IJMS
Vol 49, No 12, December 2024

Case Report

Monostotic Fibrous Dysplasia with Rare Histopathologic Features: A Case Report

Abbas Karimi[1], MD, DDS, OMFS; Mahboube Hasheminasab[1,2], DDS, OMFS; Samira Derakhshan[3,4], DDS, MSc; Dariyoush Goudarzi Pour[5], DDS, MSc; Mahdis Khazaeli Najafabadi[6], MD, MSc; Razie Hosseininavaz[5], DDS

[1]Department of Oral and Maxillofacial Surgery, Craniomaxillofacial Research Center, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran;
[2]Department of Orthodontics, University of Pacific, Dugoni School of Dentistry, San Francisco, CA, USA;
[3]Department of Oral and Maxillofacial Pathology, School of Dentistry, Tehran University of Medical Sciences, Tehran, Iran;
[4]Cancer Preclinical Imaging Group, Preclinical Core Facility, Tehran University of Medical Sciences, Tehran, Iran;
[5]Department of Oral and Maxillofacial Radiology, School of Dentistry, Tehran University of Medical Sciences, Tehran, Iran;
[6]Department of Pathology, Dr. Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran

Correspondence:
Raziei Hosseininavaz, DDS;
School of Dentistry, Tehran University of Medical Sciences, North Kargar St.,
Postal code: 14399-55991, Tehran, Iran
Tel: +98 21 22480022
Email: rshn1016@gmail.com
Received: 16 March 2024
Revised: 10 June 2024
Accepted: 18 July 2024

Abstract

Fibrous dysplasia is a slow-progressing benign condition characterized by abnormal bone formation that leads to some skeletal disorders. Although some of the fibrous dysplasia have unusual clinical and radiographic features that can lead to a challenging diagnosis, most lesions reveal an expansile bone defect due to cortex thinning.

This report presented a case of monostotic fibrous dysplasia of a 43-year-old woman with involvement of the right maxillary jaw and sinuses, which indicated unusual histopathological features. The patient was referred to the Department of Oral and Maxillofacial Surgery of Tehran University of Medical Sciences (Tehran, Iran) in 2022. Radiological and histological findings were discussed. The unusual histomorphic pattern of the lesion caused uncertainty and resulted in a missed definitive diagnosis in the primary biopsy. It was highlighted that awareness of rare histologic variants in fibro-osseous lesions especially fibrous dysplasia was required to improve diagnostic confidence.

Please cite this article as: Karimi A, Hasheminasab M, Derakhshan S, Goudarzi Pour D, Khazaeli Najafabadi M, Hosseininavaz R. Monostotic Fibrous Dysplasia with Rare Histopathologic Features: A Case Report. Iran J Med Sci. 2024;49(12):808-812. doi: 10.30476/ijms.2024.101751.3455.

Keywords ● Fibrous dysplasia of bone ● Adipose tissue ● Metamorphosis, monostatic

What's Known

- Fibrous dysplasia (FD) is a benign fibro-osseous lesion that replaces the normal bone trabeculae with connective tissue and immature woven bone.
- In most cases, histopathological examination of FD typically reveals braided bone trabeculae with immature collagen bundles and no evident osteoblastic rimming.

What's New

- This report focused on FD including fatty metamorphosis, which was extremely rare.
- In fatty metamorphosis, foam cells are found adjacent to the classical pattern of fibrous dysplasia.

Introduction

Fibrous dysplasia (FD) is a benign fibro-osseous lesion characterized by the replacement of normal bone trabeculae with connective tissue and immature woven bone.[1] The exact cause is still unknown. Men and women are affected with almost similar frequency. FD is frequently diagnosed during the second and third decades of life. When FD involves the craniofacial skeleton, it is termed "craniofacial fibrous dysplasia" (CFD). It is caused by mutation of the α-subunit of the Guanine Nucleotide binding protein, Alpha Stimulating activity polypeptide (GNAS) stimulatory protein. FD can occur in monostotic form (single bone) or polyostotic form (multiple bones). About 70%-80% of patients with FD have a monostotic form.[2] The monostatic form is about 10 times more prevalent than the polystatic form. A previous study reported that the most frequent sites of FD were the mandible and posterior maxilla.[3]

Monostatic fibrous dysplasia is diagnosed through unrelated radiographic examinations, and these lesions are often asymptomatic.[4]

