DESMOPLASTIC FIBROMA OF THE MANDIBLE: A RARE CASE REPORT AND REVIEW OF THE LITERATURE

Ege Erdiner[a] , Ömer Faruk Kocamaz[b] , Mert Özlü[c] ,Serpil Altundoğan[d]

A- Ankara University, Faculty of Dentistry, Department of Oral and Maxillofacial Surgery, Turkey egeerdiner@hotmail.com
B- Ankara University, Faculty of Dentistry, Department of Oral and Maxillofacial Surgery, Turkey omerfkocamaz@ankara.edu.tr
C- Ankara University, Faculty of Dentistry, Department of Oral and Maxillofacial Surgery, Turkey, mertozlu94@gmail.com
D- Ankara University, Faculty of Dentistry, Department of Oral and Maxillofacial Surgery, Turkey, sduran@dentistry.ankara.edu.tr

Abstract

Objectives: The desmoplastic fibroma (DF) is a rare, benign, diffuse, infiltrative proliferation of fibroblasts and mature collagen that typically occurs in the first three decades of life. Jaffe first outlined it in 1958.The mandible represents the fourth most affected site and mandibular (84%) posterior location is favored. The clinical, radiological and histological presentation of desmoplastic fibroma can resemble various other jaw bone lesions. The most common treatment for desmoplastic fibroma is block resection or wide local excision, a follow up period of 3 years is recommended.

Case Report: A 12-year-old male applied to our clinic with an expanding swelling at the anterior of his mandible. The patient had a stable medical history. Extraoral examination revealed a slight expansion of the right anterior border of the mandible. Intraoral swelling resembled a hard bony texture and there was no inflammation noted. A panoramic radiograph indicated periost reaction at the lower border of the mandible and radiolucent lesion. Incisional biopsy taken. Microscopic examination pointed DF. Treatment option was marginal resection due to aggressive behavior of DP. Resection included 3 mm of healthy bone tissue around the lesion. No recurrencies was observed after 3-and 6-month follow-ups and will be followed for several years.

Conclusion: The purpose of this case report is to examine the diagnosis and treatment approaches for desmoid tumors in the maxillofacial region, a condition with very few cases reported so far, in the context of existing literature.

Keywords: Non-odontogenic Tumor, Desmoplastic Fibroma, Tumor resection

1. Introduction

Desmoplastic fibroma (DF) is a rare, benign, diffuse, infiltrative proliferation of fibroblasts or myofibroblasts and mature collagen that typically occurs in the first three decades of life[1]. They are histologically almost identical to soft tissue fibromatoses. Although the DF can be seen in any bone, the posterior mandible is the most common location [2]. It is usually a slowly growing painless lesion. Extragnathic ones usually involve the metaphyseal region of the long bones [2][3].Radiographically the DF may be seen as a multilocular or unilocular ill-defined radiolucency without mineralization. Thinned and expanded cortex or destruction of the involved bone without a sclerotic border is a common feature.[4] [5], [6]. Due to its invasive aggressive behavior and high reccurrence rate total or marginal resection is the most solid treatment. Here we present the diagnosis, treatment and follow-up stages of desmoplastic fibroma in the mandible of a 12-year-old child.

2. Case Report

A 12-year-old caucasian male applied to the Oral and Maxillofacial Surgery Department of Dental Faculty in Ankara University with an expanding swelling at the anterior of his lower jaw.(Fig.A) According to the patient's father`s statement the swelling developed 1 month ago without any pain then after 2 weeks expanding decreased and stopped. The patient had a stable medical history there was not any systemic conditions or situations related with the case. Extraoral examination only revealed a slight expansion of the right anterior border of the mandible. Intraorally, the child was in the mixed dentition stage and the intraoral expansion covered permanent second incisor, deciduous canine and first molar. Intraoral swelling resembled a hard bony texture and there was no inflammation noted on the gums or oral mucosa even so extraoral palpation of the mandible border revealed a soft tissue replaced with the mandible border. A panoramic radiograph indicated periost

reaction at the lower border of the mandible and radiolucent lesion approximately 21,7 x 10,5mm in the bone with irregular borders including erupting permanent canine teeth (Fig. B). CBCT scan showed that base of the mandibular cortex as well as the surrounding bone of the permanent erupting canine has been totally resorbed by the lesion. Lingual cortex was perforated irregularly, and slight resorption observed in the alveoler process. (Fig.C)

First of all aspiration biopsy was considered due to periost reaction like Sclerosing Osteomyelitis of Garré however there was not any abscess formation present, therefore incisional biopsy was planned. Biopsy of the lesion was obtained in general anesthesia due to child's dental fear and submitted for histological evaluation. Microscopic examination showed a tumoral lesion consisting of partially reactive bone trabeculae and spindle-fibroblastic cells. Fibroblastic cells on the collagenized floor formed bundles (Fig.D). The border of the lesion with the surrounding bone tissue was not regular. In addition, the tissue also included follicular epithelium and follicular connective tissue. Atypia, mitotic activity or necrosis were not observed in the lesional cells, and no findings were found that would suggest a malignant lesion. In immunohistochemical examination, beta-catenin positivity was observed in lesion's cell cytoplasms. Nuclear beta-catenin positivity has not been detected. Radiological and morphological findings initially matches with Desmoplastic Fibroma.

