

Dental Management of Neurofibromatosis Type 1: A Case Report

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Introduction

Neurofibromatosis Type 1 (NF1) is a rare genetic disorder characterized by an increased risk of benign and malignant tumors and other physical and neurological manifestations. The most prevalent manifestations are multiple tumors of nerves and skin (neurofibromas), as well as areas of abnormal skin color (pigmentation) including light brown discolorations (café-au-lait spots) under the arms or in the abdominal region.⁽¹⁾ Neurofibromatosis can affect most of the oral structures, including tongue, alveolar mucosa, lip, gingiva and teeth, causing bone lesions, high score of plaque and bleeding tendencies and increase risk of caries.⁽²⁾ Children with NF1 are also reported to have class III molar relationship and unilateral posterior crossbite.⁽³⁾ This case report discusses the clinical presentation and dental management of a 5-year-old male patient with NF1 treated under general anesthesia.

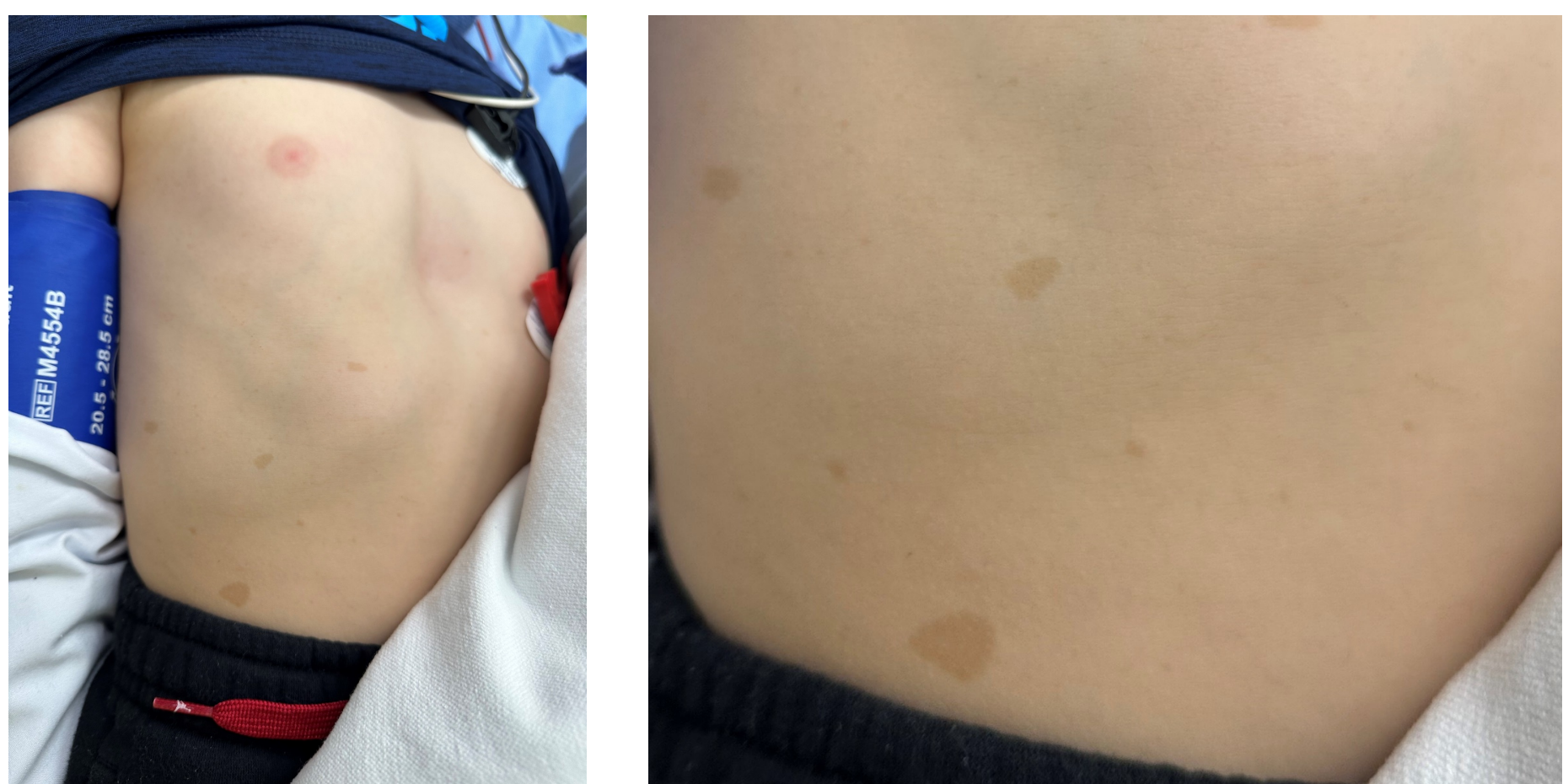


Figure 1: Light brown skin discoloration (café-au-lait spots) in the abdominal region

Case Report

This presentation discusses the case of a 5-year-old male patient referred to Tufts University School of Dental Medicine with a chief complaint of multiple caries and nocturnal pain related to lower right teeth. The patient's medical history is Neurofibromatosis type 1. Current medications include Amoxicillin and Melatonin. The patient has no known drug allergies. Extraoral examination revealed pigmented spots “café-au-lait” in the abdominal region (Figure 1). The clinical examination indicated Angle class III malocclusion; multiple caries and intra-oral swelling related to tooth #S (Figure 2). Radiographic assessment revealed bifurcation and periradicular pathology related to tooth #S (Figure 3). A full-mouth dental rehabilitation was performed under general anesthesia, with a 3-month follow-up.



