Characteristics Of Fibrous Dysplasia At Oral And Maxillofacial Surgery

Pebrian Diki Prestya, Indra Hadikrishna**, Melita Sylvyana***, Seto Adiantoro***, Kiki Achmad Rizki*****

**Resident of Oral Surgery, Faculty of Dentistry, Padjadjaran University Bandung; Staff of Department of Oral and Maxillofacial Surgery, FKG Universitas Muhammadiyah Semarang, Indonesia*
***Department of Oral and Maxillofacial Surgery, FKG Padjadjaran University, Bandung, Indonesia*
****Department of Oral and Maxillofacial Surgery, FKG Universitas Padjadjaran, Staff of Department of Oral and Maxillofacial Surgery, Hasan Sadikin Hospital Bandung, Indonesia*
*****Department of General Surgery Division of Oncology Surgery, Faculty of Medicine Padjadjaran University, Staff of Department of Oncology Surgery, Hasan Sadikin Hospital Bandung, Indonesia*

***Correspondence*: pebrian21001@mail.unpad.ac.id**

Received 4 March 2024; 1st revision 26 July 2024; 2nd revision 8 December 2024; Accepted 31 December 2024; Published online 31 December 2024

Keywords:

Fibrous dysplasia; Reshaping; Craniofacial tumor

Abstract

Background: *Medical practitioners encounter challenges in diagnosing fibrous dysplasia due to the wide variety of investigations required, such as radiographic, histological, and clinical examinations. Fibrous dysplasia typically manifests in the first or second decade of life; these lesions tend to form at a young age and cease when somatic growth is completed.*
Method: *The design of this study is a qualitative descriptive study with a retrospective approach using medical record data of patients diagnosed as fibrous dysplasia in the Department of Oral and Maxillofacial Surgery of RSUP Hasan Sadikin in the period January 2017 — March 2023. Variables examined in this study include age, gender, anatomical location, histopathological diagnosis, and treatment provided.*
Results: *This study showed Characteristics of fibrous dysplasia patients in Oral and Maxillofacial Surgery Department RSUP Hasan Sadikin in January 2017 to March 2023 with fibrous dysplasia results by sex being highest in Women as many as 6 patients (54.54%), by age indicating the most age at 17-25 years old as 5 patients (45.45%). In addition, the highest prevalence based on histopathological diagnosis is monostotic type, which is 8 patients (72.72%). Based on the anatomical location the most is the maxillary area, which is 7 patients with left maxilla, which is 5 patients (45.45%). In addition, based on the care provided the most was reshaping with 7 patients (63.63%).*
Conclusion: *The most prevalent characteristic of patients with fibrous dysplasia who receive treatment is reshaping, which is the best therapy for the diagnosis and treatment of fibrous dysplasia.*

doi: http://dx.doi.org/10.30659/odj.11.2.188-193
2460-4119 / 2354-5992 ©2024 National Research and Innovation Agency
This is an open access article under the CC BY-SA license (https://creativecommons.org/licenses/by-sa/4.0/)
Odonto : Dental Journal accredited as Sinta 2 Journal (https://sinta.kemdikbud.go.id/journals/profile/3200)
How to Cite: Prestya *et al*. Characteristics Of Fibrous Dysplasia At Oral And Maxillofacial Surgery. Odonto: Dental Journal, v.11, n.2, p.188-193, December 2024.

