Monostotic Fibrous Dysplasia of Mandible Associated with an Impacted Tooth – A Case Report and Review of Literature

Nishanth Gollamudi*

Dentist at Nishanths Dental Care, Telangana, India

*Corresponding Author: Nishanth Gollamudi, Dentist at Nishanths Dental Care, Telangana, India.

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Abstract

Fibrous Dysplasia (FD) is considered as a self-limiting developmental benign intramedullary bone lesion in which the normal bone architecture is modified. Progressive replacement of normal bone by excessive proliferating fibrous connective tissue is a characteristic feature of FD. Clinical presentation may vary from patient to another. It may occur at any age, usually detected in second and third years of life with no specific gender predilection. Common sites of involvement in the decreasing order are long bones, ribs, craniofacial bones, and the pelvis. The diagnosis is based on the history, clinical, typical radiological and histopathological features. This article reports a case of Monostotic FD in a 23 year old male patient involving the body of the mandible with emphasis and update on etio-pathogenesis, clinical, radiological and histopathological features.

Keywords: Fibrous Osseous Lesion; Finger Prints Appearance; Ground Glass; Peau d’orange

Case report

A 23 year old male patient reported to the outpatient department of Oral medicine, Diagnosis and Radiology with the chief complaint of swelling in the lower jaw on the left side. On history taking it was revealed patient noticed asymptomatic swelling at his childhood, slowly growing since last 2 months to attain the present size causing facial asymmetry. Patient had no discomfort or pain. His past dental and medical histories were insignificant. Extra oral examination revealed gross facial asymmetry due to the swelling on the lower third of the face on the left side, which on palpation was with ill defined margins, bony hard in consistency and without any signs of paresthesia or inflammation (Figure 1). Lymph node examination revealed a single solitary palpable, movable and tender submandibular lymph node measuring approximately 1 cm in diameter.

Figure 1: Extra oral examination revealed swelling on the lower third of the face on the left side.

Intra oral examination revealed a single solitary dome shaped swelling in the lower left buccal vestibule of normal mucosal color extending anteriorly from the mesial aspect of 33 to the mesial aspect of 37 posteriorly obliterating the buccal vestibule. The mucosa over the swelling was smooth, without any ulcerations or discharge. Other findings observed were Angles Class I malocclusion with Anterior

open bite, a retained deciduous tooth -75 and missing tooth – 35. Swelling on palpation was bony hard in consistency, well defined margins, non-tender with expansion of buccal and lingual cortical plates (Figure 2,3).

**Figure 2:** Intra oral examination showing a single solitary dome shaped swelling in the lower left buccal vestibule of normal mucosal color obliterating the buccal vestibule.

**Figure 3:** Angles Class I malocclusion with midline deviation and Anterior open bite.
Based on the chief complaint, past history, extra and intra oral findings of missing permanent tooth, buccal cortical plate expansion, a provisional diagnosis of a Dentigerous Cyst was given. Other lesions associated with impacted tooth like Adenomatoid Odontogenic tumor, Unicystic variant of Ameloblastoma, Calcifying Epithelial Odontogenic Tumor, Ameloblastic Fibroma and Fibro osseous lesions were considered under differential diagnosis.

Radiologic investigations such as Intra Oral Periapical Radiograph (IOPA), Mandibular occlusal, Panaromic radiograph and Computerized Tomography of the head was advised.

IOPA of the lower left posterior teeth showed retained deciduous tooth 75, permanent teeth – 36, 37 with irregular, short trabecular pattern with ill defined margins extending from the periapical regions of these teeth, showing Ground Glass or fingerprint pattern with loss of periodontal ligament space and lamina dura of the involved teeth (Figure 4). Mandibular occlusal radiograph showed expansion of the buccal and the lingual cortical plates with impacted tooth present on the lingual aspect in the body (Figure 5).

