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Original Research Paper

Dental Science

" NEW SYNDROME ASSOCIATING GINGIVAL FIBROMATOSIS AND DENTAL ABNORMALITIES – A CASE REPORT"

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ABSTRACT Gingival Fibromatosis is the overgrowth of the gingiva characterized by an expansion and accumulation of the connective tissue with the rarely presence of an increased number of cells. It is hereditary or is induced as a side effect of systemic drugs. As an inherited disorder, Gingival Fibromatosis may be part of a genetic syndrome or it may be isolated, in which case it is referred to as hereditary gingival fibromatosis. Some of the cases of Hereditary Gingival Fibromatosis associated with dental abnormalities like Amelogenesis Imperfecta. It is a general term for a number of conditions that affect enamel formation and/or calcification. The purpose of this article is to report manifesting syndrome characterized by Gingival Fibromatosis associated with dental abnormalities, including generalized thin hypoplastic Amelogenesis Imperfecta, intrapulpal calcifications, delay of tooth eruption, pericoronal radiolucencies in unerupted teeth, dental agenesis and root dilacerations.

KEYWORDS : Hereditary Gingival Fibromatosis, Amelogenesis Imperfecta.

INTRODUCTION

Gingival Fibromatosis (GF) is the overgrowth of the gingiva characterized by an expansion and accumulation of the connective tissue with the rarely presence of an increased number of cells.[1] It is hereditary or is induced as a side effect of systemic drugs.[2] As an inherited disorder, GF may be part of a genetic syndrome or it may be isolated, in which case it is referred to as hereditary gingival fibromatosis (HGF).[3] HGF is traditionally considered an autosomal-dominant disease, whereas pedigree analyses of GF syndromic forms are consistent with a mendelian transmission pattern.[4] Whereas Amelogenesis Imperfecta (AI) is a general term for a number of conditions that affect enamel formation. In most cases, an autosomal dominant mode of inheritance is involved; however, the disease is frequently found as an autosomal recessive or X-linked disorder.[5] The present case report of 19 year young boy with manifesting a new syndrome characterized by GF associated with dental abnormalities (DA), including generalized thin hypoplastic AI, intrapulpal calcifications, delay of tooth eruption, pericoronal radiolucencies in unerupted teeth, dental agenesis and root dilacerations.

CASE REPORT

19-year-old male patient came to the department with the chief complaint of swollen and excessive display of gums. On taking history patient he noticed excessive gingival overgrowth since birth, who was born at 32 weeks of gestational development via caesarean section. There were no perinatal complications. Personal history revealed he has a brother who is similarly affected with gingival overgrowth. His medical history was unremarkable, and he had never taken any medication associated with gingival overgrowth. He did not present with any mental impairment or hypertrichosis.

An intraoral examination showed a dentition with yellow discoloured teeth with diastemas and thin enamel of normal hardness. The gingiva was enlarged in both arches (Figure 1), and the posterior areas were affected more severely (Figure 2 & 3).

Panoramic radiograph of patient showed retention of several deciduous teeth because of the unerupted teeth with pericoronal radiolucent areas delineated by sclerotic borders (Figure 4). it was impossible to visualize enamel or differentiate it from dentin. Obvious coronal and radicular pulpal calcifications were present in numerous teeth. Most commonly we can see upper and lower canine, premolar, molars had pericoronal radiolucent zones.

TREATMENT

The initial treatment consisted of oral hygiene orientation and plaque control. An improvement in oral hygiene was observed. The conservative surgical treatment consisted of upper and lower anterior quadrant with external bevel gingivectomy along with gingivoplasty done (Figure 5,6,7 & 8), followed by 0.12% chlorhexidine oral rinses twice a day for 2 weeks after surgery is advised. After excision of gingiva, gingival biopsy sent for histopathological examination for pathology department. Patient under periodical observation and regular dental treatment, including dental filling, dental extractions and prosthesis rehabilitation. Scaling and prophylaxis are performed every 2 months. There was no recurrence of the gingival enlargement even after 4 months. Genetic counselling was given to all affected family members and their parents. Information regarding the pattern of gene transmission, possible ways of expression and consequences of phenotypes were emphasized. Patient referred to oral and maxio-facial surgery and orthodontic treatment for further management.