Introduction

Classification of tumors in oral and maxillofacial based on *World Health Organization* divided into *benign and malignant odontogenic tumors, beningn and malignant maxillofacial bone tumors, cartilaginous tumors, benign and malignant soft tissue lesion, fibro-osseus tumors* and *haemato-lymphoid tumors*.[1] One of the oral and maxillofacial tumors is fibrous dysplasia. Fibrous dysplasia is a non-neoplastic fibro-osseous lesion characterized by the replacement of normal bone with juvenile woven bone with irregular trabeculae and fibrous tissue. Clinicians found difficulties in diagnosing fibrous dysplasia since it necessitates a range of clinical, radiological, and histological examinations.[2] Typically appearing in the first or second decade of life, fibrous dysplasia typically begin to form at a young age and cease when somatic growth was completed.[3]

Fibrous dysplasia accounts for only 2.5% of all bone tumors and about 7% of benign bone tumors.[4,5]Mandible and maxillary bones are among the craniofacial bones that can be affected by fibrous dysplasia. Zygoma, sphenoid, frontal, occipital, ethmoid, parietal, and orbital bones can also be affected. The prevalence of fibrous dysplasia is higher in the maxilla than mandible bone. According to a study by Yang et al. (2017), the maxillary bone has the highest frequency of craniofacial fibrous dysplasia, making up 28% of cases, followed by orbital (27%), mandibular (25%), frontal (22%), and temporal (12%).[6–9] Fibrous dysplasia is often related to genetics. When mutations occur in *stem cell* which is undifferentiated during the embryological phase, osteoblasts, melanocytes, and endocrine cells that mutate all cells that will carry and accelerate the occurrence of gene mutations. The result is a clinical picture in the form of multiple lesions on the bone, skin pigmentation, and endocrine disorders.[6,7]

Curette, enucleation, or excision can be used to restore the anatomical shape when the lesion is small and localized without leaving a post-surgical deformity. If the lesion is extensive and there is malocclusion and jaw disproportion, surgery is needed using an intraoral approach to correct the facial deformity which is known as the *Reshaping* technique. In this case report, it was assessed that there was an extensive fibrous dysplasia lesion, so it was decided to undergo *Reshaping*. Fibrous dysplasia is a benign tumor for which treatment and patient age are particularly important due to its high recurrence rate. In addition, there's always the chance of metastatic change, which needs to be taken into account and monitored frequently. [2,6,10] For this reason, it is necessary to conduct research on the craniofacial characteristics of fibrous dysplasia at Departement of Oral and Maxillofacial Surgery Hasan Sadikin Hospital.

Research Methodology

The research design used in this study was a qualitative descriptive study with a retrospective approach that used medical record data of patients diagnosed as fibrous dysplasia at the Department of Oral and Maxillofacial Surgery Hasan Sadikin Hospital. With inclusion criteria in the form of medical record data of patients diagnosed with fibrous dysplasia in the maxillofacial area histopathologically at the Departement of Oral and Maxillofacial Surgery Hasan Sadikin Hospital in the period January 2017 – March 2023. While the exclusion criteria are illegible and unclear medical record data. The variables

studied in this study include age, sex, anatomical location, histopathological diagnosis, and management of fibrous dysplasia.

This study used samples, namely medical record data of patients who were diagnosed histopathologically as fibrous dysplasia in the maxillofacial area at Hasan Sadikin General Hospital in the period January 2017 – March 2023 using Total sampling *using medical records of patients diagnosed with fibrous dysplasia treated by the Department of Oral and Maxillofacial Surgery Hasan Sadikin Hospital for the period January 2017 to March 2023*. This research has received ethical approval from the ethics committee of Hasan Sadikin Hospital, Bandung with ethics committee approval number No: 49/UN6. KEP/EC/2023.

Result

The results showed the characteristics of Fibrous Dysplasia in patients at Departement of Oral and Maxillofacial Surgery Hasan Sadikin Hospital from January 2017 to March 2023. Eleven patients treated by the Departement of Oral and Maxillofacial Surgery were included in the samples. The results of data collection are presented in table 1 as follows.

Table 1: Research Results Characteristic of Fibrous Dysplasia in patients at Departement Group of Oral and Maxillofacial Surgery Hasan Sadikin Hospital from January 2017 to March 2023.

