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Case Report

Monostotic fibrous dysplasia of the mandible: A rare case report & review of literature

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ABSTRACT

Fibrous dysplasia is a lesion, characterized by the replacement of osseous tissue with fibro-osseous tissue. It mainly occurs in the calf bones and facial bones, with about 30% of cases affecting the cranial bones. Typically seen during adolescence, fibrous dysplasia can persist into adulthood. The maxilla is roughly twice as likely to be afflicted as the mandible, and it generally manifests unilaterally in the vertical rami and angle of the lower jaw. We report a case of 14-year-old adolescent with symptomatic monostotic fibrous dysplasia of the left posterior and anterior lower jaw region, supported by clinical, radiographic, and pathological findings. Radiographic examination revealed a lesion with radio-density and lucent characteristics, like ground glass. Subsequent biopsy and histological analysis confirmed the diagnosis of Fibrous Dysplasia (Aggressive Lesion). Surgical shaving and re-contouring of the jaw were performed with careful attention to protecting the left mental nerve. Regular follow-up for monostotic fibrous dysplasia showed no clear evidence of progression or malignancy during subsequent assessments.

1. Introduction

Fibrous dysplasia is an uncommon bone condition marked by aberrant growth and development of fibrous tissue within the bones.[1] This disorder usually develops in childhood or adolescence disrupting the normal process of bone growth and weakening the afflicted bones.Fibrous dysplasia affects all bones in the human body from head to toe. (For example, calf bones, face bones, etc.).[2] The actual cause of fibrous dysplasia is unspecified; however, it is assumed to be the consequence of a gene mutation that appears during the early fetal development stage. This mutation results in an overgrowth of fibrous tissue in place of a normal bone leaving the afflicted bones fragile and prone to fracture.[1]Fibrous dysplasia is expansive and diffuse has an ill-defined margin and is difficult to identify.[3]. Fibrous dysplasia symptoms might vary based on where the disorder occurs and how severe it is. Common symptoms include bone discomfort, abnormalities, uneven growth, and a higher risk of fractures. If fibrous dysplasia affects the skull and face bones, it can cause difficulties such as bone abnormalities, nerve compression, and hearing or vision problems. To diagnose fibrous dysplasia effectively, comprehensive steps such as medical history review, physical examination, and advanced imaging modalities like X-rays, CT scans, MRI, and occasionally, biopsy are utilized. Treatment for fibrous dysplasia attempts to alleviate symptoms, avoid complications, and improve quality of life. Based on the extent of the lesion and symptoms, treatment options may include pain relief, physical therapy to improve mobility and strength, surgical procedures to stabilise fractures or correct deformities, and, in some cases, medications or surgery to slow the growth of abnormal bone tissue.[4] Fibrous dysplasia is a chronic disorder that requires continuing management with the right therapy and care. Close monitoring by healthcare personnel

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is essential for detecting and managing any issues that may occur.

2. Case Presentation

A 14-year-old young girl came to the Department of Oral Medicine and Radiology with a complaint of swelling on the left side of her jaw for the past 18 months. History of presenting illness revealed that the patient was asymptomatic for about 18 months. Initially the swelling was small in size and since then progressively increased to the present form. The patient had a three-year history of a similar swelling on the right side of his jaw, and a biopsy revealed fibrous dysplasia, which was treated by surgical shaving and recountering of the right side of the mandible. The patient had no significant medical history, and his vital signs were all within normal range.

An extraoral examination (Figure 1) reveals a diffuse oval swelling of size 12x10cm on the left side of the face spanning medially from the midline to the posterior border of the ramus and superiorly from the ala tragus line to the lower border of the mandible with normal overlying skin.

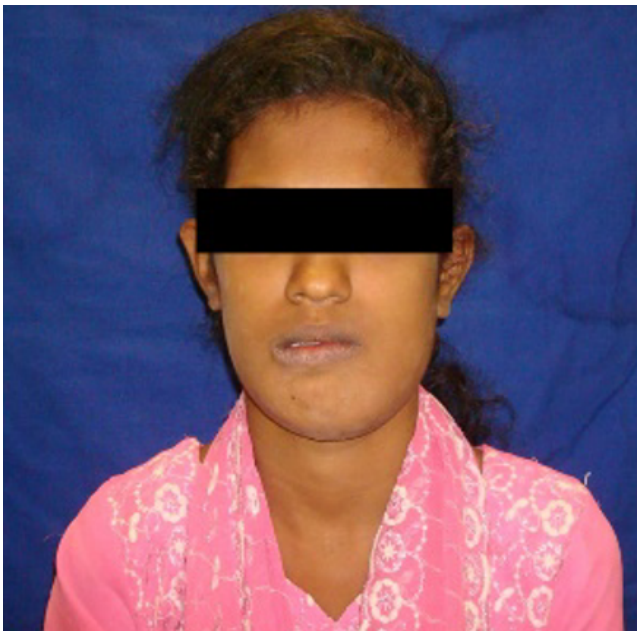

Figure 1: Extraoral image shows prominent swelling and facial asymmetry on the left side

On intra oral examination (Figure 2) shows obliteration of buccal sulcus in relation to left side with positive buccal and lingual expansion. On bimanual palpation the swelling was tender, hard, non-fluctuant, non-compressible with no pus discharge sinus and ulceration.

