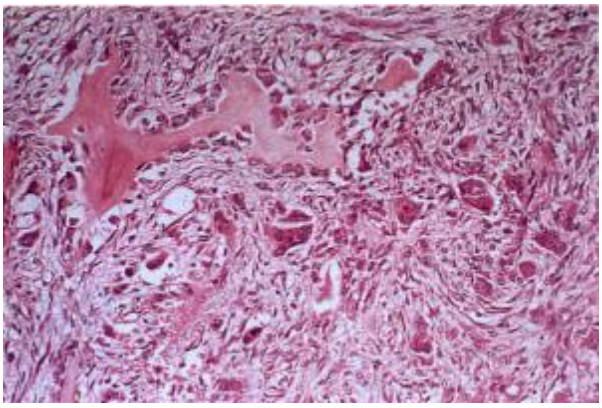

**Figure 11:** In a late lesion there is formation of woven bone by the fibrous tissue and giant cells are less numerous. Eventually bone remodelling will restore the contour and quality of the bone.

## 5. Conclusion

Fibrous dysplasia (FD) represents a spectrum of benign fibro-osseous lesions where normal bone is replaced by fibrous tissue and irregular bone. It may be monostotic (single bone) or polyostotic (multiple bones), with syndromic variants such as McCune-Albright syndrome and Cherubism. These lesions often affect craniofacial bones and can result in deformity, dental complications, and endocrine abnormalities. Diagnosis relies on clinical, radiological, and histopathological correlation. While treatment is usually conservative, surgical intervention may be required for cosmetic or functional reasons. Prognosis varies based on extent, location, and presence of systemic involvement.

## 6. Patient Consent

Patient consent has been taken prior to publications.

## 7. Source of Funding

None.

## 8. Conflict of Interest

None.

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