EurAsian Journal of Oral and Maxillofacial Surgery
Official publication of AÇBİD (Association of Oral and Maxillofacial Surgery Society)

CASE REPORT

Soft and Hard Tissue Changes in the Oral and Maxillofacial Region in Patients with Neurofibromatosis: Rare Case Report

Günay YAPICI YAVUZ,DDS,Phd[1], Mahmut KOPARAL,DDS,Phd[1], Aydin KESKINRUZGAR,DDS,Phd[1], Şeyma BAYAZIT,Dr [1]

[1]Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, Adiyaman University, Adiyaman, Turkey

Abstract

Introduction

Neurofibromatosis is a group of autosomal dominant genetic disorders characterized by multiple cutaneous lesions and tumors of the nervous system. NF is categorized into two, NF type-1 and NF type-2. Neurofibromatosis-1 is a neurocutaneous-skeletal disorder with variable phenotypic description. Craniofacial skeletal symptoms of the disease are observed in the jaw deformations and impacted, missing and displaced teeth can also be seen. The purpose of this report is to present a case of NF1.

Case report

A 23-year-old male patient was referred to the Department of Maxillofacial Surgery, Faculty of Dentistry, Adiyaman University due to abscess of molar teeth. As a result of the panoramic radiography taken from the patient, the second and third molar teeth were impacted in the left mandibula.

Conclusion

In neurofibromatosis, bone and soft tissue involvement can be seen in the maxillofacial region. They are also lesions that can cause the impacted teeth.

Keywords: Neurofibromatosis, maxillofacial region, autosomal dominant.

Introduction

Neurofibramatosis is an autosomal dominant disease.[1] The term neurofibromatosis (NF) is used for a genetic disorders that affect cell growth of neural tissues. The most common are 2 NF forms: Neurofibromatosis type 1 (NF-1) and Neurofibromatosis type 2 (NF-2). [1,2]

Neurofibromatosis type 1 (NF-1), also known as Von Recklinghausen disease.[2] NF-1 is the most common type of NF and accounts for approximately 90% of all cases, with one occurrence at 3000 births.1-3 There is a mutation in the 17q11.2 chromosome known as the NF-1 gene.[4] Neurofibromatosis 2 (NF-2) results from mutations in chromosome 22 of the NF-2 gene.[5] 50% of NF-1 patients have a family history of the disease.[6]

The most common symptoms of NF-1 in the skin are cutaneous neurofibromas and cafe au lait points.[5] The cafe-au-lait spots are clinically yellow-brown spots. The presence of at least 6 diameters greater than 1.5 cm in diameter is pathognomonic for NF-1.[7]

Superficial nodular lesions resulting from the proliferation of neural tissues in the skin are called neurofibroma. It is especially localized in the head and neck region. Due to of their size, nodules may cause problems with aesthetics and function.[8] In this study, we present the findings of oral and maxillofacial areas in a patient diagnosed with NF-1.

Case Report

A 23-year-old male patient who was diagnosed as NF-1 by the dermatologist applied to the our clinic due to swelling and infection on the left side of the mandible. The patient was contacted with help from his family because of the difficulty of sensing and speaking.

In the extraoral examination, facial asymmetry was observed. Due to the mass in the left posterior region of the mandible, bone contact could not be palpated in the left mandibula. Left lateral canthus of the left eye and sagging in the left ear, as well as hypertrophy in the left ear were observed. There were cafe au lait spots on the neck. (Figure 1 and Figure 2)

In intraoral examination, revealed growth on the left side of the tongue. There were dilated tongue papillae on the related side. In the mandible anterior teeth region and left posterior region,

Corresponding Author: Aydin Keskinruzgar
Associate Professor
Address: Adiyaman University, Faculty of Dentistry, Oral and Maxillofacial Surgery, Adıyaman
Mobile: +90(416).2251920
e-mail: akeskinruzgar@gmail.com

normal colour, non-ulcerated, painless were observed gingival hyperplasia. The patient's oral hygiene was deficient. In the area of the patient's complaint, deep caries were detected teeth 33 and 34, also there was infection due to.

The patient's panoramic radiography revealed radiolucent lesion with together 2 and 3 molar teeth impacted in the left mandibular posterior region. And lesion appeared to infiltrative growth pattern with no prominent borders. Foramen mandible and foramen mentale had expansion. (Figure 3)

The patient was prescribed antibiotics due to infection. Then, under general anesthesia, teeth 16, 26, 33, 34, 46 and 47 were taken. Impacted teeth 37 and 38 were taken with surgical intervention. The lesion was excluded.

