HEREDITARY GINGIVAL FIBROMATOSIS: A CASE REPORT

ABSTRACT

Hereditary gingival fibromatosis (HGF) is characterized by the slowly progressive fibrous enlargement of gingival tissue that causes aesthetic and functional problems. It is usually transmitted as an autosomal dominant trait and develops as an isolated disorder but can also be one of the features of various syndromes. The present case report describes a Turkish family with individuals having HGF for the last three generations. Treatment consisted of surgical removal of the hyperplastic fibrous tissue in a series of gingivectomies for a 54-year-old male patient who had generalized severe gingival overgrowth covering almost all maxillary and mandibular teeth.

Key words: Gingival Enlargement, Gingivectomy, Hereditary Gingival Fibromatosis

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INTRODUCTION

Hereditary gingival fibromatosis (HGF) is characterized by a slowly progressive, non-hemorrhagic, fibrous benign enlargement of maxillary and mandibular keratinized gingiva, the main feature is mature collagenous connective tissue growing. The hyperplastic gingiva may be localized or generalized, usually having normal color and firm consistency with abundant stippling, either unilateral or bilateral. Histologically, gingival tissue from HGF is characterized by a mucosa in which the epithelium shows rete ridges extending into the underlying connective tissue, which shows an increased density of collagen fibrils and a decrease in fibroblasts.

HGF may occur as an isolated finding or in association with other systemic features as part of a syndrome such as Zimmerman-Laband, Jones, Ramon, and Rutherford syndromes, Juvenile hyaline fibromatosis, systemic infantile hyalinosis, and mannosidosis. The condition is usually identified as an autosomal dominant condition though recessive forms are also described in the literature. Linkage studies have localized for isolated, nonsyndromic autosomal dominant forms of gingival fibromatosis to chromosome 2p21-p22 and to chromosome 5q13-q22.

The enlargement usually begins at the time of the eruption of the permanent dentition, although it may also occur during the eruption of deciduous teeth or even at birth. The enlarged gingivae may cover a portion or all the crowns of teeth, thus potentially interfering with speech, closure of the lips, and mastication. It may delay eruption of teeth and make cleaning of the teeth virtually impossible. When the gingiva is the only tissue involved, gingival fibromatosis might be in association with hypertrichosis, and/or mental retardation, and/or epilepsy.

This case report presents HGF in a Turkish family for three-generation.

CASE REPORT

A 54-year-old male presented with a complaint of excessive gingival hyperplasia involving both the mandibular and maxillary arches. On examination, a severe and generalized diffuse gingival hyperplasia involving the marginal, interdental and attached gingiva was observed covering almost all the surfaces of crowns causing severe eating, speaking and aesthetic problems (Figure 1). Gingiva was pink with a firm and dense consistency. Poor Oral hygiene was observed in response to tissue overgrowth. Radiographic evaluation demonstrated severe alveolar bone loss (Figure 2). The patient’s medical history has revealed no signs of hypertrichosis or mental retardation, and no history of epilepsy or intake of medication known to cause gingival overgrowth. He referred to our clinic to have a better esthetic appearance. The patient stated that he had such a psychological and emotional stress until he reaches to this age with a fair of surgery. There was a family history of hereditary gingival overgrowth as his mother and son had gingival enlargement, involving the maxilla as well as the mandible. The patient revealed that his mother and son had several gingival excisional surgeries because of slowly progressing hyperplasia. None of the individuals in the family had epilepsy or any type of seizure disorder, and had not taken any medications associated with gingival hyperplasia. The mother’s age is 79 and was operated three times about 40 years ago. The son stated that he had several gingival excisional surgeries 12 years ago. The recurrence was seen after the surgeries in both of them, the son and mother (Figures 3 and 4).

After giving detailed information to the patient about the management and taking an informed consent, the patient decided to initiate the treatment. Multiple surgical procedures including maxillary and mandibular vestibular and palatal/lingual gingivectomies, with reverse bevel incisions, were performed under local anesthesia, to obtain...
a smoother gingival contour (Figure 5). A biopsy sample of the gingival tissues was submitted for histologic evaluation. Microscopic examination of the specimens confirmed for hereditary gingival hyperplasia. Sections showed a hyperparakeratinized hyperplastic stratified squamous epithelium with the underlying fibrous connective tissue showing bundles of collagen fibers. The patient was diagnosed with gingival fibromatosis based on the microscopic findings.