Treatment option was marginal resection due to aggressive behavior of DP, also permanent canine teeth extraction was included into the treatment on account of invasion of its periodontal surrounding by the lesion to prevent DP's highly recurrent behavior. Surgery was scheduled 1 week after biopsy with a general anesthesia. Resection approach was performed intraorally because of the lesion's accessible location. Sulcular incision was performed with flap relieving incisions and the anterior symphysis and lower border of the mandible was exposed (Fig.E). Resection took place in such a way as to include 3 mm of healthy bone tissue around the lesion then continued through the alveolar process while preserving the dentition. Permanent canine which is in erupting stage was also extracted. Lower border of mandible was curratated and alveoplasty was performed to the sharp edges of the bone. Continuity of the mandible preserved by the dentition and accompanying

alveolar process since the border of the mandible was proned to resorption. (Fig.F) Patient endured the operation well and was discharged post operative day two. First month evaluation results were promising. There was no complaint by the patient and no signs of extraoral swelling, asymmetry and collapsed tissue or paresthesia. No recurrence was observed after 3- and 6-month follow-ups (Fig.G). Extraoral palpation of the lower mandible border showed a continous hard tissue healing and the structure of the bone in the region where the lesion is located is in line with the original contours. Patient consulted to pediatric dentistry division for his occlusion management and will be followed for several years.

3. Discussion

Desmoplastic fibroma of bone has been described as a rare, locally aggressive, benign lesion that histologically resembles a desmoid tumor of the soft-tissue. Jaffe first outlined it in 1958, isolating it as a unique entity distinct from other intraosseous fibrous tumors. Since his original description, a number of small series and case reports have brought the total number of published cases to approximately 150. This tumor constitutes <1% of bone tumors and about 0.3% of all benign osseous tumors and it usually involves the tibia, scapula and femur. [7]The mandible represents the fourth most affected site. %56 of diagnosed cases is female [2]. Most of the cases are below the age of 30. The mandibular (84%) posterior location are favored [2]. Gnathic lesions are usually painless, slow-growing, asymptomatic swellings [2], [3]. Facial asymmetry, tooth displacement, root divergence, pain, trismus, and infections were the other symptoms[2], [3]. In our case we can see the male patient is 12 years old and have an asymptomatic swelling on the anterior mandibula , lesion engulfed the permanent canine.

Typically, both nuclear and cytoplasmic beta-catenin expression are observed in almost all cases of desmoid-type soft tissue fibromatosis.[8], [9], [10]. In this case nuclear beta-catenin was not observed nevertheless cytoplasmic beta-catenin had presented according to histological findings. In 2005, Hauben et al proposed that unlike desmoid-type soft tissue fibromatosis, the presence of beta-catenin mutation was not crucial for diagnosis [8]. Genetically, there is no indication to regard DF as the intra-osseous equivalent of desmoid-type soft tissue fibromatosis.[10],[11],[12].

The clinical and radiological presentation of desmoplastic fibroma (DF) can resemble various other jaw bone lesions, such as ameloblastoma, odontogenic fibroma, odontogenic cysts, aneurysmal bone cyst, chondromyxoid fibroma, central hemangioma, eosinophilic granuloma, osteomyelitis, cemento-ossifying fibroma, giant cell granuloma, and occasionally primary malignant lesions like low-grade fibrosarcoma and osteosarcoma [13].Averna et al suggested that the gold standard for the diagnosis of DF is the histologic appearance [14]. Radiographic characteristics are not definitive. They range from unilocular to multilocular radiolucencies with either well-defined or diffuse borders, often showing cortical thinning and sometimes perforation of the buccal or lingual cortex[3]. In our case, there were also perforations and thinnings in the lingual and base cortex. Histopathologically, the presence of spindle-shaped fibroblasts and collagen containing stroma played a significant role in the differential diagnosis of this lesion with Sclerosing Osteomyelitis of Garré.

The histologic differential diagnosis of DF includes myofibroma, odontogenic fibroma, cemento-ossifying fibroma, and fibrous dysplasia. The absence of nuclear pleomorphism, very low mitotic activity, lack of traces of odontogenic epithelium, and lack of cementum or bone components should help differentiate DF from other jaw bone lesions histologically [14]. The differential diagnosis included other benign spindle cell tumors as well as low-grade sarcomas, particularly fibrosarcoma. As noted, a positive immunohistochemical reaction to β-catenin and a low Ki67 index can aid in establishing a definitive diagnosis[15].