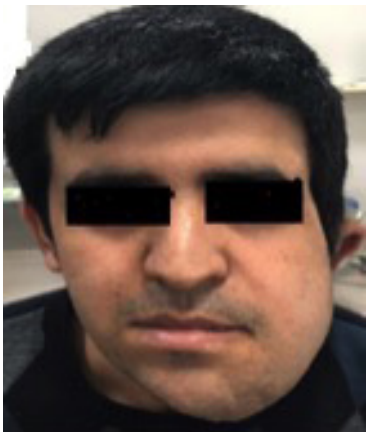

Figure1: NF-1 extraoral image

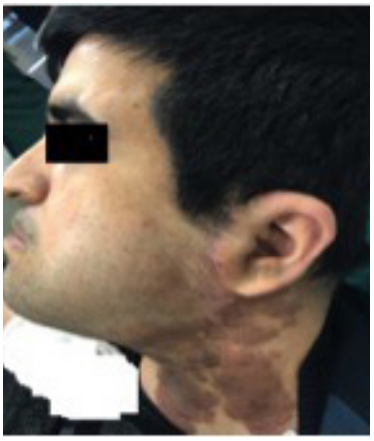

Figure 2: Cafe au lait spots on the neck

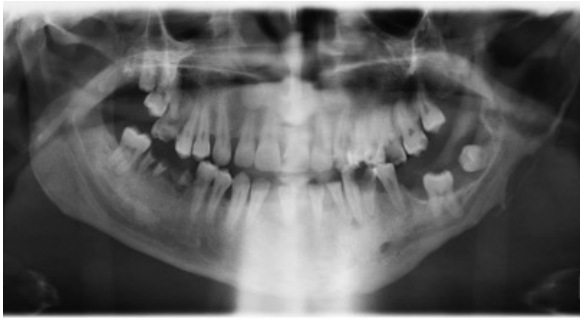

Figure 3: Left mandible affected by NF-1

Discussion

NF1 patients may exhibit hypoplasia or hyperplasia of the jaws and facial changes caused by the buccal bone and temporomandibular joint. Osseous changes can be in the bone or a product of soft tissues that grow against the bone.[6] In our case, due to the pressure on the surface of the NF bone, deformation of the bone was observed in the left mandible. Localized neurofibroma is the most common type of neurofibroma in NF-1 patients. The most frequently affected areas in the head and neck region are the scalp, cheek, neck and oral cavity.[2]

Oral findings can be found in almost 72% of NF1 patients.[6] Neurofibromas occurring in craniofacial structures can spread into the mouth, causing abnormalities that will prevent the function of oral structures, disrupting the teeth series and causing the teeth to be positioned incorrectly.[9] In our patient, NF-1 caused teeth 37 and 38 to be impacted. In particular, the expansion of the alveolar duct, foramen mandible and foramen mentality in the mandible is considered to be an oral manifestation of NF1.[3] Periodontal localizations may cause mobility in the teeth and hyperplasia in the gums. The most common oral manifestation of NF-1 is dilation of tongue fungiform papillae that occurs in about 50% of cases.[6] Dilation of tongue papillae was observed in our case report in accordance with the literature.

NF-1 patients may have abnormal speech rate, hoarseness and / or hypernasal speech.[5] In our case, mental retardation and perception deficits were observed in the patient.

Conclusion

NF-1 may show symptoms in the hard and soft tissue in the maxillofacial region. Diagnosis and follow-up of the disease is important for the patient's life functions.

References

1. Akoğlu G, Gazyağcı AS, Kutluer İ. Neurofibromatosis: Evaluation of Clinical Features of 11 Cases. Turk J Dermotol 2013;7:196-200.
2. Lee L, Yan YH, Pharoah MJ. Radiographic features of the mandible in neurofibromatosis A report of 10 cases and review of the literature. Surg Oral Med Oral Radiol Endod 1996;81:361-7.
3. Visnapuu V, Peltonen S, Tommissalo T, , Pentonen J, Happonen RP. Radiographic Findings in the Jaws of Patients With Neurofibromatosis. J Oral Maxillofac Surg 2012; 70:1351-1357.
4. Visnapuu V, Pienihäkkinen K, Peltonen S, Happonen RP, Peltone J. Neurofibromatosis 1 and dental caries. Clin Oral Invest 2011;15:119-121.
5. Visnapuu V , Peltonen S , Alivuotila L , Happonen RP, Peltonen J. Craniofacial and oral alterations in patients with Neurofibromatosis 1. Orphanet J Rare Dis. 2018; 13: 131.
6. Cunha K G, Barboza E P, Dias E P, Oliveira F M. Neurofibromatosis type I with periodontal manifestion. A case report and literature review. Br Dent J 2004;24;196(8):457-60.
7. Ucok C, Tuncer N, Günhan O. Neurofibromatosis (von reclinghausen's disease) a case report. Journal of Istanbul University, Fakulty of Dentistry 2012;31.76-79.
8. Çalık Yılmaz BC, Alaçam A. A Pathology mimicking dental abscess: facial plexiform neurofibroma. Acta Odontol Turc 2019;36(1):28-32.
9. Ayhan Bani A, Türköz Ç. Syndromes And Accompanying Craniofacial Anomalies. A.Ü.Dis.Hek. Fak. Derg. 2012; 39(1):35-47.