

Case Report

Recurrent Monostotic Fibrous Dysplasia Affecting Mandible: A Case Report

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ABSTRACT:

Fibrous dysplasia (FD) is a skeletal developmental abnormality characterised by the replacement of normal bone with increased fibrous tissue proliferation in irregular bony trabeculae. The maxilla is more commonly affected than the mandible in Monostotic FD. On the affected side, patients may experience swelling, pain, or numbness. We present a recurrent case of monostotic FD affecting the left mandible in a female patient aged 14 who had steadily worsening swelling on the left mandibular side one year following surgical management. This case also emphasises the significance of cone beam computed tomography (CBCT) as an investigative imaging modality since we could conduct the finer evaluation of the lesion and its relationship with adjacent structures with great precision and accuracy.

Key words: fibrous dysplasia; monostotic; ground glass; CBCT; bone recontouring.

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INTRODUCTION

Fibrous Dysplasia (FD) is a benign intraosseous tumourlike disease in which fibrous connective tissue replaces medullary bone, resulting in immature and poorly calcified bone. ¹A genetic mutation in the GNAS Protein causes an imbalance in osteoblastic and osteoclastic activity, which results in FD. ²It can manifest as a monostotic or as a polyostotic form. Monostotic FD, which is usually unilateral, is approximately ten times more common than polyostotic FD. ³⁻⁴ The absence of stable bone matrix can lead to a number of different symptoms including facial asymmetry and distortion. ⁵

In this case report, we report a recurrent case of MFD involving the mandible of a 14-year-old female patient. The diagnosis is established using CBCT imaging characteristics & supported by histopathology. The treatment opted was surgical bone recontouring to correct the patient's facial contour.

CASE REPORT

A 14-year-old female patient presented to our department complaining of swelling over the lower left back region of the jaw since 1 year. Patient noticed the swelling 5 years ago, which was initially small and was progressively increasing in size. There was no associated pain, fever or pus discharge with the swelling. Patient was surgically treated for fibrous dysplasia 4 years ago. One year after the surgery patient noticed the presence of swelling in the same region which gradually increased to the current size. Extraoral examination showed facial asymmetry due to diffuse swelling on the left side of the mandible, measuring approximately 4 x 5 cm. There were no cutaneous pigmentations on the face, mouth opening was adequate, and nonpalpable submandibular lymph nodes were noted. (Figure.1)



Figure 1: Extraoral photograph showing facial asymmetry & diffuse swelling on left side of the jaw.

Intraoral examination displayed bi-cortical expansion of the cortical plates, extending from the mandibular left canine to the retromolar region. Mandibular left second molar was absent and associated teeth showed displacement. (Figure 2) On palpation, it was firm in consistency, non-tender, fixed, non-mobile, non-compressible with no signs of paraesthesia.



Figure 2: Intraoral image showing bi-cortical expansion and obliteration of the vestibule

An intraoral periapical radiograph (IOPA) of the region of interest revealed the loss of normal trabecular structure and a diffuse mixed radiopacity imparting the “ground-glass” appearance. (Figure 3)

For further radiographic assessment of the lesion Cone Beam computed tomography (CBCT) was advised. Concerned axial images showed a unilateral mixed expansile lesion involving left mandible with blending borders. Thinning of the cortical plates with perforation of buccal cortical plate was noted at multiple sites were noted. peripheral part of the lesion showed multiple flecks of calcifications and impacted first and second molar. It also showed displacement of mandibular canal superiorly and lingually. (Figure 4a, b, c)

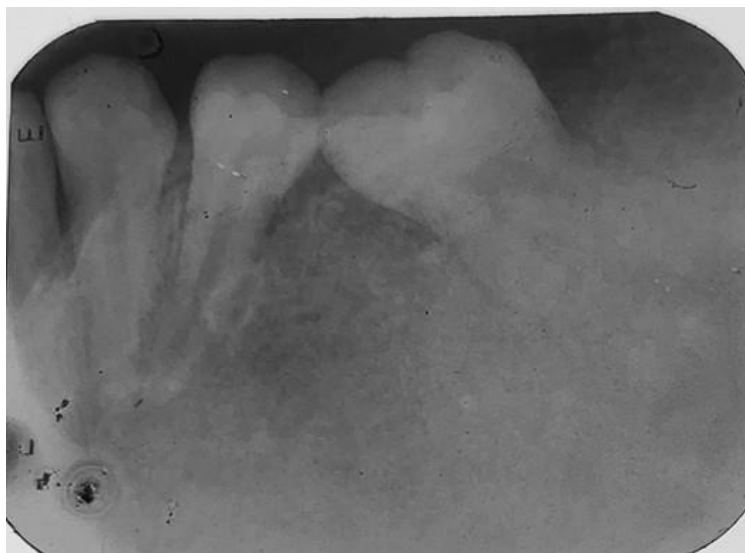


Figure 3: IOPA of affected region showing typical 'Ground glass appearance'

A provisional diagnosis of fibrous dysplasia was made after reviewing the patient's past medical history, clinical, and radiological data. Differential diagnoses of central ossifying fibroma & McCune's Albright's syndrome (MAS) were considered.

