

Hereditary Gingival Fibromatosis and its management: A Rare Case of Homozygous Twins

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ABSTRACT

Hereditary gingival fibromatosis (HGF) is a rare condition which manifests itself by gingival overgrowth covering teeth to variable degree i.e. either isolated or as part of a syndrome. This paper presented two cases of generalized and severe HGF in siblings without any systemic illness. HGF was confirmed based on family history, clinical and histological examination. Management of both the cases was done conservatively. Quadrant wise gingivectomy using ledge and wedge method was adopted and followed for 12 months. The surgical procedure yielded functionally and esthetically satisfying results with no recurrence.

KEYWORDS: Gingival enlargement, Hereditary, homozygous, Gingivectomy

INTRODUCTION

Hereditary Gingival Enlargement, being a rare entity, is also known as familial elephantiasis, elephantiasis gingivae, diffuse fibroma.¹ It may initiate/aggravate soon after eruption of permanent teeth.² It may occur as an isolated entity or may be associated with various syndromes such as Zimmer–man Laband Syndrome, Ramon Syndrome, Rutherford Syndrome, Cross syndrome, Jones syndrome.³ Clinically, it present as pale pink color gingival enlargement, firm in consistency, which may be localized or generalized. A familial history may be reported in few cases. Histological features consist of highly fibrous connective tissue with numerous fibroblasts. Connective tissue is avascular with thickened acanthotic hyperkeratotic stratified squamous epithelium.⁴ Gingival enlargement may pose consequences such as esthetic problems, delayed eruption of permanent teeth, retained primary teeth, speech problems, and malocclusion.² The management of gingival fibromatosis include gingivectomy using conventional technique (scalpel), laser, electrocautery followed by gingivoplasty.⁵

Here we report a case of nonsyndromic hereditary gingival fibromatosis in homozygous twins which was treated using quadrant wise conventional surgical technique

CASE REPORT

Case Report 1: A 10-year-old female child with her sister was brought by her mother with the chief complaint of

swollen gums. The patient gave a history of swelling of upper gums that started 2 years back which gradually increased in size. The child's mother denied prenatal exposure to tobacco, alcohol, and drug. She also reported no exposure to radiation and no illness during her pregnancy. Both the daughters were born via caesarean delivery at 38 weeks of pregnancy and were homozygous twins (Fig 1). Post-natal history revealed the history of loose stools at 8 months of age to both sisters for which they took unknown medication in the form of syrup for 2 years. There was no contributing paternal/maternal history with the similar complaint. The patient was referred for genetic and other systemic disease evaluation. The chromosomal analysis revealed no anomaly, hence ruled out the suspicion of associated systemic disease. Extraoral examination revealed convex profile, frontal bossing, broad and depressed nasal bridge, prominent premaxilla and potentially incompetent lips. (Fig 2)

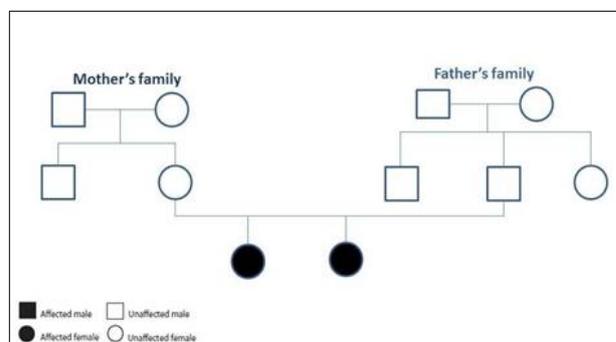


Fig 1- Pedigree analysis of the family

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Fig 2- Extraoral photos (Case 1)

Intraoral examination revealed a generalized diffuse enlargement of gingiva affecting the labial/buccal & palatal/lingual gingiva (attached, marginal & interdental gingiva) involving both the arches covering almost two thirds to three-quarters of all teeth (Grade 3- as per Bokenkamp 1994)⁶. Stippling was preserved. Gingiva was pink, and firm, dense, fibrous in consistency that caused difficulty in tooth eruption. However, there were no signs of bleeding or suppuration. (Fig. 3) Panoramic radiograph did not reveal any bone lesion. No bone loss around teeth was evident. (Fig. 4)