**Figure 4:** IOPA showing retained deciduous tooth 75, permanent teeth – 36, 37 with irregular, short trabecular pattern with ill defined margins showing Ground Glass or fingerprint pattern with loss of periodontal ligament space and lamina dura of the involved teeth.
Panoramic radiograph revealed an ill-defined radiopaque lesion extending anteriorly from roots of the left lower canine to root apexes of left second permanent molar posteriorly, superiorly from the alveolar crest to the inferior border of the mandible with a gradual blending of normal trabecular bone into an abnormal trabecular pattern. The abnormal trabeculae found are shorter, thinner, irregularly shaped and more numerous than the normal trabeculae imparting ground glass appearance or shattered windshield appearance or Peau d’orange appearance or salt-and-pepper or thumb print appearance. The involved teeth on the left side of the lower jaw were displaced superiorly and this expansile lesion could be the possible reason for the deviation, midline shift and presence of anterior open bite in relation to 32, 31, 41, 42, 43 and 44 was observed. Other findings include slight distal displacement of teeth – 33 and 34, a retained deciduous tooth – 75 and obliquely placed impacted tooth in relation to 35 is seen at the inferior border of the mandible and thinning of the lower border of the mandible is noted (Figure 6). To obtain the extents of this fibro-osseous lesion in three dimensions computed tomography scan was advised which showed a well-defined radiopaque expansile lesion on buccal and lingual aspects with ground glass attenuation and narrow zone of transition noted in the body of the left hemi mandible suggestive of a fibro osseous lesion possibly FD (Figure 7). Other radiological differential diagnosis included Ossifying Fibroma, Cementifying or Cemento ossifying Fibroma etc. Patient was informed about the surgical recontouring; informed consent was obtained and was advised hematologic investigations prior to the surgery. Complete blood picture, differential Leukocyte count, Clotting and Bleeding time, serum electrolytes were normal with raised levels of Alkaline Phosphatase in the range of 110 U/L.
**Figure 6:** Panoramic radiograph showing an ill defined radiopaque lesion with gradual blending of normal trabecular bone into an abnormal trabecular pattern imparting a ground glass appearance. Obliquely placed impacted tooth in relation to 35 is seen at the inferior border of the mandible and thinning of the lower border of the mandible.

**Figure 7:** Well defined radiopaque expansile lesion on buccal and lingual aspects with ground glass attenuation and narrow zone of transition noted in the body of the left hemi mandible.
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**Treatment**

After explaining the surgical procedure and obtaining an informed consent from the patient, he was admitted after Pre anesthetic surgical profile. Intra oral approach was preferred, incision is made in the lower left buccal vestibule, flap is raised, the bone is exposed and surgical recontouring of the involved bone is done till symmetry is achieved. Excised specimen was sent for histopathological examination. The given H and E stained soft tissue section under microscope showed fibrillar stroma within which plenty of trabeculae of coarse, woven immature bone, irregular in shape that are evenly spaced. Bone trabeculae contained osteocytes within lacunae without osteoblastic rimming seams of osteoid are evident. Osteoclastic activity is also evident extending to the surface of the trabeculae confirming the diagnosis of Fibrous Dysplasia (Figure 8). Post-operative patient swelling subsided after a week with uneventful healing. A six month postsurgical follow-up of the patient showed no evidence of recurrence.

![Figure 8: H and E section showed fibrillar stroma within which plenty of trabeculae of coarse, woven immature bone, irregular in shape that are evenly spaced. Bone trabeculae contained osteocytes within lacunae without osteoblastic rimming seams of osteoid are evident.](image)

**Discussion**

FD is a benign developmental non-inherited intra medullary bone disease which is characterized by progressive replacement of the normal bony trabeculae, bone marrow and cancellous bone to excessive fibrous connective tissue leading to formation of woven or immature bone mediated by the abnormal osteoblasts due to change in their maturation and differentiation [1,2]. FD of bone was first described by Von Recklinghausen in 1891. Lichtenstein and Jaffe introduced the term "Fibrous Dysplasia" in 1938 [3].

**Etiopathogenesis**

FD develops as a result of disturbance and derangement in the function of the osteoblasts. Mutations occur in the alpha sub unit of the G signaling coupling protein encoded by gene GNAS (Guanine Nucleotide binding protein Alpha Stimulating) which is located on chromosome 20q13.2-13.3. Mutations occur in the form of replacement of the Cysteine or Histidine amino acids by Arginine at position 201 which leads to the loss of activity of the enzyme Guanosine triphosphatase, activation of Adenylate cyclase and increased synthesis of 3’-5’Cyclic Adenosine monophosphate (Figure 9). This mutation is responsible for the increased cellular activity, bony metaplasia and increased and disorganized fibrotic content which manifests as hap hazardly arranged bony trabeculae on radiographs [4-6].

Based on the number and extent of the bones involved, FD can be classified into two types namely, Monostotic and Polyostotic. Monostotic FD goes by the name involving usually a single bone, unilateral and most common among all the types contributing to 70% of the total FD cases reported [7,8]. It is 4 times more common than the Polyostotic variant [5]. It is most commonly seen in the second decade of life with no specific sex predilection and becomes dormant thereafter. It usually involves femur, tibia and skull and facial bones [7,8].