Variable	**Measurement Results**	**N (%)**
Gender	Man	5 (45,45%)
	Woman	6 (54,54%)
Diagnose	Monostotic dysplasia fibrous	8 (72,72%)
	Polyostotic dysplasia fibrous	3 (27,27%)
Anatomical Location	Left Maxilla	5 (45,45%)
	Right mandible	2 (18,18%)
	Left Mandible	2 (18,18%)
	Right Maxilla and Right mandible	2 (18,18%)
Management	*Reshaping*	7 (63,63%)
	Debulking	1 (9,09%)
	Segmental Resection	1 (9,09%)
	Hemimaxillectomy	1 (9,09%)
	Hemimandibulectomy	1 (9,09%)

Based on the data from this study, it was found that 5 out of 11 patients (45.45%) had fibrous dysplasia monostotic type in their left mandible. In addition, two patients (18.18%) had fibrous dysplasia on both sides of the mandible, and two patients (18.18%) had polyostotic fibrous dysplasia. Reshaping was the most common procedure, performed on up to 7 patients (63.63%). Debulking, segmental resection, hemimaxillectomy, and hemimandibulectomy were performed on 1 patient each (9.09%). Nine patients (81.81%) underwent the first procedure, one (9.09%) underwent the second, and one (9.09%) underwent the third.

Discussion

Fibrous dysplasia is a very rare pathological condition that accounts for about 5-7% of all benign tumors, with a prevalence of about 1 in 30,000 people. Craniofacial fibrous dysplasia is a form of fibrous dysplasia that often occurs, among 50-100% of patients with polyostotic type have craniofacial abnormalities, while only 10-25% of monostotic types also affect craniofacial structures. Factors that caused fibrous dysplasia can be related to genetics and hormones. Fibrous dysplasia can occur with or without endocrine abnormalities and cafe-au-lait spots (McCune-Albright Syndrome). Moreover, fibrous dysplasia is a skeletal disorder arising from somatic mutations in *GNAS* associated with bone marrow stromal cells. *GNAS* mutations in fibrous dysplasia are gain-of-function alterations, which activate adenylyl cyclase (AC) to generate excess cyclic adenosine monophosphate (cAMP) through loss of GTPase activity of GTP-bound Gαs and abnormal activation of GDP-bound Gsα protein, generate abnormal proliferation and differentiation of mutation-bearing BMSCs.[5,11] Additionally, the management of fibrous dysplasia aims to achieve good function and aesthetics while ideally taking time, technique, and surgical indications into account. The main treatment option for fibrous dysplasia is surgery, though it is not always used for isolated, small, asymptomatic lesions. Surgery on Dysplastic fibrous lesions is performed on lesions that are painful, causing progressive deformity, and functional disorders. [12,13]

This study showed that the outcome of fibrous dysplasia in male patients was 5 patients (45.45%) while female patients were 6 patients (54.54%). According to Davidova et al (2019) states that the sex predilection between females and males is 2:1, Often fibrous dysplasia can be related to genetics and hormones, where female more than male affected by hormonal disorders. Hence, related to fibrous dysplasia possible etiology.[5,14] Fibrous dysplasia lesions can be found at various locations in the maxillofacial, usually found in the maxilla and the lesions are solitary. The study showed the maxilla with the highest prevalence being left Maxilla 5 patients (45.45%), right mandibular 2 patients (18.18%), Left mandible 2 patients (18.18%), right maxilla and right mandible 2 patients (18.18%), this could be due to the Maxilla being most susceptible to trauma compared to other parts, although the pathological mechanism is not yet so clear. Yang et al explain that there is a relationship between the characteristics of fibrous dysplasia and its pathophysiology that occurs due to the expansion of cortical bone with the gradual replacement of bone with fibrous tissue.[9]

According to Yang et al, the polyostotic type had a prevalence of 47%, while the monostotic type had a slightly higher prevalence of 56%. Riddle and Bui state that 75–80% of cases of fibrous dysplasia are of the monostotic type.[15] This study showed the prevalence of fibrous dysplasia monostotic type as

much as 8 (72.72%) and polyostotic type as many as 3 patients (27.27%) the reason for the difference in prevalence between monostotic and polyostotic is still not clearly known, there is a possibility that it occurs due to gene determination.[9]