Fig 3- Pre operative intra oral photos showed gingival enlargement in both the arches

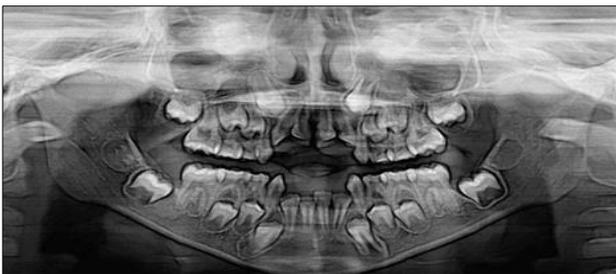


Fig 4: Orthopantomogram shows no sign of bone loss associated

On the basis of medical, family and drug history and clinical examination, a provisional diagnosis of Hereditary Gingival Fibromatosis was made

Case Report 2: A 10-year-old female, sibling of the same patient reported with the chief complaint of swollen gums since 2 years. The family history of gingival enlargement was present since it was present in both the siblings. Post natal history didn't reveal the history of systemic illness or medication for same. The genetic evaluation didn't exhibit the presence of any systemic condition, which may be associated with gingival enlargement. Extra oral and Intra oral features were same (Fig. 5) as the first patient, suggesting a provisional diagnosis of Hereditary Gingival Fibromatosis.



Fig 5- Extraoral photos (Case 2)

Management: Quadrant wise gingivectomy was planned, and after explaining to the parents/patients, due consent was taken before carrying out the treatment. Pre-surgical mouth rinse with 0.2% chlorhexidine was done before every appointment for both the cases. External bevel gingivectomy was performed using ledge and wedge procedure using scalpel method under local anesthesia (2 % lignocaine with 1:100,000 Adrenaline), and the periodontal dressing was given post-surgery. (Fig 6) Over



Fig 6: Post surgical procedure photograph after gingivectomy using conventional surgical method

retained teeth and were also extracted during the procedure. After each dressing antibiotics (Amoxicillin 250 mg three times a day) and Analgesics were prescribed to the patients. The periodontal pack was removed after seven days, and postoperative instructions were given to the patients. The patient was recalled after seven days and the postoperative period was uneventful. Oral hygiene instructions were emphasized for patient and parents. The interval of 3-4 weeks was given between successive surgeries. The tissue obtained was sent for histological examination. Histopathologic examination revealed abundant dense connective tissue in which markedly thickened collagen fibers, and numerous fibroblasts were present. The surface epithelium displayed hyperplasia of stratified squamous epithelium. (Fig 7)

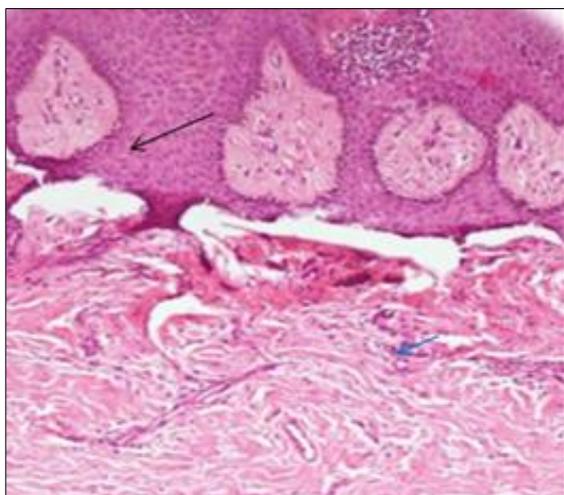


Fig 7: Histopathological slide showing hyperplastic epithelium (black arrow) with fibro collagenous tissue (Blue arrow)

The post surgical follow-up after 6 months did not show any recurrence (Fig 8). After 12 months of follow up, when no recurrence was observed, both the patients were given anti-tongue-thrust correction appliance (cribs) for anterior open bite correction and kept under observation. (Fig 9)