Polyostotic form of FD is seen younger individuals with definite female predilection [9]. Polyostotic type involves multiple bones (such as femur, skull and pelvic bones) and shows definite female predilection. Wide spread and diffuse involvement of the multiple bones leading to compression of the nerves and occupying physiologic spaces in the craniofacial form and may manifest as pain, paresthesia, malocclusion, diplopia, loss of vision, epistaxis, auditory disturbances and diffuse pain mimicking the sinusitis [10-12].
In few cases, the Polyostotic variant is seen in association with patients with variations in female sex hormones particularly during pregnancy [11]. Polyostotic FD is seen associated with a condition called McCune – Albright syndrome with other features such as café au lait pigmentation, multiple endocrinical disturbances such as Precocious puberty, hyperthyroidism, acromegaly, hyperprolactinemia, Cushing syndrome, hyperparathyroidism, gynaecomastia and hypophosphatemic rickets are seen with definite female predilection. It was first described by Donovan James McCune and Fuller Albright and was named after them.

Café au lait pigmentation (called as coffee in milk in French) is one of the presenting sign though not characteristic is brownish colour macules on the skin especially on the trunk with irregular edges and is referred as Coast of Maine. Café au lait pigmentation is also seen in other conditions such as Neurofibromatosis type I in which the spots are smaller rounded with smooth edges referred to as Coast of California. Cutaneous pigmentation in Polyostotic FD is ipsilateral to the side of the bony lesions, a feature that differentiates this from the pigmentation seen in Neurofibromatosis type I. Mazabraud’s syndrome is another condition associated with Polyostotic FD and benign intra muscular myxomas [10,13].

**Clinical presentation**

FD is rare but severe bone deforming disease which causes asymptomatic painless swellings of the bones causing asymmetry, deformity of long bones and bone pain in advanced cases [14]. With initial development of FD the patient usually reports facial swellings and asymmetries. Although the lesion is usually asymptomatic, encroachment on canals and foramina, as well as restrictions of movement, may engender complaints of pain and discomfort.

FD is usually asymptomatic manifests as painless swellings of the face causing non-tender visible asymmetry. When the swellings approach the anatomical neurovascular bundles and impinge on them, causing considerable pain and discomfort [3,10]. FD is usually seen in children and young adults in their growing age periods of either sex i.e., first and second decades of life with mean average onset of age is 10 years. Generally as a thumb of rule, the growth of this lesion usually stops when the skeletal growth ceases but few cases continue to increase in size even after the cessation of the skeletal growth [3].

It affects maxilla more commonly than in mandible in the ratio of 2:1 and posterior areas are more commonly involved than the anterior areas. Swelling usually occurs as dome shaped smooth swelling of the mucosa with obliteration of the vestibule. When the swelling is traumatized surface ulceration may be noticed. Pain and discomfort are seen in minority of the cases. Teeth in the region of the swelling are vital and permanent teeth fail to erupt as in the present case. The involved teeth may also show minimal displacement and the development of malocclusion as in our case which showed midline deviation and anterior open bite. Maxilla is more commonly affected than its counterpart and when it involves the maxillary sinus it gets collapsed and obliterated. Visual problems may be encountered when the orbital cavity is involved. Diplopia or epiphora or exophthalmos or hearing impairment may be seen due to pressure from the lesion on the surrounding structures [10].

**Radiographic features**

Radiographic appearance of FD on the films is manifestation of its histologic structure. Based on the qualitative and quantitative data the radiographic appearance may vary. If there is increased osseous content, the lesion appears radio opaque and increased fibrous tissue appears as radiolucencies on films. Plain film radiography is preliminary radiological investigation which shows mottled and irregular appearance of the involved bone.

Cancellous bones present as increased radio opacity when compared to the cortical bones where the trabeculae are heavily altered. Cortical bones also becomes thinned, expanded but without any perforation maintaining the continuity and without any periosteal reaction. In younger individuals the lesion appears homogenous with ground glass appearance and in elderly patients it appears more sclerotic due to increased bone content [3].