In addition, the most common locations of monostotic type fibrous dysplasia are in the maxillary region by 28% and mandible by 25%, as well as in the polyostotic type by maxilla by 30% and mandible by 21%.[9] Similarly, with this study, the monostotic type was higher than the polyostotic with the results showing monostotic as many as 8 patients (72.72%) and polyostotic type as many as 3 patients (27.27%). This difference in prevalence between monostotics and polyostotics is closely related to genetic factors. Moreover, pathological mechanism of their involvement more at maxilla are still unclear, there is some clinical evidence indicating that local infection or trauma may result at maxilla fibrous dysplasia. As fibrous dysplasia lesions are characterized by expansion of cortical bone with gradual replacement by fibrous tissue that is firm, rubbery, and gritty, there is another possibility that the bones that are connected and closed to the maxilla are easy to be aggressively implicated.[9,14]

According to this study, the youngest age at which fibrous dysplasia is diagnosed is eleven years old. The condition's hallmark is a lesion that typically appears at ten years old, allowing for the monitoring of lesions' development from an aesthetic and functional point of view.[10,16] Age distribution also plays an important role, the results of research on fibrous dysplasia characteristics based on age revealed that there was only one patient (9.09%) aged 6–11, three patients (27.27%) aged 12–16, five patients (45.45%) aged 17–25, and one patient (9.09%) aged 36–45.

This study showed that the number of patients diagnosed with fibrous dysplasia under the age of 30 years was 9 patients (81.81%) compared to the age over 30 years which was 2 patients (6.5%). Although the fact that fibrous dysplasia can develop at any age, Chen's study Et al. revealed that 75% of individuals had the condition before the age of thirty.[17] The nature of lesions, which expand during childhood and cease in growth when the skeletal bones are grown, is responsible for this condition. Furthermore, apoptosis of mutation-carrying mesenchymal cells leading to causes the lesion activity of fibrous dysplasia to diminish with aging. The literature indicates that fibrous dysplasia manifests in the second and third decades and is brought on by delayed or incorrect diagnosis as well as by symptoms that are not readily apparent. In other ways, although most monostotic cases are detected for the first time later in life due to absence of earlier symptoms. The earlier the mutation, the more widespread and the more severe will be the manifestations. In the other hand, Most cases of fibrous dysplasia develop slowly, the early oral and maxillofacial symptoms are not obvious, and they can easily be ignored by patients.[14,18–20]

Conclusion

Characteristics of fibrous dysplasia patients in Departement of Oral and Maxillofacial Surgery RSUP Hasan Sadikin in January 2017 to March 2023 with fibrous dysplasia results by sex being highest in Women as many as 6 patients (54.54%), by age indicating the most age at 17-25 years old as 5 patients (45.45%). In addition, the highest prevalence based on histopathological diagnosis is monostotic type,

which is 8 patients (72.72%). Based on the anatomical location the most is the maxillary area, which is 7 patients with left maxilla, which is 5 patients (45.45%)