The patient was advised orthopantomogram (OPG) along with Mandibular occlusal radiograph & Scintigraphy. The orthopantomogram (Figure 3) revealed mixed radio-opaque radiolucency involving the mandible in relation to 46 to 37 with increased expansion in relation to left side when compared to the right.

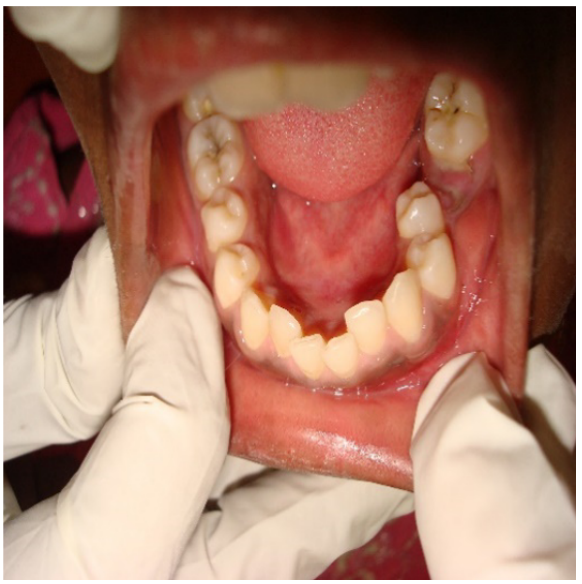

Figure 2: Intraoral photograph showing obliteration of buccal sulcus in relation to left side with positive buccal and lingual expansion

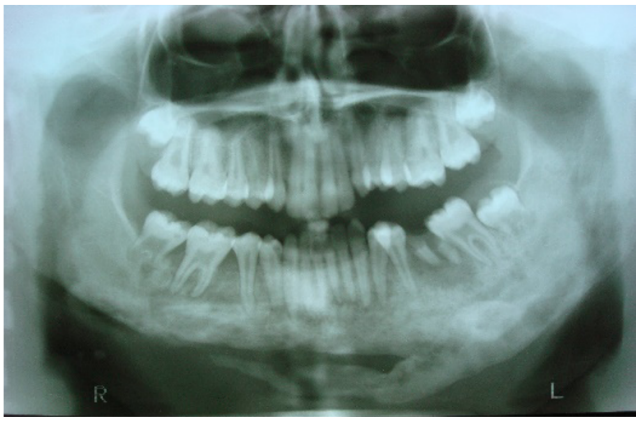

Figure 3: OPG showing mixed radio-opaque radiolucency involving the mandible in relation to 46 to 37 with increased expansion in relation to left side when compared to the right

Mandibular occlusal radiograph (Figure 4) revealed buccal & cortical expansion from 34 to 37 & lingual expansion in relation to the anterior mandibular aspect.

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Scintigraphy (Figure 5) demonstrated an increase in tracer concentration when comparing the left and right sides of the mandible.

The patient was sent to an endocrinologist for further confirmation. Routine tests such as complete blood count, erythrocyte sedimentation rate, serum calcium, and serum

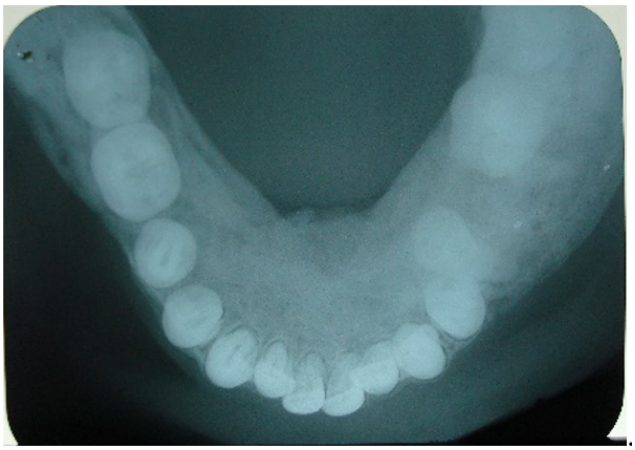

Figure 4: Mandibular occlusal radiograph showing buccal & cortical expansion from 34 to 37 & lingual expansion in relation to the anterior mandibular aspect