A 0.12% chlorhexidine gluconate rinse was prescribed for administration twice a day for 2 weeks. The patient was seen at 1, 3 and 4 weeks postoperatively. There were no postoperative complications. After 30 months of follow-up, poor oral hygiene was observed (Figure 6).

To achieve optimal esthetic appearance, we recommended orthodontic and prosthetic treatment but the patient refused these treatments. Also patient’s son refused any treatment because of overgrowth of the gingival tissue following the operations.

**DISCUSSION**

Hereditary gingival fibromatosis is rare, affecting only one in 750,000 people.\textsuperscript{16} It usually develops as an isolated disorder but can be one feature of a syndrome.\textsuperscript{17} In this case, the patient did not exhibit any signs or symptoms suggesting that the condition was syndromic.

HGF can be inherited as an autosomal dominant though recessive forms are described in the literature.\textsuperscript{13} The mode of genetic transmission in our case, points to an autosomal dominant gene, because family members (the patient’s mother and son) of both sexes were affected.

Reports of the histologic, morphologic, and cellular characteristics of gingival tissues from HGF patients compared with controls are not always consistent, although an increase in the amount of collagen is generally reported.\textsuperscript{18} In recent years, studies aimed to clarify the pathogenic mechanisms of collagen accumulation in HGF gingivae but there has been controversy about this mechanism: gingival fibrosis is a result of increased collagen synthesis and/or decreased degradation, or alteration in fibroblast proliferation.\textsuperscript{19-21}

The proliferation and expression of growth factors of HGF keratinocytes are abnormal. However, the exact role of keratinocytes in HGF pathogenesis is still unknown. According to Meng et al.\textsuperscript{22} HGF keratinocytes have an important role in HGF pathogenesis by inducing extracellular matrix (ECM) accumulation by fibroblasts. These authors reported that results strongly suggest that keratinocyte–fibroblast interactions contribute to the pathogenesis of HGF.

The hyperplastic gingiva has usually got a normal color and firm consistency.\textsuperscript{2} The clinical presentation of HGF is
variable in both the distribution (number of teeth involved) and in the degree (severity) of expression. Buccal and lingual tissue may be involved in both the mandible and maxilla, and the severity of hyperplasia may range from slight enlargement to total coverage of the dentition and may vary between individuals within the same family.\(^{11}\) In our case, the hyperplastic gingiva was of normal color and had a firm consistency. The patient was not able to chew especially at the anterior region, as the gingiva was almost covering the maxillary teeth.

Gingival tissue enlargement usually begins with the eruption of the permanent dentition. There are also cases which have been reported to occur during the eruption of the deciduous dentition; however, it rarely appears at birth.\(^{16}\) Since recurrence could be expected within a few months after surgery and may return to the original condition within a few years, the patient may undergo to repeated gingivectomy procedures. Reports about recurrence rates are conflicting,\(^{23-25}\) so the long-term benefit of periodontal reduction surgery cannot be predicted. In severe cases of hereditary gingival fibromatosis, full-mouth tooth clearance has been advocated, as some evidence\(^{24,25}\) suggests that the condition does not recur if the teeth have been extracted.

It was stated that there is less chance of recurrence if the gingivectomy is delayed until the permanent dentition is in place.\(^{26}\) However, one report recorded that deciduous teeth were retained and permanent teeth were entrapped in the overgrown gingival tissue. Thus, delaying the gingivectomy with the goal of avoiding recurrence was contraindicated. These authors have also stated that as long as patient is informed about the likelihood of recurrence, repeated gingivectomies is generally well accepted and well tolerated. On the other hand, Shekar\(^{27}\) reported that repeated gingivectomy procedures often causes further increase in the patients and parents' psychological and emotional stress.

Kelekis-Cholakis et al.\(^{28}\) stated that orthodontic treatment might stimulate recurrence in some patients, especially if periodontal hygiene is impeded by the orthodontic appliance but they did not observe significant recurrence during the first 3 years.

Poor oral hygiene is one of the factors that may aggravate gingival enlargement as it is seen in the members of this family. Therefore, the importance of postoperative follow-up and adequate hygiene should be emphasized to HGF patients.\(^{29,30}\)

**CONCLUSION**

Hereditary gingival fibromatosis is a rare disease characterized by the proliferative fibrous overgrowth of the gingival tissue. Enlargements in the gingiva may cover the dental crowns resulting in both aesthetic and functional problems. Multiple surgical procedures, monthly periodontal examinations and prophylaxis are an essential part of the treatment protocol. However, additional effort by the patient is needed to cope with HGF.

**REFERENCES**