The most common treatment for desmoplastic fibroma is block resection or wide local excision, but enucleation is rarely used[16]. In a separate report by Karimi A et al. in 2020, the recurrence rate was about 40% to 47% for lesions treated by curettage or intralesional resection [17]. However, due to its aggressive behavior as demonstrated by cortical perforation, soft tissue involvement and high recurrence rate, many authors recommend complete surgical resection of the lesion with safety margins[18]. Following surgical treatment, a follow up period of 3 years is recommended[18].

In this case we preferred wide marginal resection in view of the fact that there were cortical perforations, soft tissue involvement and a deciduous tooth trapped inside the lesion. We aimed to prevent complications as fractures, malocclusions, paresthesia etc. due to the lesions' location.

4. Conclusion

Case of a 12-year-old male patient with a mandibular DF is reported. Due to its invasive behavior and soft tissue involvement, treatment approach has been wide marginal resection. A rapid hard tissue healing was observed in the relevant area, no signs of recurrence were observed in the first six-month follow-ups.

DF is a rare, locally aggressive benign bone tumor that often mimics various facial bone tumors in radiological images. To make an accurate diagnosis, it is crucial to correlate clinical, pathological, and radiographic findings. Surgical excision with a wide margin is the preferred treatment due to the tumor's high recurrence rate. In some cases, enucleation and curettage may also be effective treatment options.

5. References

1. Wippold II, F. J., White, F. V., Jamroz, G., Haughey, B., & Forsen, J. (2005). *Desmoplastic fibroma of the mandible in an infant. Pediatric Radiology, 35(9), 906–909.* doi:10.1007/s00247-005-1479-6

2. Said-Al-Naief N., Fernandes R., Louis P., Bell W., Siegal G.P. Desmoplastic fibroma of the jaw: a case report and review of the literature. *Oral Surg. Oral Med. Oral Pathol. Oral Radiol. Endod.* 2006;101:82–94. doi: 10.1016/j.tripleo.2005.03.034.

3. Inwards C.Y., Unni K.K., Beabout J.W., Sim F.H. Desmoplastic fibroma of bone. *Cancer.* 1991;68(9):1978–1983. doi: 10.1002/1097-0142(19911101)68:9<1978::aid-cncr2820680922>3.0.co;2-h.

4. Ikeshima A., Utsunomiya T. Case report of intra-osseous fibroma: a study on odontogenic and desmoplastic fibromas with a review of the literature. *J. Oral Sci.* 2005;47:149–157. doi: 10.2334/josnusd.47.149.

5. Hauben E.I., Jundt G., Cleaton-Jansen A., Yavas A., Kroon H.M., Marck E.V., Hogendoorn P.C.W. Desmoplastic fibroma of bone: an immunohistochemical study including beta-catenin expression and mutational analysis for beta-catenin. *Human Pathol.* 2005;36:1025–1030. doi: 10.1016/j.humpath.2005.07.004.

6. Flucke U., Coleman H. Who classification of head and neck tumors. In: El Naggar A., Chan J.K.C., Grandis J.R., Takata T., Slootweg P.J., editors. *Lyon.* 4th ed. 2017. p. 250.

7. Crim JR, Gold RH, Mirra JM, Eckardt JJ, Bassett LW.Desmoplastic fibroma of bone: Radiographic analysis. Radiology 1989;172:827-32.

8. Tejpar S., Noller F., Li C. Predominance of beta-catenin mutations and beta-catenin dysregulation in sporadic aggressive fibromatosis (desmoid tumor) *Oncogene.* 1999;18:6615–6620. doi: 10.1038/sj.onc.1203041.

9. Saito T., Oda Y., Tanaka K. Beta-catenin nuclear expression correlates with cyclin D1 overexpression in sporadic desmoid tumors. *J. Pathol.* 2001;195:222–228. doi: 10.1016/j.humpath.2006.01.017.

10. Saito T., Oda Y., Nakamori M., Tamiya S., Yamamoto H., Yokoyama R., Iwamoto Y., Tsuneyoshi M. Possible association between higher beta-catenin mRNA expression and mutated beta-catenin in sporadic desmoid tumors: Real-time semiquantitative assay by TaqMan polymerase chain reaction. *Lab. Invest.* 2002;82:97–103. doi: 10.1016/j.humpath.2008.05.005.

11. E. Hauben, A.M. Cleton-Jansen. Desmoplastic fibroma of bone. In WHO Classification of Tumors of Soft Tissue and Bone. Edited by Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F. IARC Lyon, 2013. p. 298. https://hdl.handle.net/2066/132068.

