Case Report

Hereditary Gingival Fibromatosis – A Case Report

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Abstract
Hereditary gingival fibromatosis (HGF) is a rare condition which shows slowly progressive enlargement of gingiva characterized by a generalized enlargement of the buccal and lingual aspects of the attached and marginal gingiva due to accumulation of connective tissue. The condition may occur isolated or as part of a syndrome. Here we present a rare case of unilateral gingival overgrowth of the maxillary and mandibular arches. The diagnosis of the case was based on clinical, radiographic and familial history.

Key words: Gingival fibromatosis, hyperplastic gingival, Gingival enlargement

Introduction
Hereditary gingival fibromatosis (HGF) is a rare condition characterized by progressive enlargement of the gingivae, with a reported incidence of 1 in 7 50,000. Gingival fibromatosis may be familial or idiopathic affecting both the arches. It normally develops as an isolated finding but can be one of the features of several multisystem syndromes. Gingival fibromatosis, also known as Elephantiasis gingivae, Hereditary gingival hyperplasia, Idiopathic fibromatosis, and Hypertrophied gingivae, is a rare condition characterized by slow, progressive enlargement of the gingivae. Clinically, the onset is consistent with the eruption of permanent dentition. At times, it is correlated to the eruption of primary dentition. It rarely presents at birth. Overgrowth can be observed varying in extent and severity. The excess gingival tissue may cover partial or whole crown, resulting in diastemas, teeth displacement, retention of primary teeth, or impacted teeth. The hyperplastic gingiva is usually normal in color, with firm consistency and heavy stippling.

Both autosomal dominant and autosomal recessive modes of inheritance have been reported. The gingival enlargement may occur alone or in combination of other symptoms as part of a syndrome. Examples of syndromic GF are Zimmerman-Laband...
syndrome (GF, hypoplastic distal phalanges, hepatosplenomegaly, epilepsy, hypertrichosis, and mental retardation), Jones syndrome (GF and progressive neural deafness), Klippel-Trenaunay syndrome (GF, hemihypertrophy, Nevus flammeus, hemangioma, hypertelorism, and macrocephaly), Ramon syndrome (GF, unerupted teeth, corneal dystrophy, and mental retardation), and Cross syndrome (GF, nanophthalmos, microcornea, and severe mental retardation).11

Pedigree analyses of HGF families were consistent with simple mendelian transmission pattern, although autosomal recessive cases have been reported in the literature. Recently, Son-of-sevenless (SOS-1) has been identified as the prime etiology for nonsyndromic HGF. SOS-1 is a guanine nucleotide-exchange factor that functions in the transduction of signals that control cell growth and differentiation.12,1

Case Report

An 18-year-old male patient reported to the Department of Oral Medicine and Radiology with a chief complaint of gingival swelling in his mouth. On further questioning, the patient revealed that he had first noticed the swelling after eruption of his permanent teeth. The swelling slowly progressed involving the gingiva mainly in the left upper and lower arches and attained the current size. He complained of bleeding from the gums while brushing, pain in gums while eating hard food. Patient had reported to the hospital due to esthetic concerns. He has not consulted any dentist or physician for his problem previously.

The patient was thoroughly questioned about his physical and mental status to rule out any syndromes associated with the swelling. His medical history was non contributory. His family history revealed significant information that his mother (38 years) and sister (11 years) had similar clinical features of the gingiva. Patient’s family members were requested to report for clinical examination. Similar clinical presentation was noticed in patient’s mother and sister. He also revealed habit of eating Gutkha 1 packet per day since 5 years in his personal history.

On intraoral examination patient revealed nodular, diffuse enlargement of gingiva on both maxillary and mandibular arches with increased involvement of left maxillary and mandibular gingiva compared to right, which were pink in color, firm and fibrous in consistency. The crowns of the teeth on left side of the arches were barely visible because they were almost embedded deep within the enlarged gingiva. There was supragingival plaque, extrinsic and intrinsic stains of teeth, moderate level of calculus and generalized bleeding on probing of gingiva noticed. He had fractured crown in relation to 11 and 21 due to previous trauma. Dental caries with 47 and 16. Grade I mobility with 11. Maxillary and mandibular anterior teeth were displaced from their normal positions.

Correlating the family history and clinical presentations, it was diagnosed as hereditary nonsyndromic gingival fibromatosis.
Figure 1: Frontal view of the patient.

Figure 2: Photograph showing gingival enlargement in maxillary anteriors and completely embedded mandibular anterior within the gingiva.

Figure 3: Photograph showing gingival enlargement in maxillary anteriors and completely embedded mandibular anterior within the gingiva.

Figure 4: Clinical picture showing normal right posterior gingiva.

Figure 5: Photograph showing prominent enlargement of left posterior gingiva.

Figure 6: Clinical picture showing enlarged mandibular left posterior gingiva.

Figure 7: Clinical picture showing enlarged maxillary left posterior gingiva.
Figure 8: Maxillary occlusal radiograph

Figure 9: Mandibular occlusal radiograph

Figure 9: Intra oral periapical radiographs

Figure 10: Lateral cephalogram

Figure 11: Clinical pictures of 38-year-old mother showing similar gingival features who presented diffuse involvement of right and left maxillary and mandibular gingivae.
Discussion
Gingival enlargement, either localized or generalized might be attributed to a number of reasons, ranging from inflammation, leukemic infiltration, and association with use of medicines like phenytoin, cyclosporine, and nifedipine etc. HGF occurs due to congenital or hereditary causes. Studies reveal an autosomal dominant inheritance with chromosomal abnormality in 2p21-p22 and 5q13-q22. It could arise due to nutritional and hormonal factors. In our present case of HGF which may be transmitted as either autosomal dominant or recessive. We justify our diagnosis of the gingival fibromatosis as hereditary and as an autosomal dominant, as this condition is seen in both his mother and sister. Our case showed occurrence of Hereditary Gingival Fibromatosis as an isolated condition but it can also occur in association with various syndromes. A thorough examination of the patient, revealed no association with any of the clinical features associated with the syndromes. Hematological examination reveals all parameter were within normal limits. On the basis of medical, familial, drug histories, radiological examination and the clinical findings, our case was diagnosed as hereditary gingival fibromatosis. Gingivectomy with open flap debridement was advised for enlarged quadrants to restore esthetic, functional and masticatory needs of the patient.

Conclusion
Hereditary gingival fibromatosis is a rare, highly recurrent condition which should be managed as early as possible to avoid difficulty in proper oral hygiene maintenance leading to further inflammation which hinders normal occlusion, mastication, speech and esthetics. Gingival enlargement can recur even after successful treatment hence, regular and thorough follow-up is important in patients showing this rare condition.

References
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