Case Report

Supernumerary Premolars in a Patient with Neurofibromatosis Type-1

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Abstract

Neurofibromatosis is a relatively common dominantly inherited neurodermal dysplasia that affects many parts of the body including oral cavity. The patient, an 18-year-old girl diagnosed as neurofibromatosis Type-1 (NF1) exhibited four supernumerary premolars. In addition, panoramic radiograph of the patient showed enlarged right mandibular canal and foramen, unilocular radiolucent areas in the rami of the mandible, and increase in the dimension of coronoid notch on both sides of the mandible. The concomitant presentation of supernumerary teeth with NF1 reported here might be due to genetic linkage or could be a chance occurrence.

Keywords: Neurofibromatosis Type-1, premolars, supernumerary teeth

Introduction

Neurofibromatosis Type-1 (NF1) is a dominantly inherited neurodermal dysplasia caused by mutations in the NF1 gene. The NF1 gene controls neurofibromin production which is believed to act as a tumor suppressor.[1] The condition is clinically classified into two main types: peripheral type or NF1 and central or NF2. NF1 also known as von Recklinghausen's disease is a relatively common autosomal dominant disorder carried on chromosome 17q11.2 and affects the growth of neural tissue. NF1 accounts for 90% of all cases and has an incidence of 1 in 3000–3500 live births with no gender or racial predilection.[2-5] Neurofibromas are the hallmark of the disease. The National Institutes of Health Consensus Development Conference 1988 has defined criteria for diagnosis in 1988.[6] An individual is said to have NF1 if two or more of the following features are observed: café au lait pigmentations of skin, neurofibromas, freckling in the axilla or in the inguinal regions, optic glioma, pigmented iris hamartomas, osseous lesion, and a first-degree relative with NF1. The patients with NF1 may also have scoliosis, learning disabilities, short stature, and brain lesions. Intraoral lesions are common in NF1 accounting for 72% of cases.[3-5,7] They are frequently located on the tongue and buccal mucosa. Numerical aberrations in dentition have also been reported in NF1 patients and these are mostly in the form of hypodontia.[8] The purpose of this article is to present a case of NF1 with supernumerary premolars.

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Case Report

An 18-year-old female reported to our department with the chief complaint of gum bleeding on brushing. She had a short stature. On examination, multiple small nodules were observed on her face and neck [Figure 1]. History revealed that these soft tissue masses were not present since birth, but appeared after puberty. The patient reported that she had been diagnosed as NF1 4 years ago. Her family history revealed that her father also had NF1.

Intraoral examination of the patient showed four erupted supernumerary premolars. Morphologically, the two maxillary supernumeraries were conical, while the mandibular supernumerary premolars were supplemental in shape [Figures 2 and 3]. The patient gave a history of teeth extraction.

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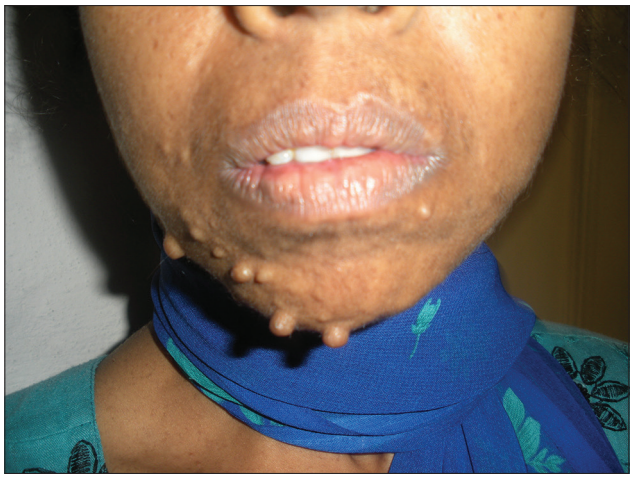

Figure 1: The patient with multiple neurofibromas.

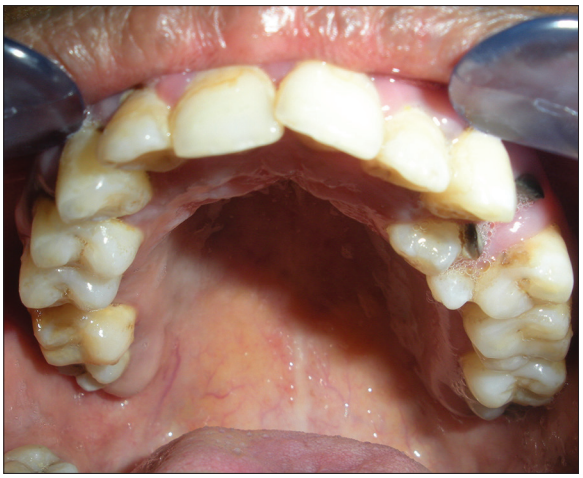

Figure 2: Two palatally erupted conical maxillary supernumerary premolars.

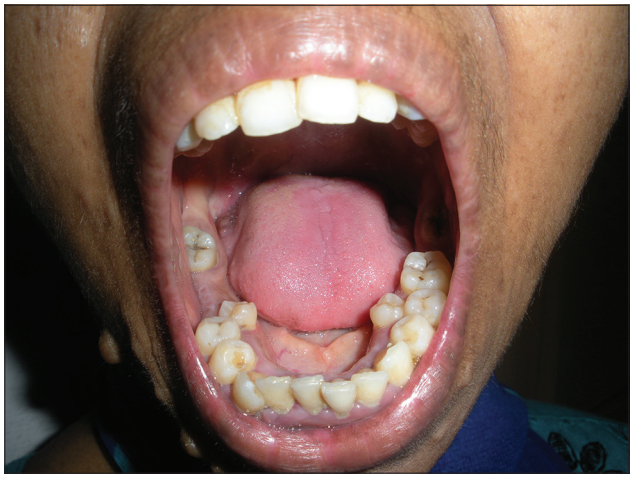

Figure 3: Lingually positioned supplemental supernumerary mandibular premolars.