Figure 2: Intra-oral Photographs



Figure 3: Preapical Radiograph for tooth #S



Figure 4: Upper Occlusal Radiograph

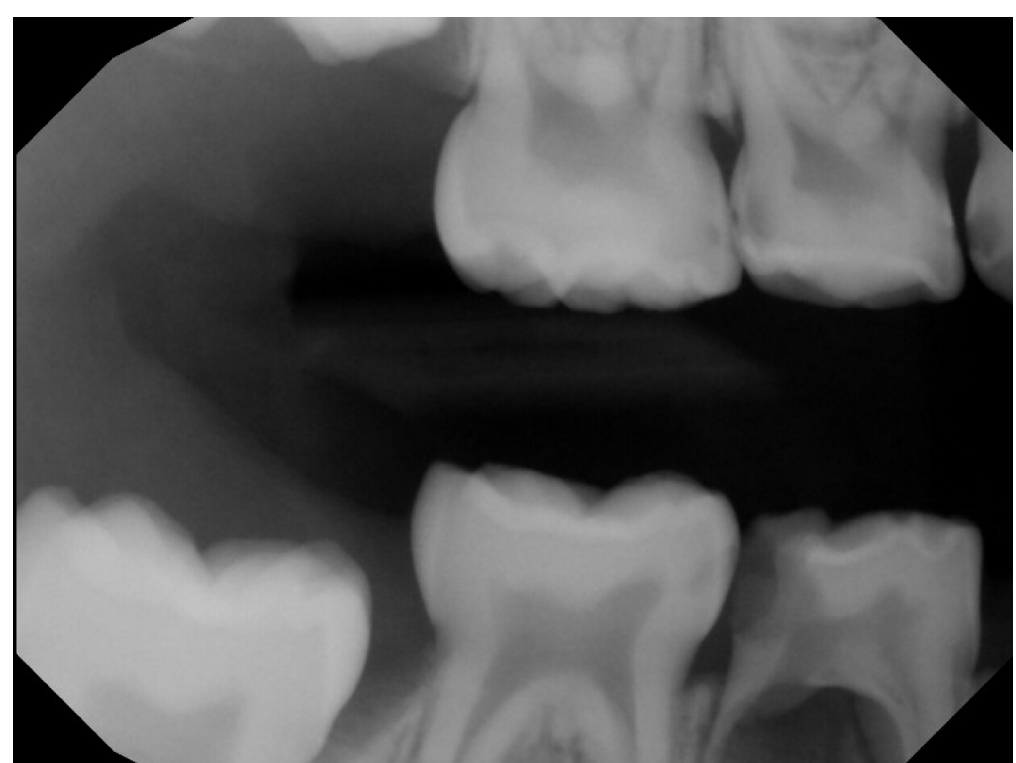


Figure 5: Right Bitewing

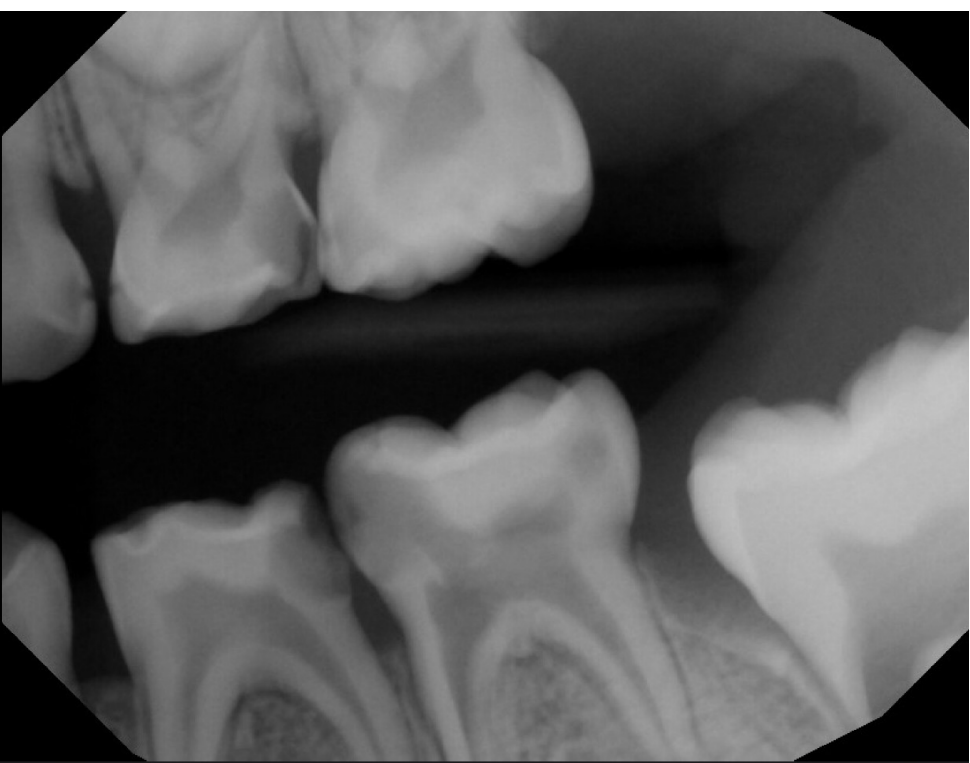


Figure 6: Left Bitewing

Managment

Patient is 5-year-old with Frankl I score and multiple caries. Therefore, dental rehabilitation under general anesthesia was done.

Treatment phase:

- Resin restoration on tooth #C(DF), #E(MFL), #F(MFL), #H(DF) was performed.
- Pulpotomy was done on tooth #I, K and L.

- Stainless steel crowns were placed on teeth #A, B, I, J, K, L and T.
- Extraction was done for tooth #S.

Maintenance phase:

Due to patient's high caries risk, a periodic exam, prophylaxis, and fluoride varnish application every 3 months were recommended.

Conclusion

Patients with NF1 exhibit many oral manifestations, such as an increased tendency of plaque accumulation, gingival bleeding and caries. Therefore, it's important for the oral healthcare providers to place more emphasis to the oral hygiene education and the important of recare appointments. Children with NF1 also are more likely to have class III malocclusion, thus, early orthodontic evaluation needs to be done to limit future invasive orthodontic treatments.

References

1. National Institutes of Health: Consensus Development Conference. Neurofibromatosis. Conference Statement. Arch Neurol. 1988;45:575-578.
2. Shapiro S, Abramovitch K, Van Dis M, Skoczylas L, Langlais R, Jorgenson R, et al. Neurofibromatosis: Oral and radiological manifestations. Oral Surg. 1984; 58:493-498.
3. Lammert M, Friedrich RE, et al. Early primary tooth eruption in neurofibromatosis 1 individuals. Eur J Oral Sci. 2007 Oct;115(5):425-426.