The histopathologic variants of FD are restricted. In most cases, histopathological features of FD include woven, often separated bone trabeculae with no prominent osteoblastic rimming, interspersed with immature collagen bundles.[4]

There are certain rare histopathologic subtypes in the literature, the absence of awareness of this fact, particularly in small and fragmented samples in incisional biopsies, might result in a misdiagnosis of a variety of FD.[5]

This study presented a case of monocytic FD in a 43-year-old woman affecting the right maxillary jaw and sinus with rare histopathological findings. It was highlighted that the diagnosis of FD required simultaneous analysis of clinical, radiological, histopathological, and surgical findings, such as other fibro-osseous lesions, as well as familiarity with rare variants of FD.

Case Presentation

In February 2022, a 43-year-old Iranian woman with a chief complaint of swelling for an unknown duration was referred to the Department of Oral and Maxillofacial Surgery of Tehran University of Medical Sciences (TUMS). Except for hypertension, there was no remarkable past medical history. The intra-oral examination revealed a mild swelling with firm consistency in the right posterior area of the maxilla.

Radiographic features revealed a non-aggressive ground glass density lesion in the right maxillary sinus and alveolar process, with a well-defined border (figure 1A). The osteomeatal complex, nasal cavity (midline septum), lacrimal duct and sac, foramen rotundoum oval, vidian canal, and facial bone were all intact with no abnormality (figure 1B). Differential diagnoses included fibrous dysplasia, ossifying fibroma, and other fibro-osseous lesion. For a definitive diagnosis, an incisional biopsy was performed. Microscopic examinations revealed remnants of lamellar bone trabeculae demonstrating prominent reversal lines with fibrofatty marrows.

The diagnosis was mature bone trabeculae with adipose tissue. According to radiographic findings, this might not be an accurate representative of the main lesion. Therefore a deep rebiopsy or evaluation of the entire excised lesion was recommended.

The patient had an excisional biopsy of the whole lesion. The microscopic evaluation revealed a classical pattern of FD pattern consisting of several separated thin mature bone trabeculae admixed with prominent fatty connective tissue.