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In FD the bony architecture is disturbed, disrupted disorganized and is replaced by fibrous tissue which causes abnormal distribution of trabeculae. The trabeculae are short, thinner, numerous, small, irregular, not oriented to the occlusal stress and gradually blending with adjacent normal healthy bone. FD appears as an irregular mass with ill defined borders without any distinct borders or any surrounding capsule. This feature differentiates FD from Ossifying Fibroma which has a well defined capsule surrounding it.

FD has been described radiographically as ground glass (granular) or shattered wind shield or peau d’orange or stippling or thumb print (swirling bony trabeculae) or salt and pepper which describes the abnormal and unorganized arrangement of the trabeculae [10,15]. Based on the distribution of the osseous and fibrous component three radiographic patterns are described such as pagetoid, sclerotic and radiolucent. Pagetoid appearance is more common followed by sclerotic type. Radiolucent variant is least common among all radiographic appearances. The present case had pagetoid appearance involving the mandible [9,16,17]. Upward or superior displacement of the inferior canal may be considered to be a pathognomonic. Loss of lamina dura is again considered a characteristic feature when the mass involves the tooth bearing region. However in our case the position of the canal was unaltered but lack of lamina dura on periapical radiographs was seen [18].

Rationale of Computed Tomography and Magnetic Resonance Imaging

Computerized tomography (CT) forms a valuable diagnostic radiographic aid in localizing and confirming the extensions and relationships to the vital anatomical structures. Recent advances in the field of cranio facial radiology have made possible to locate and delineate the boundaries exactly and reconstruct the mass in three dimensions (axial, coronal and sagittal) which plays an important role in pre-operative assessment and possible post treatment outcome.

A regular craniofacial non-contrast CT scan with customized sections of 3-4 mm is preferred to know the margins of mass of the facial bones and skull [19]. Heterogeneous pattern with enlarged and intact cortical plates without any periosteal reaction is a characteristic feature of FD. Depending on the distribution of the fibrous and osseous tissues the relative radio density of the lesion varied in the range from 34 - 513 hounsfield units [20].

CT is the modality of choice when the region of interest is seen involving hard tissues however Magnetic Resonance Imaging (MRI) is particularly helpful when fibrotic and collagen content dominates the osseous tissue which appears as homogenous moderately low signal hypo dense lesions on T1 weighted images and appears hyper dense with high signal intensity on T2 weighted images [20].

Diagnosis and differential diagnosis

Diagnosis is based on asymptomatic painless swelling of the bones, occurrence in second decade of life, growth of which ceases when the skeletal maturity is reached, typical fusiform like growth, expansion of the cortical plates but with no perforation or any periosteal reaction. Typical radiographic features such as loss of lamina dura, abnormal trabeculae appearing as ground glass or thumb print appearance do contribute in arriving at diagnosis. Superior displacement of the canal is also an important feature.

Biochemical marker such as Alkaline Phosphatase levels may rise as in our case but it is not a specific marker. Levels of Alkaline Phosphatase are lowered when Pamidronate is administered. It may serve in measuring the treatment outcome, prognosis and periodic estimation of levels of Alkaline Phosphatase is done to rule out the recurrence. Sudden rise with Alkaline Phosphatase along with bone pain and other associated symptoms may indicate malignant transformation [21].

Differential diagnosis of FD include Ossifying Fibroma, Central Giant cell Granuloma, Paget’s disease, Osteomyelitis. Ossifying Fibroma is a neoplasm arising from the periodontium seen more commonly involving the mandible, in 3 - 4th decades of life with a well defined capsule surrounding the mass with centrifugal growth pattern. It has radiographic features almost same as FD. Paget’s disease also mimics FD has similar radiographic appearance but it has bilateral or generalized involvement of the bones with male predilection, occurrence commonly in elderly age group and it also shows increased levels of Alkaline Phosphatase. Early cases of FD may be confused with Central

Giant cell Granuloma which has definite female predilection, common in mandible anterior to molar, manifests as painless expansion of bone and appears multilocular radiolucency with well defined borders on radiographs [22].

Early stages of Osteomyelitis may resemble FD but Osteomyelitis is inflammatory in origin, pain, swelling, pus discharge and presence of draining sinus tract are the cardinal signs. As the tooth is treated either endodontically or surgically inflammation subsides. But FD is considered as Developmental or self limiting anomaly [10].