12. Okubo T., Saito T., Takagi T., Suehara Y., Kaneko K. Desmoplastic fibroma of the rib with cystic change: a case report and literature review. *Skeletal Radiol.* 2014;43:703–708. doi: 10.1007/s00256-013-1772-7.

13. Kahraman D, Karakoyunlu B, Karagece U, Ertas U, Gunhan O. Desmoplastic fibroma of the jaw bones: A series of twenty-two cases. J Bone Oncol. 2020 Oct 21;26:100333. doi: 10.1016/j.jbo.2020.100333. PMID: 33204607; PMCID: PMC7653059.

14. Averna R., De Flippo M., Ferrari S., Bacchini E., Rossi C. Desmoplastic fibroma of the mandible. *Acta Biomed.* 2011;82:77–81. doi: 10.1007/s12105-014-0561-5.

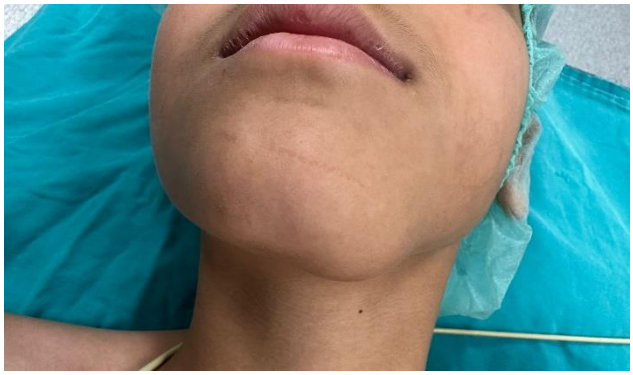
15. Mohammadi F, Shirani G, Derakhshan S, Faghihi T. Desmoplastic fibroma of the lower jaw in a 2-year-old patient; report of a rare case. Dent Res J (Isfahan). 2020 May 23;17(3):231-234. PMID: 32774802; PMCID: PMC7386374.

16.Reid EN, Lawoyin DO, Suresh L, Longwe E. Desmoplastic fibroma of the anterior mandible. Case report and review of literature. N Y State Dent J 2009;75:32-3.

17. Desmoplastic fibroma of the jaws: a case series and review of literature. Karimi A, Derakhshan S, Moradzadeh Khiavi M, Mosavat F, Mirjalili F. *Iran J Pathol.* 2020;15:134–143

18. Griffith WB. Irby Desmoplastic fibroma: report of a rare tumor of the oral structures. *Oral Surg Oral Med Oral Pathol.* 1965:269–275.

6. Figures



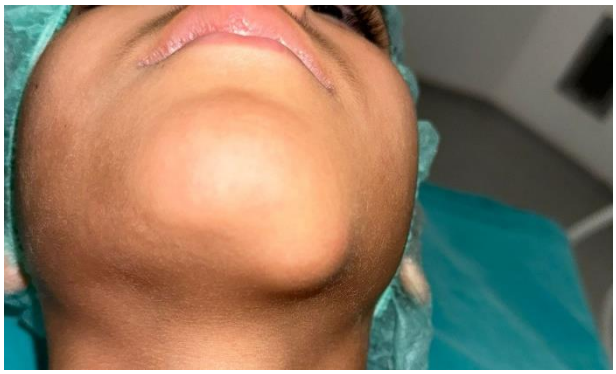


Figure A.