HISTOPATHOLOGIC AND ULTRASTRUCTURAL FEATURES

The tissues removed during the surgical procedures, were fixed in formalin and embedded in paraffin; the sections were subjected to hematoxylin and eosin (H&E) stain. The histologic features of the gingival overgrowth showed a well-structured epithelium with elongated and thin papillae inserted in fibrous connective tissue(Figure 9). Areas with mild chronic inflammatory infiltrates were observed in the subepithelial connective tissue. The connective tissue showed an increased amount of collagen fibre bundles. Numerous psammomatous calcifications were observed in the connective tissue (Figure 10).

DISCUSSION

This case report documented a family affected by an undescribed syndrome characterized by GF and DA with thin generalized hypoplastic AI as the main dental feature. Other variant features include pulpal calcifications, root dilacerations, delay of tooth eruption, and pericoronal radiolucencies in unerupted teeth can be seen.

It is known that isolated GF may result from a single gene mutation, whereas syndromic forms may result from alterations in multiple genes or a single gene dosage effect. GF as an inheritance disorder shows a heterogeneous pattern of transmission. Chromosomal abnormalities reported for syndromes with GF include duplications, deletions, and/or other anomalies of chromosomes, suggesting that syndromes with GF are genetically different from isolated GF, which is traditionally transmitted by an autosomal dominant gene.[6] The clinical findings of the patients described there were consistent with genetic GF. GF can vary from focal sites of gingival enlargement to generalized involvement, and the degree of overgrowth may vary from slight to severe.

The gingival enlargement may result in aesthetic and functional problems for affected individuals. In our case, patient exhibited a generalized gingival overgrowth, and the posterior and anterior region of the maxilla and mandible were severely affected. he had functional discomfort, but they were unhappy with the appearance of their gingiva. When we studied previous reported cases that the condition usually begins at the time of eruption of the permanent dentition, but it can develop with the eruption of the deciduous dentition and rarely is seen at birth.[7]

The histologic features of the gingiva were composed of fibrous connective tissue with collagen fiber bundles, and all samples contained a significant proportion of myofibroblasts as revealed by histochemical staining.[8] We also observed a broad distribution of calcified psammomatous structures associated with odontogenic epithelial remnants in the overgrown gingiva.

Enamel alterations were compatible with generalized thin hypoplastic autosomal-recessive AI. Affected members showed teeth with yellow colour, smooth surface, lack of contact points, no enamel apparent radiographically, and a thin enamel layer with normal structure alternating with rough areas with severe porosity and irregularly shaped empty spaces.[9] AI has been described as part of several syndromes, but there are only few reports in the literature demonstrating the association between AI and gingival overgrowth.

Generalized gingival overgrowth was also described in a patient affected by the syndrome associating AI with nephrocalcinosis.[10] None of our patient had any history of renal impairment.

GF may result in esthetic and functional problems for affected individuals, and the only treatment available is surgical resection of the overgrowth tissue, but recurrence is anticipated. Recently, demonstrated in vitro that interferon gamma significantly inhibited HGF myofibroblastic cell metabolism, as revealed by the decreased synthesis of type I collagen, supporting that local delivery of this cytokine may be useful to prevent gingival overgrowth in affected patients.[11] In our patient he was unhappy with the appearance of his gingiva, so that we undergone surgical treatment of gingivectomy and gingivoplasty procedure. We were in regular follow-up the patient with professional scaling and oral hygiene instructions, and recurrence was not seen since 6 months.

CONCLUSION

These are the rare case report of gingival overgrowth associated with enamel dysplasia, unerupted teeth and intrapulpal calcifications shows similarities to this family. The above-mentioned features are sufficiently unique to characterize a new syndrome associating GF and DA including generalized thin hypoplastic AI. Finally, conclude in this type of case a genetic investigation is essential to clarify the defect behind this syndrome.