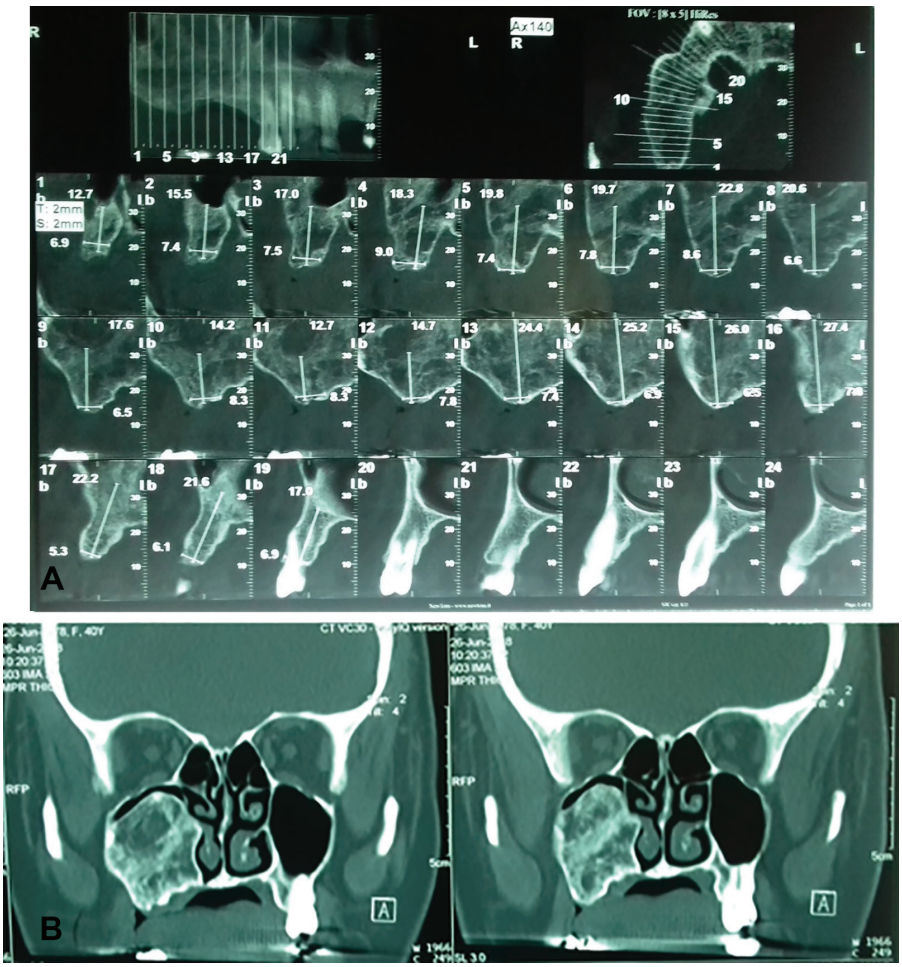

Figure 1: A) A radiopaque lesion with a ground-glass internal structure was observed. B) The computed tomography revealed a well-defined mixed lesion with no prominent expansion.

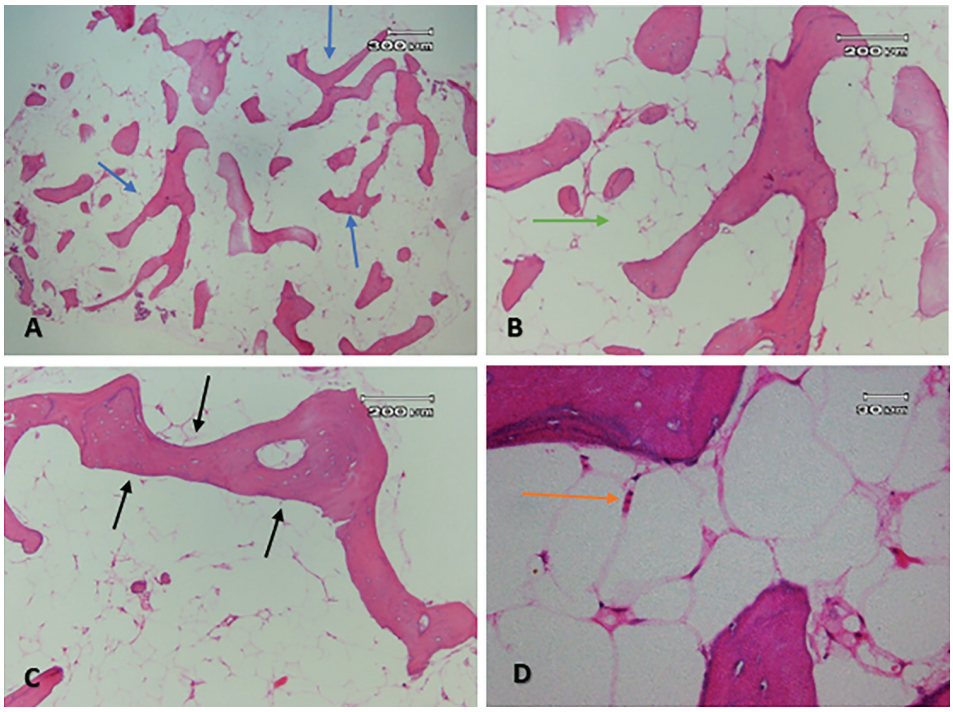

Figure 2: The histopathologic view shows A) a classical view of fibrous dysplasia, characterized by separated bone trabeculae with a "Chinese character" appearance, as indicated by the blue arrows (×40). B) Prominent fatty connective tissue demonstrated mature adipocytes among bone trabeculae, as indicated by the green arrows (×100). C) The image depicts various-sized lamellar bone trabeculae devoid of osteoblastic rimming (black arrows) within a multitude of adipocytes (×100). D) Large mature adipocytes occupied the connective tissue between bone trabeculae, as indicated by the red arrows (fatty metamorphosis), (×400).

The stroma had aberrant mature adipocytes with scar collagen bundles and fibroblastic connective tissue (figure 2). Although these microscopic views were observed in incisional biopsy of the lesion, a conclusive diagnosis of FD with rare variants might be impossible, particularly in small biopsy specimens.

Based on radiographic and histopathologic features, the final diagnosis was fibrous dysplasia with lipid infiltration (fatty metamorphosis) (figures 1 and 2). The patient's last follow-up, about 2.5 years after surgical treatment, revealed no evidence of recurrence.