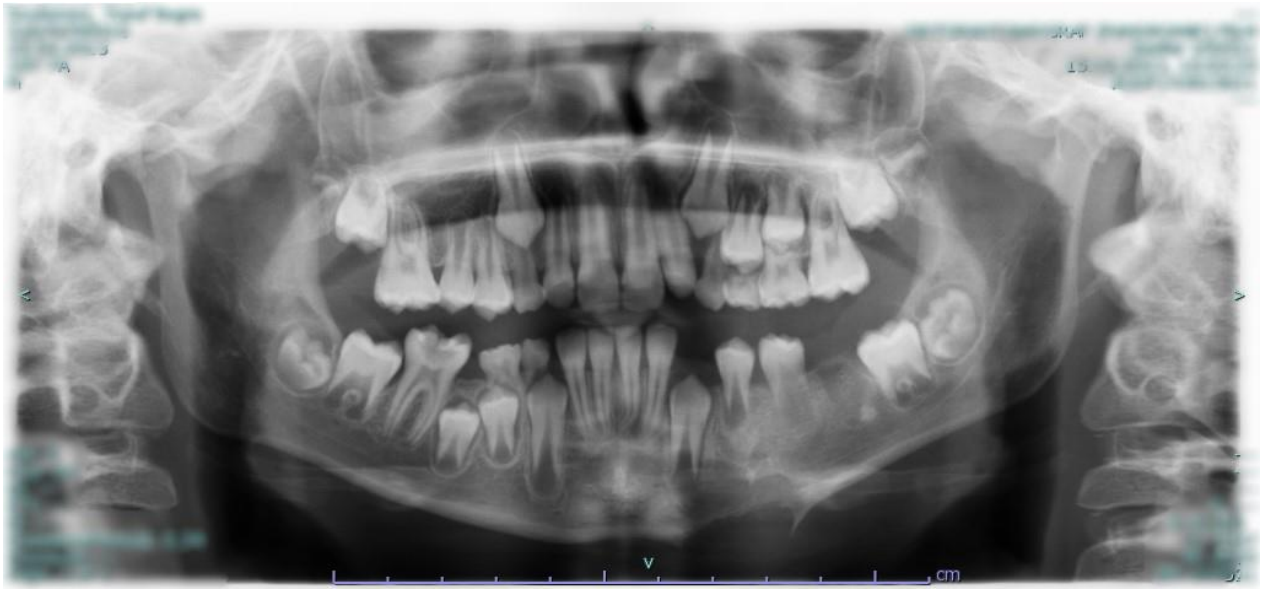


Figure B. Periost reaction at the lower border of the mandible and radiolucent lesion.

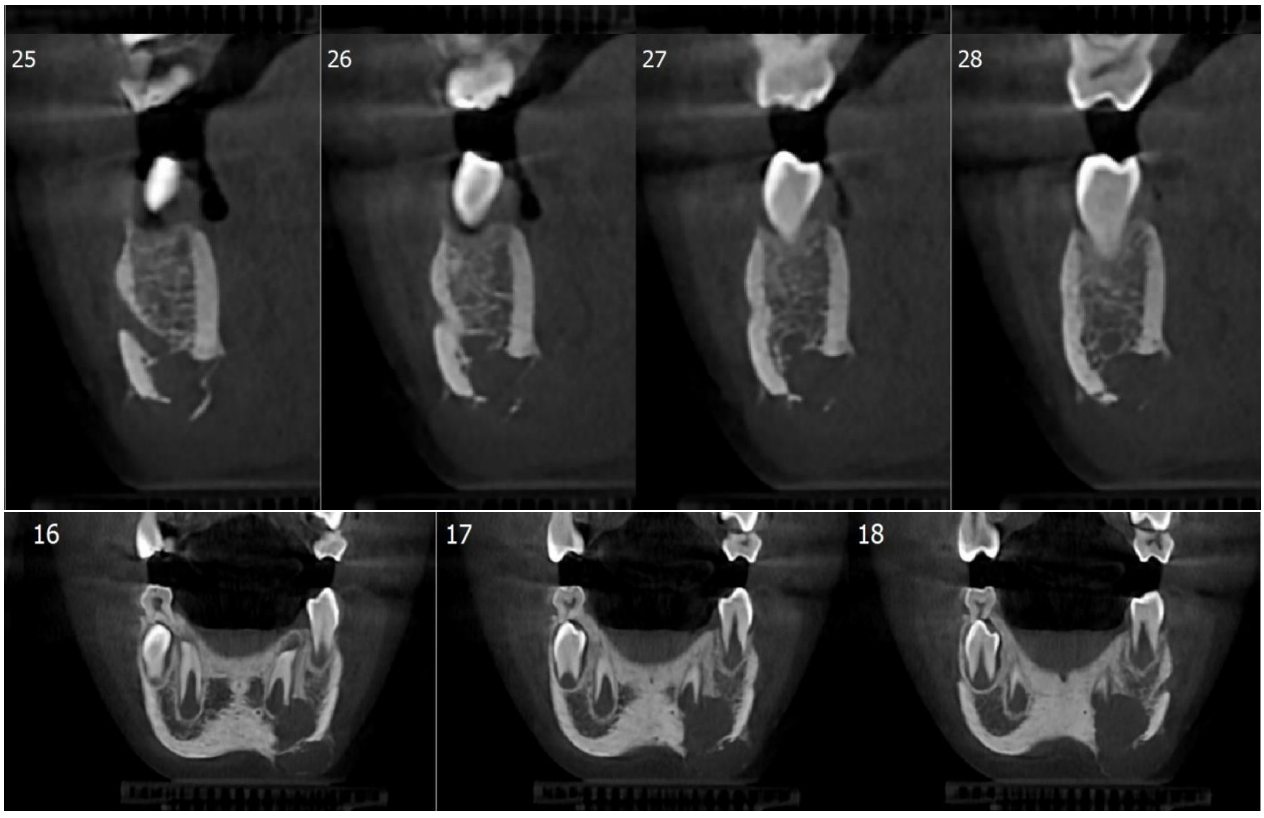


Figure C.