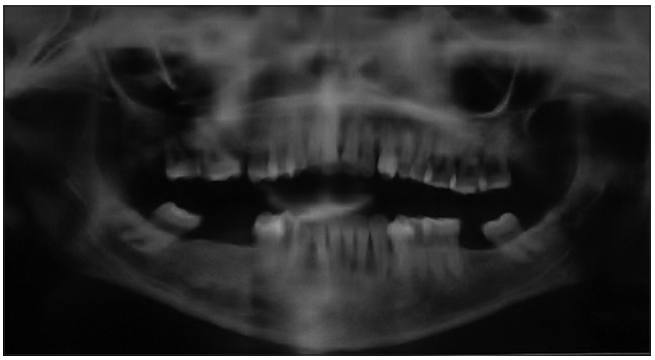

Figure 4: Orthopantomograph showing radiolucent changes in the mandible.

Orthopantomogram was advised. The panoramic radiograph of the patient showed enlarged right mandibular canal and foramen. In addition, unilocular radiolucent areas were observed in the rami of the mandible. There was an increase in the dimension of coronoid notch on both sides of the mandible [Figure 4].

Retained root of the maxillary left first premolar was extracted under local anesthesia. Oral prophylaxis was performed and oral hygiene instructions were given.

Discussion

NF1 is an autosomal dominant trait with high penetrance and variable expressivity. In 50% of cases, NF1 patients do not have a positive family history of the disease. In the present case, the father of the patient also had NF1. Although neurofibromas are the principal trait of NF1, it is a disease of the bone and jaw bones are sometimes involved.[2,8]

Intraoral lesions are common in NF1. They are frequently located on the tongue and buccal mucosa. The clinical features of NF1 are usually identifiable during early adolescence or even in childhood and the condition worsens slowly.[2] Therefore, early detection and appropriate therapeutic measures should be taken to minimize complications. Dental abnormalities of NF1 include hypodontia, supernumerary teeth, and impacted teeth. The patient in this report showed supernumerary premolars.[1-4] In addition, bone abnormalities such as intrabony neurofibromas, hypoplasia of the jaw, enlarged mandibular foramen, coronoid notch deformity, and soft tissues abnormalities such as oral neurofibromas are also reported in patients with NF1.

The most frequent radiographic features of mandible are enlargement of mandibular canal and/or foramen.[1,2] In this report, panoramic radiograph exhibited enlarged right mandibular canal, mandibular foramen, and an increase in the dimension of coronoid notch.

Supernumerary premolars account for 3%–9% of all supernumerary teeth, and their prevalence varies from 0.29% to 0.64% of general populations.[9,10] Although 90% of all supernumeraries occur in the premaxillary area, supernumerary premolars are more common in the mandible and are three times more frequent in males than in females.[9,10] This patient showed four supernumerary premolars. NF1 is an autosomal

dominant disorder with a variable phenotypic expression. The etiology of supernumerary teeth is not clearly understood. Both genetic and environmental factors are suspected. The presence of multiple supernumerary teeth in a NF1 patient might have a genetic linkage, or it could be a chance occurrence. Further genetic studies are required in this direction.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Neville BW, Damm DD, Allen CM, Bouquot JE. Oral and Maxillofacial Pathology. 3rd ed. Philadelphia: Elsevier; 2009. p. 528-31.
2. Bardellini E, Amadori F, Flocchini P, Conti G, Piana G, Majorana A. Oral findings in 50 children with neurofibromatosis type 1. A case control study. Eur J Paediatr Dent 2011;12:256-60.
3. Javed F, Ramalingam S, Ahmed HB, Gupta B, Sundar C, Qadri T, *et al.* Oral manifestations in patients with neurofibromatosis type-1: A comprehensive literature review. Crit Rev Oncol Hematol 2014;91:123-9.
4. Cunha KS, Barboza EP, Dias EP, Oliveira FM. Neurofibromatosis type I with periodontal manifestation. A case report and literature review. Br Dent J 2004;196:457-60.
5. Kaplan I, Calderon S, Kaffe I. Radiological findings in jaws and skull of neurofibromatosis type 1 patients. Dentomaxillofac Radiol 1994;23:216-20.
6. Neurofibromatosis. Conference statement. National Institutes of Health Consensus Development Conference. Arch Neurol 1988;45:575-8.
7. Geist JR, Gander DL, Stefanac SJ. Oral manifestations of neurofibromatosis types I and II. Oral Surg Oral Med Oral Pathol 1992;73:376-82.
8. Friedrich RE, Reul A. Supernumerary molars and wisdom tooth shape alterations in patients with neurofibromatosis type 1. J Oral Maxillofac Res 2017;8:e5.
9. Cervellera C, Del Pin D, Dissaux C. Neurofibromatosis type I and supernumerary teeth: Up-to date review and case report. Med Buccale Chir Buccale 2017;23:84-9.
10. Cho SY. Supernumerary premolars associated with dens evaginatus: Report of 2 cases. J Can Dent Assoc 2005;71:390-3.