This study was approved by Tehran University of Medical Sciences as a case study with an approval code of 1402-4-494-68928. The Ethics Committee of Tehran University of Medical Sciences approved this study (code: IR.TUMS.DENTISTRY.REC.1402.092). Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Discussion

Fibrous dysplasia is a developmental, tumorlike condition that can be presented in monostotic or polyostotic forms. Monocytic is more frequent, accounting for about 80-85% of all cases.[6] Bone scintigraphy is a practical method to rule out the polyostotic type of FD. Previous research indicated that the prevalence of fibrous dysplasia was about 7% of all benign bone disorders.[2] In some cases, it might be a component of Jaffe-Lichtenstein, McCune-Albright, or Mazbraud syndromes. FD primarily affects children and young adults, with the maxilla outnumbering the mandibles.[6] The most common symptom of FD patients is a slow-growing, painless swelling. However, FD has a spectrum of clinical presentations depending on the anatomical location of the lesion. In most cases, FD is painless and unilateral. Nonetheless, in some rapid growth, there may be cases of pain, paresthesia, tooth involvement, occlusion problems, or inhibition of tooth eruption.[3,5] In most cases, radiological findings revealed a ground glass pattern surrounded by a narrow rim of sclerotic bone.[1] Cortical thinning and endosteal scalloping are typical features of FD.[5] There is evidence that the typical radiographic appearance of FD can shift from less radiolucent to more mixed lesion over time, depending on age.[5]

Definite diagnosis relies on histological findings. Typical histopathologic features of FD include cellular fibrous connective tissue and trabeculae of immature bone.[6]

The features of FD might be variable and difficult to interpret. Although they are rare, awareness of patterns would facilitate accurate interpretation.[5]

Some of the distinguishing features are focal fatty metamorphosis, collagenization of stroma, stroma-rich pattern, and myxoid stroma.[5]

Fibrous dysplasia with a classical pattern has nondemarcated hypocellular, loosely arranged stroma, with spindle cells, admixed with bone trabeculae, with no prominent osteoblastic rimming. In most cases, these trabeculae are described as a "Chinese character". The fatty metamorphosis subtype features have extensive sheets of adipose tissue between the osseous trabeculae.[7]

Fibrous dysplasia with collagenized stroma demonstrates less plump cells than the classic pattern and a high level of intercellular collagen fibrosis.

Few osseous trabeculae were found in a paucitrabecular pattern with significant hypercellular stroma, including plump spindle cells.[7]

Myxoid stroma is a less prevalent pattern of FD, which is characterized by a few scattered spindle and stellate-shaped cells. This pattern shows fewer collagen bundles than collagenase stroma.[8]

In a recent publication, Shidham and colleagues reported the percentage prevalence of different types of FD.[5] Focal fatty metamorphosis accounted for 23% of all cases, followed by classical pattern (58%), stroma-collagenization (12%), stroma-rich pattern (12%), and myxoid stroma (16%).[7]

This research presented a rare type of FD with fatty metamorphosis, which indicated adipocytes in the vicinity of the classical type of FD. In cases with prominent fatty metamorphosis, small specimens might be non-representative and confusing for pathologists, necessitating an entire biopsy to make a definitive diagnosis.

A rare variant of FD might present a significant challenge for pathologists in terms of diagnosis. Understanding the histopathological spectrum or other rare histopathological patterns of FD might lead to an accurate diagnosis and interpretation. Shidham and colleagues reported that the most prevalent histopathological pattern among the rare patterns was fatty metamorphosis.[5]

It seems that being more familiar with rare variants of FD, particularly fatty metamorphosis, is essential for enhancing the accuracy of the diagnosis.

Bony fibrous dysplasia can be treated surgically or non-surgically, such as wait and watch technique or medical therapy. The severity of clinical characteristics determines the treatment strategy.[9] Medication included bisphosphonates, which were demonstrated to enhance functionality, alleviate pain, diminish the risk of fracture, and reduce bone resorption.[2] Radiotherapy and chemotherapy were contraindicated for FD due to the possibility of malignant transformation and were not curative for lesions, respectively.[8] The overall prognosis was optimal and follow-up was advised.[1] In follow-up of the patients, clinical and radiographic evaluations, particularly CT scans, are essential, given that the lesions have a considerable rate of recurrence.[8]

Fortunately, malignant transformation is observed in less than 1% of monostatic FD patients.[10] The incidence of malignant transformation was higher in patients with polyostotic FD than in those with monostotic involvement of FD.[10]

Conclusion

To establish an accurate diagnosis, it is essential to complete the following steps, including a complete patient history, precise clinical examination, perfect radiographic assessment, exact histopathological evaluations, and finally, being aware of and familiar with rare histopathological variants.