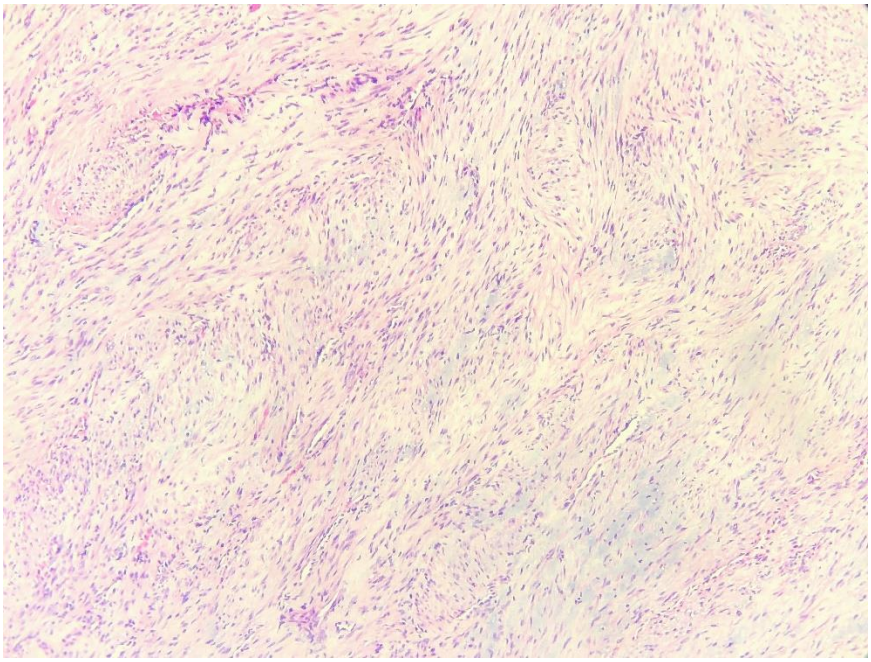


Figure D. Consists of spindle-shaped myofibroblastic/fibroblastic cell arrays in a stroma containing collagen fibers (HEX100).

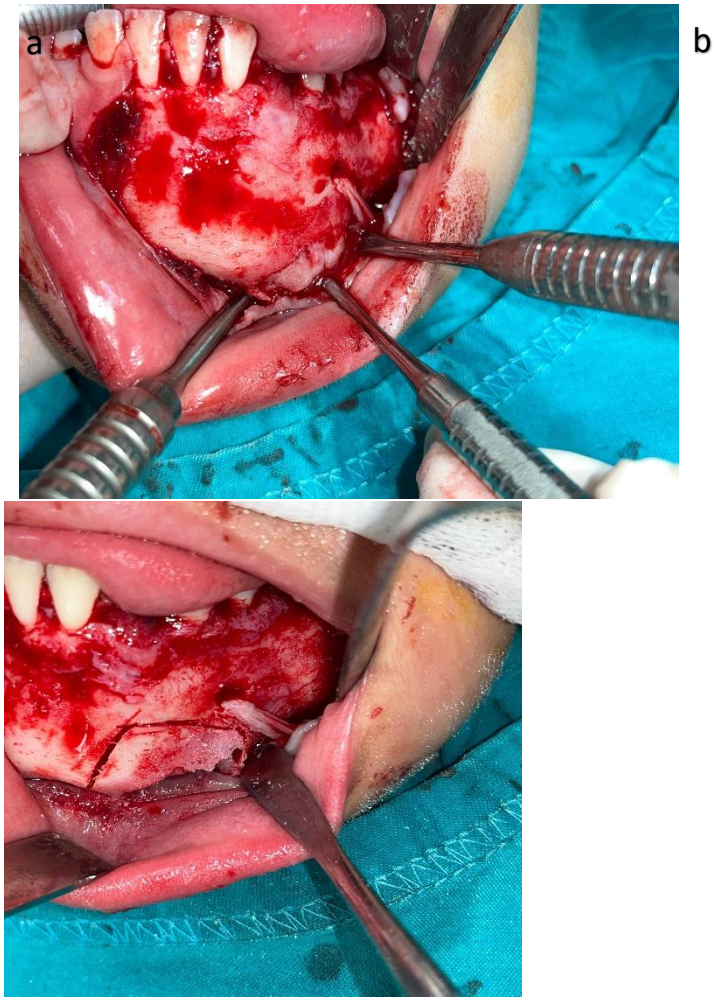


Figure E. a. Sulcular flap incision made and reached the base of mandible.

Figure E. b. Marginal resection borders lined with steel burr.