Fig 8: Follow up after (6 months) showing no sign of recurrence



Fig 9: 12 months follow up with anti tongue thrusting cribs' with no recurrence

DISCUSSION

The case report presents a symmetric case of Grade 3 gingival enlargement of two young siblings suggesting autosomal recessive penetrance. Although the etiology is still not understood, the majority of researchers have attributed this condition to hereditary factors. The mode of genetic transmission in patients with HGF is both autosomal dominant and autosomal recessive. Recently four genetic separated loci on chromosome 2p, Chromosome 5q, and chromosome 11p have been determined in association with HGF.⁷ Gingival enlargement may be localized or generalized. In the present cases, gingival enlargement was generalized (Grade 3) involving both the arches. In the present cases, the diagnosis of HGF was made based on family history and clinical examination. Clinical examination revealed that the attached and marginal gingiva was pink and firm in consistency which helped it in differentiating it from drug induced gingival enlargement. Moreover, the absence of inflammation in the present cases ruled out the diagnosis of inflammatory gingival enlargement.

HGF may occur as an isolated phenomenon or in association with other systemic illness. The cases presented with anterior open bite, frontal bossing, broad and depressed nasal bridge, and potentially incompetent lips. HGF usually occurs in association with hypertrichosis (Cowden Syndrome) and mental retardation (Cross Syndrome). The other rare associations such as multiple hyaline fibromas, osteolysis of terminal phalanges and recurrent infections (Murray-Puretic-Drescher syndrome), corneal opacities and retarded tooth eruption (Rutherford syndrome), ear, nose, bone and nail defects with hepatosplenomegaly (Zimmermann- Laband syndrome), progressive deafness (Jones syndrome), microphthalmia, mental retardation, athetosis and hypopigmentation (Cross syndrome) and giant fibroadenoma of breast, hypertrichosis and multiple hamartomas (Cowden syndrome) were not observed in either of two cases.³ The present case reports did not show any of the above-mentioned features that rule out these systemic conditions in association with HGF.

The histological findings are nonspecific with hyperkeratosis, acanthotic epithelium with elongated papillae. The abundance of fibroblasts is present in

connective tissue. This increased production of extracellular matrix molecules, type I collagen and fibronectin could lead to bulk of gingiva clinically.⁴ Histopathological examination in the present case report were similar to HGF.

Gingival enlargement can cause speech alteration, diastema in teeth, cessation of eruption of permanent teeth leading to difficulty in mastication, malocclusion, and unaesthetic appearance.² In the present cases, gingival enlargement has led to non-eruption of permanent teeth, malocclusion and unaesthetic appearance. Management of gingival enlargement depends on the age of occurrence and cause of this condition. Gingival overgrowth usually begins with the eruption of permanent teeth which progresses rapidly with the stage of active eruption. In order to address patients' functional and esthetic needs, surgical excision of the enlarged gingiva is usually necessary. Other methods of removing large quantities of gingival tissue have been used in a number of studies, such as carbon dioxide laser and electrocautery.⁵ In the present case, gingivectomy using scalpel method using wedge procedure was adopted. Conventional surgical excision has advantages over other modes that bulk of tissue can be removed in single visit, technique is not sensitive and faster healing rates have been observed.⁶

There is no definite consensus regarding the time of surgical intervention in young and adolescent patients. Emerson suggested to wait till all the permanent teeth have erupted i.e. at the ages of 3, 6 and 12 years in order to maintain oral hygiene effectively after gingivectomy.⁸

The recurrence rate of HGF is very high. Hence patient should be followed for a considerable period of time and may require repeated surgeries. Cuestas-Carneiro and Fletcher recommended excision of the excess tissue combined with removal of all teeth in severe chronic cases, because it appears that permanent cure is most likely to be obtained.^{9,10} In the present cases, we trended toward a more conservative approach, and no recurrence was seen in both the cases.

CONCLUSION

HGF is a rare entity, conferring to pedodontist and

periodontist an important role in correct diagnosis and treatment of affected patients. Esthetic and dental associated alterations can affect the quality of life of affected children. Hence quadrant wise gingivectomy using scalpel method was used along with proper oral hygiene instructions would be a better treatment option in these cases. Follow up after 12 months revealed satisfying results with no recurrence.

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