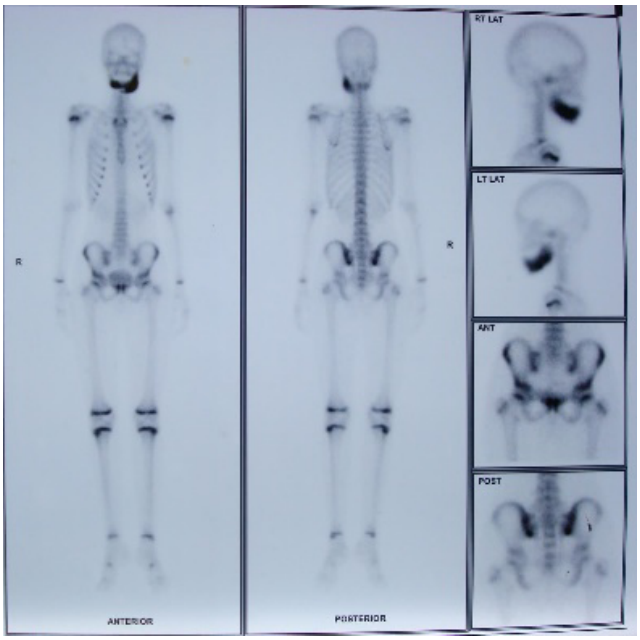

Figure 5: Scintigraphy demonstrated an increase in tracer concentration when comparing the left and right sides of the mandible

alkaline phosphatase were done. All parameters except serum alkaline phosphatase were within normal ranges. Laboratory investigations revealed increased alkaline phosphatase level which was approximately 231iu/l and decreased Vit D (250 H) which was 14.29 ng/dl.

An incisional sample was taken from the lesion on the mandible for histological investigation. Macroscopic examination revealed two fragments of bony hard tissue, largest measuring of size 1x0.4cm of size firm to hard in consistency black in colour another measuring of size 0.2x0.3cm black in colour firm in consistency.

Microscopic examination of the histopathological specimen revealed immature woven bony trabeculae with osteocytes and osteoblastic rimming interspersed with rich fibrous connective tissue stroma. Focal areas show the presence of multinucleated giant cell, inflammatory calls and pleomorphic cell.

Tablets given: T. Sandoeal 500 mg once a day and Calcirol 60,000 IU in 1 glass of milk once a week followed by once a month. And bisphosphonates were planned in 6 weeks. The patient received 5mg of zoledronic acid intravenously. One month after treatment, bone-specific alkaline phosphatase levels dropped and a repeat CT of the head revealed significant improvement. Based on the clinical history, radiographic examination, and histological aspects of the lesion, craniofacial fibrous dysplasia was diagnosed. Surgical recontouring was limited to the mandible. The contour of the alveolar bone was corrected with a stainless-steel drill and soft tissue reduction was also performed.

3. Discussion

Fibrous dysplasia arises when osseous structure is replaced by fibro osseous tissue. Trabeculae or spherules of weakly calcified non-lamellar bone generated by osseous metaplasia can be seen in well-vascularized, cellular fibrous tissue. Its pathophysiology is linked to a GNAS (Guanine nucleotide-binding protein, alpha stimulating) gene mutation from post-zygotic activation at codon 20q 13.2 - q13.3. This encodes a Gs subunit (G-coupled protein receptor). Differentiation and proliferation of bone marrow stromal cells is caused due to disruption in Gsa signalling due to which normal bone is replaced with fibrous tissue. During the early stages of pregnancy gene mutation occurs which is responsible for fibrous dysplasia.

Fibrous dysplasia can be of two types 1. Monostotic 2. Polyostotic

1. Monostotic: affecting one bone
2. Polyostotic: multiple bone affected

Adolescents and young adults are more likely to have a single bone involved. Symptoms of numerous bone deterioration (polyostotic) occur before the age of ten.[5] However, fibrous dysplasia is a genetic disorder caused due to mutation, curing the disease is not found still but it is not passed from parents to child unless both the parent or one carry the gene.

Surgery, pain relief drugs, and bone restoration or stabilization are all options for treatment. This disease has a vast clinical range, from a solitary, asymptomatic monostotic lesion found by chance to a very crippling condition affecting the entire skeleton, loss of eyesight, hearing, and movement. This lesion typically stops developing after puberty but may re-grow during pregnancy.

Symptoms of fibrous dysplasia include:

1. Mild to severe bone discomfort.
2. Swelling.
3. Bone malformation.
4. Bone fractures.
5. Curvature of the leg bones.

Fibrous dysplasia can affect every bone in the body, with the most common being the femur, tibia, ribs, humerus, pelvis, and skull. Rarely, fibrous dysplasia is related to hormone-producing gland syndromes such as Lichtenstein-Jaffe, which involves numerous bones and in rare cases, Café au lait pigmentations on the skin, as well as Albright Syndrome.