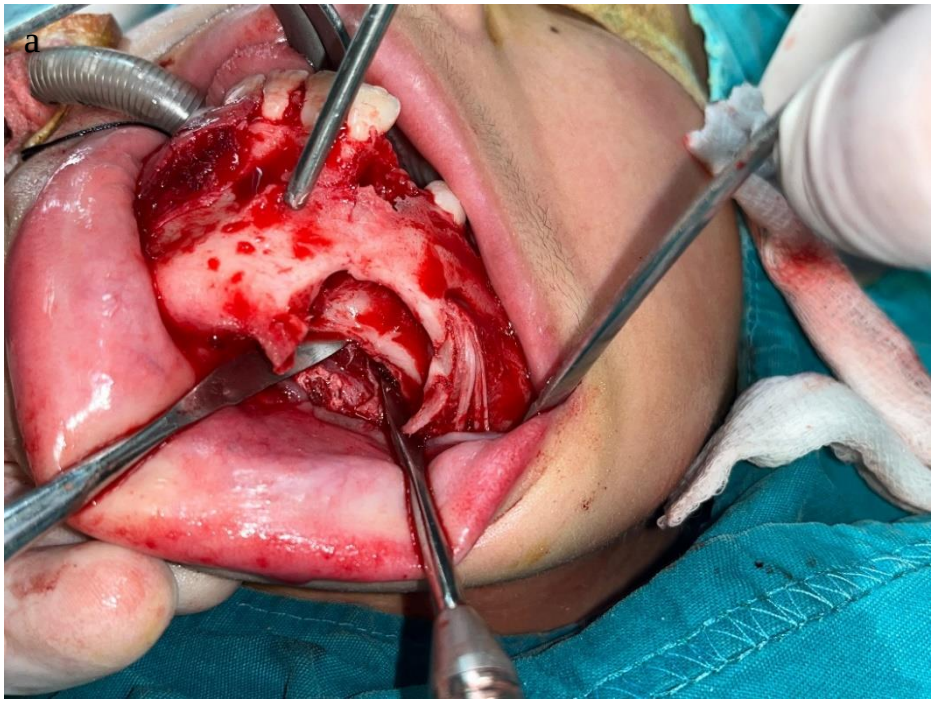


Figure F. a. Bone, lesion and effected soft tissue removed.

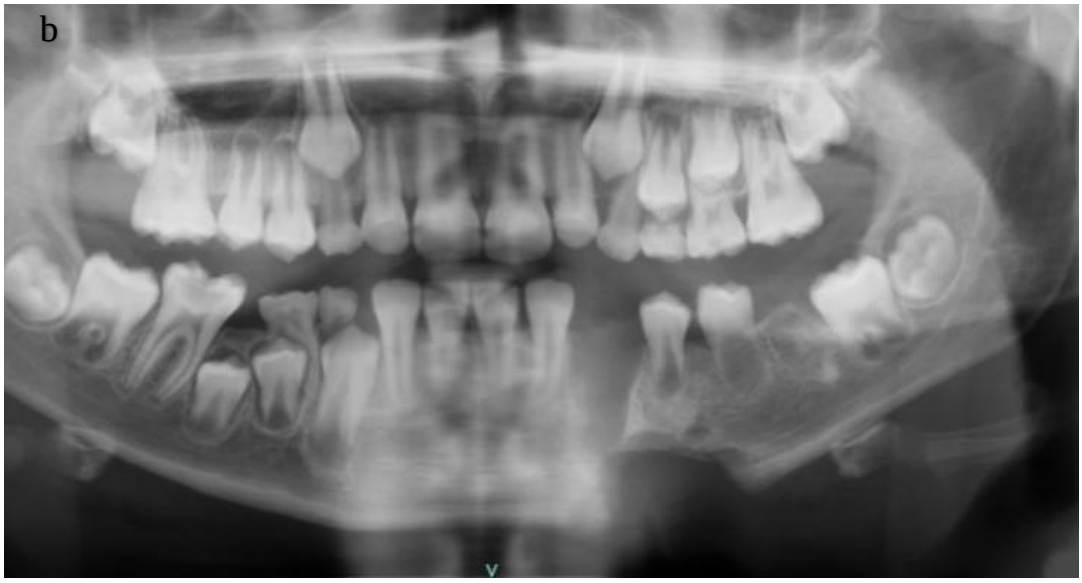


Figure F. b. Post-op 24h panoramic radiograph.