Figures

Figure 1

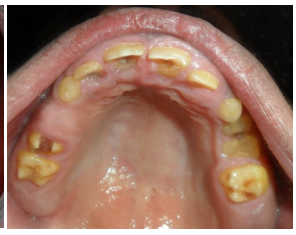

Figure 2

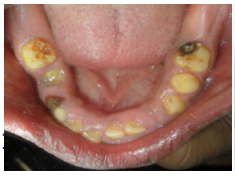

Figure 3

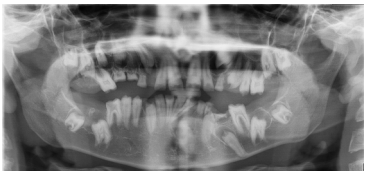

Figure 4

Figure 5

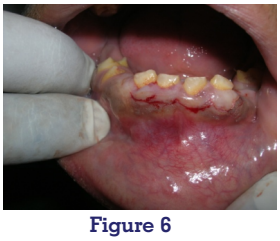

Figure 6

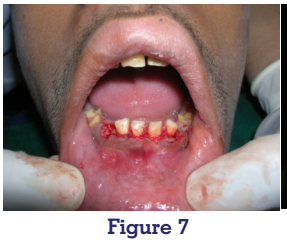

Figure 7

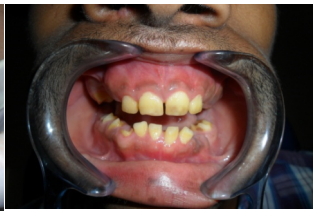

Figure 8

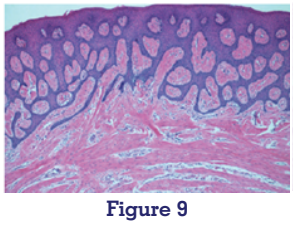

Figure 9

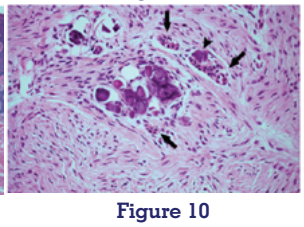

Figure 10

REFERENCES

1. Takagi M, Yamamoto H, Mega H, Hsieh KJ, Shioda S, Enomoto S. Heterogeneity in the gingival fibromatoses. Cancer 1991;68:2202-2212.
2. American Academy of Periodontology. Drug-associated gingival enlargement (informational paper). J Periodontol 2004;75:1424-1431.
3. Gorlin R, Cohen M, Henekam R. Syndromes of the Head and Neck, 4th ed. Oxford Monographs on Medical Genetics. New York: Oxford University Press; 2001: 847-857.
4. Bhowmick SK, Gidvani VK, Rettig KR. Hereditary gingival fibromatosis and growth retardation. Endocr Pract 2001;7:383-387.
5. Aldred MJ, Savarirayan R, Crawford PJ. Amelogenesis imperfecta: A classification and catalogue for the 21st century. Oral Dis 2003;9:19-23.
6. Casavecchia P, Uzel MI, Kantarci A, et al. Hereditary gingival fibromatosis associated with generalized aggressive periodontitis: A case report. J Periodontol 2004;75:770-778.
7. Crawford PJ, Aldred M, Bloch-Zupan A. Amelogenesis imperfecta. Orphanet J Rare Dis 2007;2:17.
8. Farrer-Brown G, Lucas RB, Winstock D. Familial gingival fibromatosis: An unusual pathology. J Oral Pathol 1972;1:76-83.
9. Collins MA, Mauriello SM, Tyndall DA, Wright JT. Dental anomalies associated with amelogenesis imperfecta: A radiographic assessment. Oral

Surg Oral Med Oral Pathol Oral Radiol Endod 1999;88:358-364.
10. Paula LM, Melo NS, Silva Guerra EN, Mestrinho DH, Acevedo AC. Case report of a rare syndrome associating amelogenesis imperfecta and nephrocalcinosis in a consanguineous family. Arch Oral Biol 2005;50: 237-242.
11. Kavvadia K, Pepelassi E, Alexandridis C, Arkadopoulou A, Polyzois G, Tossios K. Gingival fibromatosis and significant tooth eruption delay in an 11-year-old male: A 30-month follow-up. Int J Paediatr Dent 2005;15:294-302.