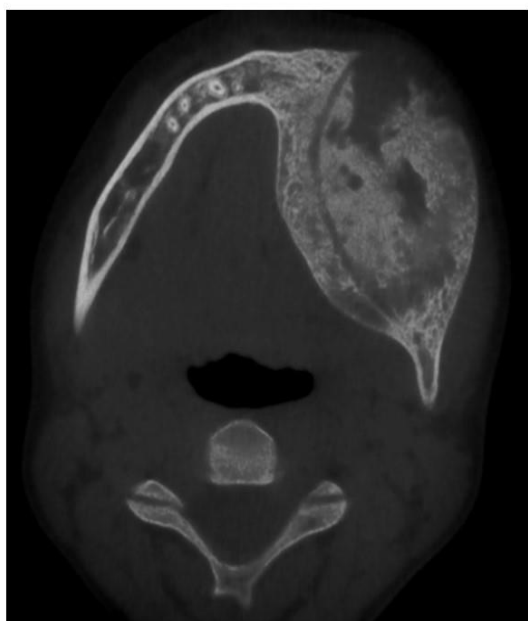


Figure 4a

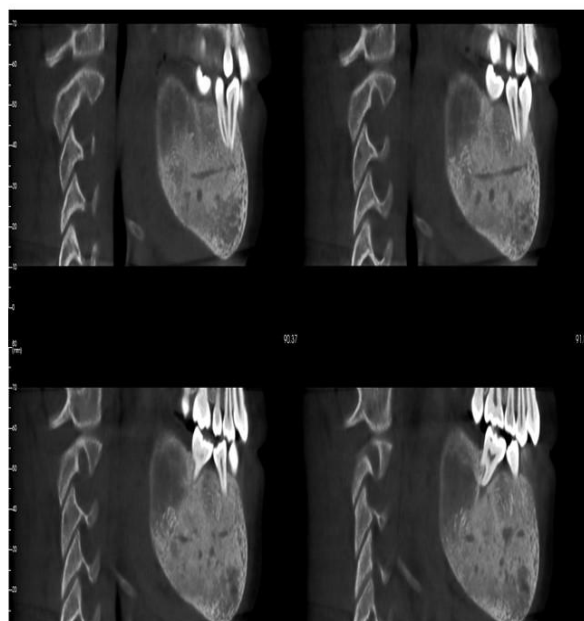
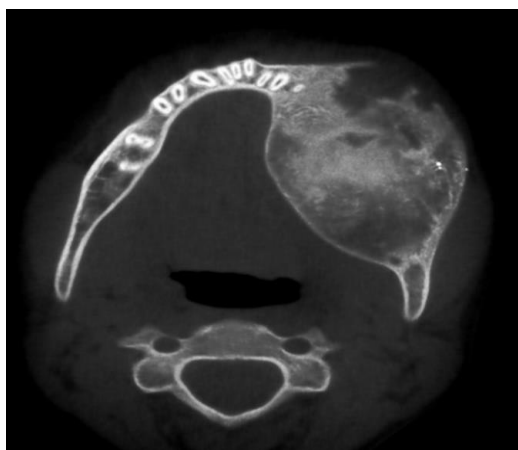


Figure 4b

Figure 4a) & 4b): Axial & sagittal section of the CBCT showing the expansile mixed lesion with left mandible with haphazardly arranged trabecular pattern. Also note the lingual and superior displacement of the mandibular canal.



Blood investigations showed no signs of endocrinopathies which ruled out the possibility of MAS. As the patient was a young female and aesthetics were essential to her, surgical bone recontouring of the affected side of the mandible under general anesthesia was performed. Later the specimen was sent for histopathologic evaluation and the results were consistent with FD. The Haematoxylin and Eosin-stained section showed lesional tissue composed of fibro cellular stroma with plump proliferating fibroblast and numerous trabeculae of course, immature bone with wide osteocytic spaces. The bone deposition showed no orientation to functional pattern. Typical Chinese letter trabecular pattern was noted (Figure 5 a,b)

Patient was followed up for 1 year both clinically and radiologically and the lesion appeared stable with no evidence of regrowth.