**Histopathology**

The characteristic histopathological feature consists of relatively constant ratio of osseous and fibrous tissue in the lesion. It contains spicules of woven bone with typical osteoblastic rimming and foci of irregularly arranged unorganized bony trabeculae in the fibrous connective tissue stroma which is loose, arranged in whorled pattern [6]. The fibrous connective tissue contains predominantly collagen fibers arranged in storiform fashion. The spindle- shaped fibroblasts show star shaped nucleus and bony trabeculae exhibit Chinese letter pattern or Alphabet soup but not specific to FD. The bone (cortical or cancellous) is replaced is by the immature bone called as woven bone which forms from the collagen, in which the trabeculae do not exhibit any functional orientation. The irregular shaped trabeculae are not lined by osteoblasts but focal osteoblastic rimming may be seen. However few cystic areas are also seen due to degeneration of the connective tissue. Osteoclastic activity is also evident extending to the surface of the trabeculae [6].

**Treatment**

As the etio pathogenesis is obscure and the disease is self limiting and sometimes continuous producing facial asymmetry and deformation, there are no standard guidelines for the treatment. However treatment options include the following, Observation and monitoring, Medical and Surgical.

**Observation and monitoring**

As FD is seen in second decade of life and known to stop it by the end of skeletal maturity it can be considered as self-limiting disease. However, regular periodic checkup at every 3 - 6 months needs to be done to assess the growth of the mass. Non-expensive tools such as estimation of Alkaline Phosphatase can be performed. CT scans and Scintigraphy also helps in monitoring the disease progression [23].

**Pharmacological**

Corticosteroids act as anti-inflammatory drugs such as Cortisone which helps in alleviating the pain of the bone lesions in advanced cases when the mass impinges on the nerves. Bisphosphonates such as Alendronate or Zoledronic acid or Pamidronate etc are used when the site is inaccessible to surgery. It has been shown to reduce the rate of growth of these lesions and arrest the resorption by inhibiting action of acid Phosphatase and have been tried in cases with limited success. Bisphosphonates are safe with minimal side effects and usually well-tolerated [5,19]. Matarazzo, *et al*. found decrease in the bone pain, decrease in the markers such as Serum Alkaline Phosphatase, urinary hydroxy proline and increased bone density was observed on the DEXA scan when treated with bisphosphonates such as Pamidronate [24]. Calcitonin causes bone calcification and is an adjuvant to surgery.

Earlier along with steroids, bisphosphonates and Calcitonin, External Beam Radiation therapy was tried. Radiation therapy induced malignant transformation in these lesions and is contraindicated now. Usually sarcomas, Osteosarcoma, Fibro sarcoma and Chondrosarcoma develop post to radiation therapy (radiation induced sarcomas) [25].

**Surgery**

The mainstream treatment of FD is surgical intervention. Based on the patients age treatment is planned. If the patient is in his second decade of life, surgery is postponed until he or she reaches skeletal maturity [7,26]. Aims of the surgical approach include restoring the occlusal deformities, stabilize the occlusion, correct the dentofacial abnormalities restoring the aesthetics, prevent pathological fractures.
and also prevent recurrence of post-surgical relapse [7]. Surgical aspect is of two types. One is surgical contouring or shaving which is conservative approach consists of debridement, Osteoplasty and shaving of the lesion till the desired anatomy is obtained and other is radical excision with post-surgical reconstruction.

When there is a diffuse and wide spread involvement of the facial bones, Radical surgical excision is preferred but it has the disadvantages of extensive loss of anatomical structures and even teeth in the vicinity [14]. Surgery usually involves a multidisciplinary approach consisting of an Oral and Maxillofacial Radiologist, Oral and maxillofacial surgeon, Neurologist, Plastic surgeon, Anesthesiologist and a Orthodontist [9]. Recurrence of FD is common during the growth period and rare in adult age groups. The probability of recurrence increases with conservative surgical approach where the total mass cannot be excised. Radiation is known to trigger Radiation induced malignant transformation and hence it is contra indicated [27].

Conclusion

Isolated cases of FD are rare in the oro facial region. It is considered as self-limiting fibro osseous lesion involving the long bones predominantly and rarely the craniofacial bones. It occurs due to mutation in the GNAS gene which causes abnormal proliferation of fibrous connective tissue and replacement of bone with fibrous tissue. Thus new bone formed in this response is termed as woven bone which appears as ground glass or Peau d’Orange on radiographs. The possible relation of FD to the endocrinal disturbances is to be further studied and caution to rule out endocrinal disturbances in a child with FD is to be emphasized. Oral Physician and maxillofacial radiologist plays an important role in early diagnosis and formulates a proper treatment plan based on the observations of necessary radiological investigations.

Bibliography