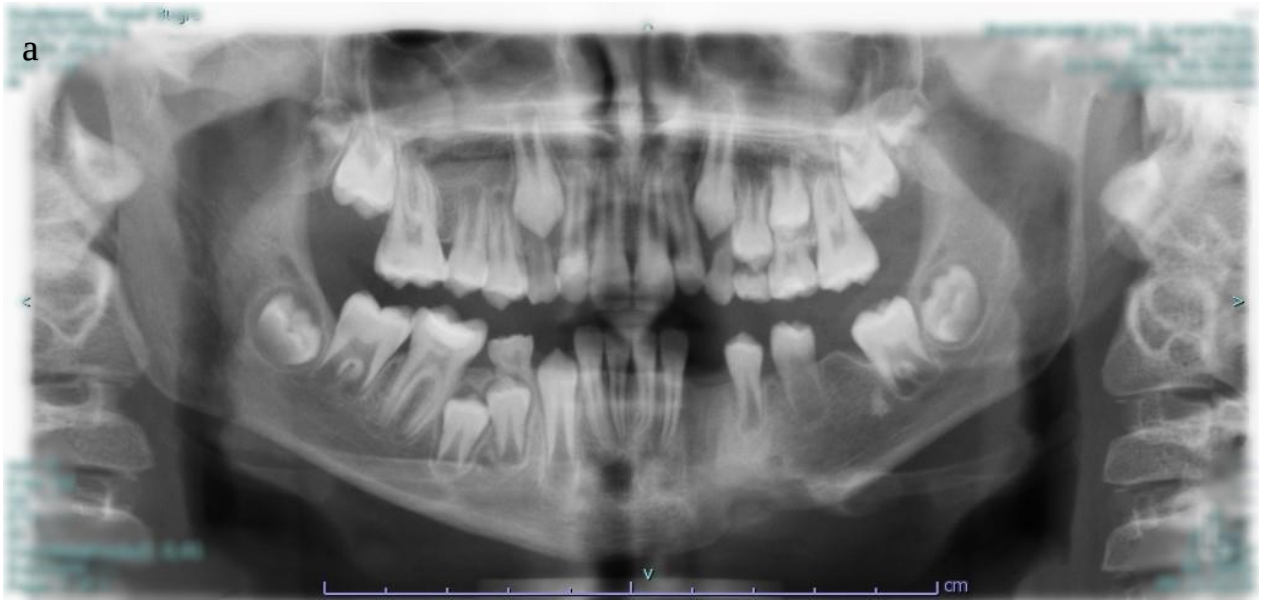


Figure G. a. Post-op third month panoramic radiograph.

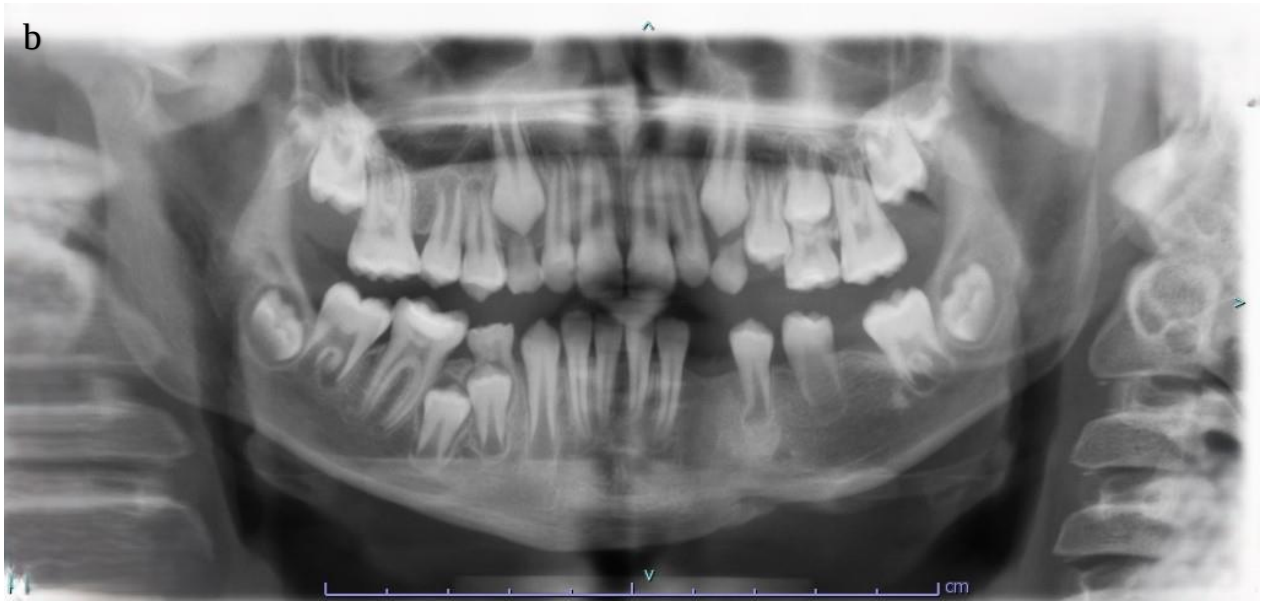


Fig. G. b. Post-op sixth month panoramic radiograph.