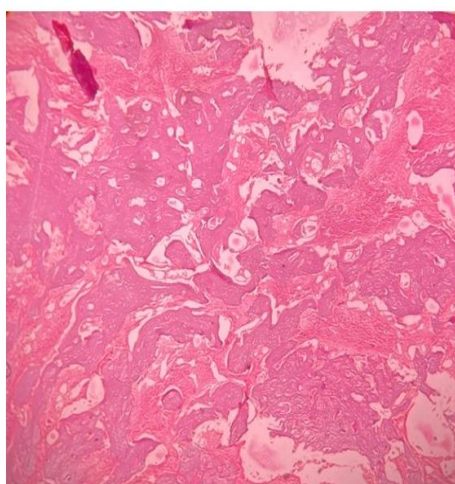


Figure 5a

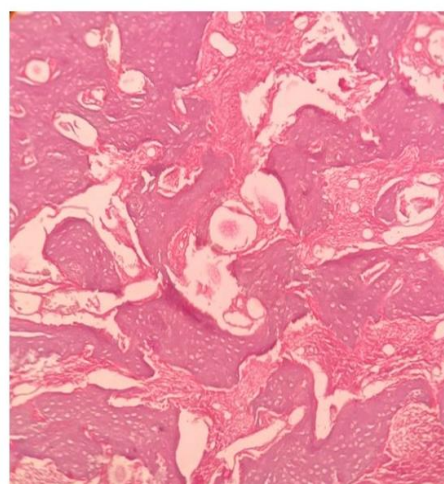


Figure 5b

Figure 5a) Section shows irregular bony trabeculae interspersed within the connective tissue stroma. (H&E, x4).

Figure 5b) Section showing bony trabeculae consisting numerous lacunae containing osteocytes and surrounding connective tissue stroma is fibro-cellular, showing Chinese letter appearance. (H&E, x10).

DISCUSSION

Lichtenstein coined the term "fibrous dysplasia" in 1938.⁶In general, FD manifests in two forms: monostotic, polyostotic. Polyostotic FD with endocrinopathies is known as McCune-Albright syndrome.⁷Although less serious than polyostotic FD, monostotic FD is of greater concern to dentists due to the relatively high frequency of occurrence in the jaws. In monostotic forms, the maxilla is more

commonly involved than the mandible. It can appear in the premolar and molar region and the anterior part of the mandible shows least reported involvement.⁸ Our case showed involvement of mandible with the lesion showing extension into the anterior part, this is a rare incidence in the FD.

The most common radiographic feature of FD is a "ground glass" appearance with blending borders into the normal bone; This is the type of image that was

discovered during our patient's examination. Ogunsalu et al.⁹ suggested the following radiographic appearance of FD: (1) orange peel/ground glass appearance; (2) antrum opacification; (3) radiolucent mandibular lesion; and (4) varying degrees of opacification (not ground glass and not orange peel). Although computed tomography (CT) can be used to assess the lesion, its utility is limited by the high dose that gets absorbed by the patient during exposure.¹⁰ Because of its advantages, such as high resolution and low radiation dose, CBCT is an important diagnostic tool for maxillofacial imaging. In this patient, CBCT guided us in determining the exact extension of the lesion.

FD shows diffuse blending borders into the surrounding normal bone which cannot be assessed accurately on two-dimensions conventional radiographs. CBCT also played an essential role in determining the association of the lesion with the vital structures. Displacement of the associated teeth without resorption and Classical superior displacement of the mandibular canal was noted in our case.

There are several different treatment options for FD, including conservative surgery, radical surgery, and bisphosphonate therapy.¹¹ In our case, the patient was young, and because the asymmetry was bothering her, a conservative surgical approach was taken, keeping in mind that she was 14 years old. After puberty, disease activity usually ceases. 25% to 50% of cases with surgical recontouring show continued growth of the lesion,¹² which may have been the reason for recurrence in our case. As a result, surgical management should be postponed for as long as possible.

A rare but distinct possibility of malignant transformation exists in FD and varies from 2.5% to 7%.¹³ The follow-up is crucial (in terms of time) in order to spot relapses or potentially malignant changes early on.

CONCLUSION

Fibrous dysplasia should be considered in the differential diagnosis of young patients who have unilateral facial swelling. Radiographic evaluation is required because the presentation and distribution of the lesions can help to define the diagnosis. CBCT is useful in assessing a lesion and determining its exact extent, as well as in pre-surgical evaluation and patient follow-up. Increased knowledge of the imaging characteristics of fibrous dysplasia can aid dentists in the prompt diagnosis and management of patients.

LEGENDS

- 1) Extraoral photograph showing facial asymmetry & diffuse swelling on left side of the jaw.
- 2) Intraoral image showing bi-cortical expansion and obliteration of the vestibule.

3) IOPA of affected region showing typical 'Ground glass appearance'.

4a) & 4b) Axial & sagittal section of the CBCT showing the expansile mixed lesion with left mandible with haphazardly arranged trabecular pattern. Also note the lingual and superior displacement of the mandibular canal.

4c) Axial Section depicting sporadic flecks of calcifications noted in the lesion.

5a) Section shows irregular bony trabeculae interspersed within the connective tissue stroma (H&E, ×4)

5b) Section showing bony trabeculae consisting numerous lacunae containing osteocytes and surrounding connective tissue stroma is fibro-cellular, showing Chinese letter appearance. (H&E, ×10)

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