In addition, deep and adequate sampling is required for accurate diagnosis and preventing additional surgical intervention prior to primary treatment.

Acknowledgment

This study was supported by Tehran University of Medical Sciences and Health Services (Tehran, Iran).

Authors' Contribution

A.K: Study design, drafting; M.H: Study design, drafting; S.D: Data interpretation, reviewing the manuscript; D.G: Data interpretation, reviewing the manuscript; R.H: Data Gathering, Data interpretation, and drafting; All authors have read and approved the final manuscript and agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Conflict of Interest: None declared.

References

1 de Oliveira Mendes FH, dos Santos TJS, Paz JER, Evangelista IG, Carvalho FSR, Alves APNN, et al. Role of dental clinical and imaging exam during McCune Albright Syndrome

diagnosis process: Case Report. Brazilian Journal of Health Review. 2019;2:4668-81. doi: 10.34119/bjhrv2n5-066.
2 Kushchayeva YS, Kushchayev SV, Glushko TY, Tella SH, Teytelboym OM, Collins MT, et al. Fibrous dysplasia for radiologists: beyond ground glass bone matrix. Insights Imaging. 2018;9:1035-56. doi: 10.1007/s13244-018-0666-6. PubMed PMID: 30484079; PubMed Central PMCID: PMCPMC6269335.
3 Berberi A, Aoun G, Khalaf E, Aad G. Monostotic Fibrous Dysplasia of the Mandible in a 9-Year-Old Male Patient Treated with a Conservative Surgical Treatment: A Case Report and 15-Year Follow-Up. Case Rep Dent. 2021;2021:9963478. doi: 10.1155/2021/9963478. PubMed PMID: 34007492; PubMed Central PMCID: PMCPMC8110395
4 Burke AB, Collins MT, Boyce AM. Fibrous dysplasia of bone: craniofacial and dental implications. Oral Dis. 2017;23:697-708. doi: 10.1111/odi.12563. PubMed PMID: 27493082; PubMed Central PMCID: PMCPMC5292317.
5 Shidham VB, Chavan A, Rao RN, Komorowski RA, Asma Z. Fatty metamorphosis and other patterns in fibrous dysplasia. BMC Musculoskelet Disord. 2003;4:20. doi: 10.1186/1471-2474-4-20. PubMed PMID: 12946277; PubMed Central PMCID: PMCPMC200977.
6 Schoenau E, Rauch F. Fibrous dysplasia. Horm Res. 2002;57:79-82. doi: 10.1159/000058106. PubMed PMID: 12065933.
7 Davidova LA, Bhattacharyya I, Islam MN, Cohen DM, Fitzpatrick SG. An Analysis of Clinical and Histopathologic Features of Fibrous Dysplasia of the Jaws: A Series of 40 Cases and Review of Literature. Head Neck Pathol. 2020;14:353-61. doi: 10.1007/s12105-019-01039-9. PubMed PMID: 31079311; PubMed Central PMCID: PMCPMC7235148.
8 Pinto M, Braz G, Santos R, Polido D, Paixão J, de Melo S. Fibrous Dysplasia in Maxillary Bone: Case Report. Int J Oral Dent Health. 2018;4:72. doi: 10.23937/2469-5734/1510072.
9 Assiri KI. Monostotic Fibrous Dysplasia Involving the Mandible: A Case Report. SAGE Open Med Case Rep. 2020;8:2050313X20936954. doi: 10.1177/2050313X20936954. PubMed PMID: 32974022; PubMed Central PMCID: PMCPMC7493254.
10 Kaspar L, Balko J, Strnadova M, Krskova L, Maska D, Zamecnik J. Mazabraud's syndrome: A case report supported by molecular studies and review of the literature. Bone Rep. 2023;18:101685. doi: 10.1016/j.bonr.2023.101685. PubMed PMID: 37250204; PubMed Central PMCID: PMCPMC10220459.