Fibrous dysplasia can be diagnosed by X-rays, computed tomography, scintigraphy, or biopsy:

1. X-rays indicate a ground glass look or bubbly lytic lesion Depending on how much calcified material is present, the radiopacity and lucency may vary. The defective bone fuses with normal bone, resulting in loss of circumscription or delineation of the lesion.
2. Computed tomography demonstrates the extent to which the bones are impacted.
3. Scintigraphy employs radioactive tracers are delivered into the circulation; the damaged sections of the bones absorb a greater amount of traces and look brighter in the scan.
4. Biopsy involves removing a tiny portion of the afflicted bone using a hollow needle and sending it to the laboratory for examination.

Fibrous dysplasia can lead to

1. Bone deformities and fractures due to weaker areas.
2. Vision and hearing loss: The damaged bone may be located near the nerves to the eyes and ears, resulting in hearing and vision loss.
3. Arthritis: Deformed pelvis and leg bones might cause osteoarthritis in their joints.
4. Cancer: the damaged bone seldom develops malignant, usually found in people who have had radiotherapy.

Fibrous dysplasia can be treated surgically or non-surgically.

3.1. Non-surgical therapy options

1. Medications: use of bisphosphonates. Bisphosphonates are medications used to reduce the activity of cells that disintegrate bone. These are available as tablets and can also be administered intravenously. Early research has demonstrated significant pain alleviation for this condition.
2. Bracing: Bracing helps to avoid fractures in weaker bones. However, bracing has not been shown to help prevent bone abnormalities.

3.2. Surgical therapy option

1. Curettage: this a surgical procedure commonly used to remove the affected area.
2. Bone graft: after curettage the cavity of the bone is filled with a graft.
3. Internal fixation: metal road and plates are used to fix the fracture.

Patient is also advised physiotherapy treatment such as close kinetic chain exercise and muscle strengthening exercise

Post operative care for fibrous dysplasia include:

1. Counselling
2. Passive range of motion exercise
3. Ice pack to reduce swelling
4. Muscle strengthening exercise
5. Exercise to prevent arthritis

4. Conclusion

"Fibrous Dysplasia" was coined by Lichtenstein in 1938.[6] This lesion is caused by a mutation in guanine nucleotide-binding protein gene that occurs during early development disrupting normal bone formation. Fibrous Dysplasia is a slow-progressing, benign skeletal disorder in which normal bone is replaced with fibro-osseous tissue. Approximately 2.5 percent of all bone tumours and over 7 percent of benign bone tumours are caused by this lesion.[7] Almost half of the patients are diagnosed with polyostotic fibrous dysplasia, and 10-27 percent are diagnosed with monostotic fibrous dysplasia with craniofacial involvement.[8] The incidence of fibrous dysplasia is 1:10,000.[9] It affects all age groups, regardless of gender. Typically, fibrous dysplasia affects the long and flat bones. Fibrous dysplasia commonly affects the posterior part of the jaw and is typically unilateral. Monostotic fibrous dysplasia most usually affects the maxilla rather than the mandible. Fibrous dysplasia limited to a single bone in the maxilla or mandible is uncommon. In our example, the mandible was involved in a 14-year-old female adult, which is unusual in fibrous dysplasia. This lesion was anterior, spanning the midline and spreading to the posterior portion of the jaw. Fibrous dysplasia is often seen in children and young adults.[10,11] According to some research, fibrous dysplasia has a female inclination, while others demonstrate no gender predilection and that fibrous dysplasia progresses into old age. Differential diagnoses for fibrous dysplasia include simple bone cysts, non-ossifying fibroma, osteofibrous dysplasia, adamantinoma, low-grade intramedullary osteosarcoma, and Paget's disease. As a result, doctors struggle to distinguish between benign and malignant bone disorders. Fibrous dysplasia is diagnosed based on a thorough patient history, radiographic evaluation, clinical examination, haematological testing, and histological findings. Each patient exhibits distinct symptoms and clinical findings. As a result, management

should focus on the site of participation. Treatment is generally conservative, such as shaving the afflicted area; this is the most practical and effective choice. In the current case, an incisional biopsy was performed to rule out malignancy and confirm the disorder; bisphosphonates were prescribed; the affected side was operated on while preserving the mental nerve to avoid complications such as paraesthesia and neurovascular bleeding; and regular follow-up was performed to ensure that the disorder did not reoccur.

5. Sources of Funding

None.

6. Conflict of Interest

None.

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