Reference

1. Slootweg PJ, El-Naggar AK. World Health Organization 4th edition of head and neck tumor classification: insight into the consequential modifications. Virchows Arch. 2018 Mar;472(3):311–3.
2. Ono K, Yoshioka N, Kunisada Y, Nakamura T, Nakamura Y, Obata K, et al. Craniomaxillofacial Fibrous Dysplasia Improved Cosmetic and Occlusal Problem by Comprehensive Treatment: A Case Report and Review of Current Treatments. Diagnostics (Basel). 2022 Sep 3;12(9):2146.
3. Zain-Alabdeen E, Abdelfattah A, Kordi O, Al-Sadhan R. The dilemma of juvenile fibrous dysplasia versus chronic osteomyelitis of the posterior mandible: A case report. Clinical Case Reports. 2022;10(10):e6379.
4. Balaji PP. Textbook of Oral and Maxillofacial Surgery. Elsevier India; 2018.
5. Karia HG, Jadhav A, Bhola N, Hande A. Monostotic fibrous dysplasia of mandible: A report of a case. Journal of Datta Meghe Institute of Medical Sciences University. 2020;Apr 1;15(2):317.
6. Neville BW, Damm DD, Allen C, Chi AC. Oral and Maxillofacial Pathology. Elsevier Health Sciences; 2015. 928 p.
7. Menon S, Venkatswamy S, Ramu V, Banu K, Ehtaih S, Kashyap VM. Craniofacial fibrous dysplasia: Surgery and literature review. Ann Maxillofac Surg. 2013 Jan;3(1):66–71.
8. Forbes-Haley C, Najran A, Nandra S, Bhola S. Craniofacial polyostotic fibrous dysplasia: A case report. Dent Update. 2019 Sep 2;46(8):768–74.
9. Yang L, Wu H, Lu J, Teng L. Prevalence of Different Forms and Involved Bones of Craniofacial Fibrous Dysplasia. J Craniofac Surg. 2017 Jan;28(1):21–5.
10. Lee JS, FitzGibbon EJ, Chen YR, Kim HJ, Lustig LR, Akintoye SO, et al. Clinical guidelines for the management of craniofacial fibrous dysplasia. Orphanet J Rare Dis. 2012 May 24;7 Suppl 1:S2.
11. Shi R, Li X, Zhang J, Chen F, Ma M, Feng Y, et al. Clinicopathological and genetic study of a rare occurrence: Malignant transformation of fibrous dysplasia of the jaws. Molecular Genetics & Genomic Medicine. 2022;10(1):e1861.
12. Lopez-Garibay LA, Guevara-Valmaña O, Telich-Tarriba JE, Navarro-Barquín DF, Haro-Alvarez N, Andrade-Delgado L, et al. Craniofacial Fibrous Dysplasia: Surgical Management and Long-Term Outcomes at a Referral Center in Mexico City. Indian J Plast Surg [Internet]. 2023 Mar 13 [cited 2023 Mar 18]; Available from: http://www.thieme-connect.de/DOI/DOI?10.1055/s-0042-1760251
13. Brucoli M, Garzaro M, Dosio C, Boffano P, Benech A. The surgical management of monostotic fibrous dysplasia of the inferior turbinate. J Stomatol Oral Maxillofac Surg. 2020 Sep;121(4):457–9.
14. 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 Jun;14(2):353–61.
15. Riddle ND, Bui MM. Fibrous dysplasia. Arch Pathol Lab Med. 2013 Jan;137(1):134–8.
16. Yang HY, Su BC, Hwang MJ, Lee YP. Fibrous dysplasia of the anterior mandible: A rare case report. Ci Ji Yi Xue Za Zhi. 2018 Sep;30(3):185–7.
17. Lisle DA, Monsour P a. J, Maskiell CD. Imaging of craniofacial fibrous dysplasia. J Med Imaging Radiat Oncol. 2008 Aug;52(4):325–32.
18. Boyce AM, Burke A, Peck CC, DuFresne CR, Lee JS, Collins MT. Surgical Management of Polyostotic Craniofacial Fibrous Dysplasia: Long-Term Outcomes and Predictors for Postoperative Regrowth. Plast Reconstr Surg. 2016 Jun;137(6):1833–9.
19. Tafti D, Cecava ND. Fibrous Dysplasia. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 [cited 2023 Jul 16]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK532947/
20. Vigano L, Powierża M, Viganò VA, Casu C. Fibrous dysplasia of the mandible: differential diagnosis. 2021 [cited 2024 Dec 19]; Available from: https://abacus.universidadeuropea.com/handle/11268/11123