Case Report

International Journal of Dental and Health Sciences
Volume 02, Issue 03

HEREDITARY GINGIVAL FIBROMATOSIS ASSOCIATED WITH CONSANGUINITY: A RARE CASE REPORT

Anila Sankaranarayanan[1*],Nandakumar Krishnankutty[2],Prasanth Dhanapal[3], George Jacob M[4]

[1]Reader, Department of Periodontics, P.S.M. College of Dental Science and Research, Thrissur, India
[2]Principal, Azeezia Dental College, Kollam, India
[3]Professor, Dept. Of Conservative Dentistry & Endodontics, Annoor Dental College & Hospital, Muvattupuzha, India
[4]Reader, Dept of Conservative Dentistry & Endodontics, P.S.M. College of Dental Science and Research, Thrissur, India

ABSTRACT:

Hereditary gingival fibromatosis is a rare condition manifested by enlarged gingival tissues, and can occur as an isolated disease or as part of a syndrome. The condition is inherited as an autosomal dominant trait, which is more common, or as an autosomal recessive trait. This case report reports a case of a 16 year old boy with non-syndromic Hereditary gingival fibromatosis with a probable recessive mode of inheritance, as the patient's parents reported a history of consanguineous marriage. The fibrotic gingival tissue was removed surgically, sextant wise under local anesthesia. After the surgical procedure the patient's appearance was considerably improved. The case was followed up for 18 months at regular intervals and there was no evidence of recurrence of the gingival condition.

Key words: Consanguinity, Fibromatosis, Gingival, Hereditary

INTRODUCTION:

Hereditary gingival fibromatosis (HGF) is a rare inherited condition, which results in spontaneous and progressive enlargement of the gingiva [1]. Even though traditionally considered an autosomal dominant disease, it does manifest less commonly as an autosomal recessive condition [2]. It can occur as an isolated trait or as part of a syndrome [3]

The hyperplastic gingiva usually displays a normal pink colour and has a firm consistency with presence of exaggerated stippling [4]. The usually painless enlargement may extend up to the mucogingival junction, but does not involve the alveolar mucosa [5]. The excessive gingiva can cause aesthetic problems like diastema, lip prominence, phonetic problems and problems related to mastication, in addition to psychological disturbance [6]. The enlargement can increase plaque retention thus inducing periodontitis and halitosis [7].

*Corresponding Author Address: Dr.Anila.S,. Email: anila_sp@rediffmail.com

Females and males are equally affected and the phenotype and genotype frequency is 1:175,000 and 350,000 respectively [8]. A mutation in the SOS-1 or son-of-sevenless gene on chromosome 2p21-p22 has been linked to HGF [2]. .Autosomal-dominant forms of gingival fibromatosis have been genetically linked to the chromosomes 2p21-p22 [9] and 5q13-q22 [10, 11].

Syndromes associated with HGF with autosomal recessive inheritance include Cross syndrome, Ramon syndrome, Zimmerman-Laband syndrome,[12] and Murray-Puretic-Drescher syndrome [7]. In the recessive form with no other associated defects or syndromes, consanguinity is always present in the family [13].

Union of individuals who are related to each other as close as or closer than second cousins is the arbitrary definition of consanguinity. A patient with a genetic disorder, whose parents had a consanguineous marriage, indicates a strong evidence for that genetic disorder to have an autosomal recessive pattern of inheritance [14].

Here we are reporting the clinical, histopathological features and dental management of a 16 year old boy with HGF with a probable recessive mode of inheritance.

CASE DETAIL:

A 16 year old male reported with complaints of gingival swelling since 3 years. The patient chief complaint was with regard to poor aesthetics. Patient gave a history of delayed eruption of deciduous teeth and gingival enlargement was also reported during deciduous dentition.

The patient's medical and surgical history was non-contributory. The patient did not have hypertrichosis or mental retardation and had no history of intake of medications known to cause gingival enlargement. Family history revealed consanguineous marriage of his parents. The patient had a sibling without this similar gingival manifestation.

Extraoral examination revealed a convex profile with incompetent lips. Intaoral examination revealed generalized diffuse gingival enlargement symmetrically affecting both maxillary and mandibular arches, on both buccal and palatal/lingual surfaces (Fig.1). The enlargement involved marginal, attached and interdental gingiva in both the arches, covering almost the entire clinical crown except the incisal/occlusal areas. The enlargement was most severe in lower lingual region (Fig.2). Gingiva was pale pink in colour with pigmentations present within attached gingiva, it had a firm fibrous consistency with presence of stippling and was not tender to palpation. There was no mobility of any teeth.

An orthopantomogram revealed normal development of permanent teeth with missing lower right first molar. There was no disharmony between the dental and skeletal ages and no gross bony deformity. Routine blood and urine investigation showed all parameters within normal limits. Since the patient's

parents were second cousins, a pedigree analysis was done, which indicated a strong evidence for an autosomal recessive mode of inheritance.

Full mouth gingivectomy under local anaesthesia was planned and the treatment plan explained to the patient and parent. Gingivectomy was done sextant wise under local anaesthesia in weekly intervals. An initial internal bevel incision was given, followed by a crevicular incision (Fig.3). After the tissue wedge was removed (Fig.4), the inner surface of the gingival flap was trimmed to remove excess tissue, and the flap was sutured back.

Patient was prescribed chlorhexidine 2% oral rinse (for 1 week), antibiotics and analgesics for 5 days after the surgical procedure in each sextant. There was uneventful healing post-surgically (Fig.5 & 6). Suture removal was done after 1 week and plaque control instructions were reinforced. Post-operatively patient's esthetics and masticatory function was considerably improved.

Histopathologic examination showed pseudoepitheliomatous hyperplasia of parakeratinised stratified squamous epithelium with elongated rete pegs (Fig. 7). Densely collagenous, relatively avasular connective tissue was noted with numerous fibroblasts and chronic inflammatory cells (Fig. 8). An overall impression of fibroepithelial hyperplasia was reported. (10X magnification)

The patient was put on a follow-up program and was reviewed at 1, 2, 3, 6, 12 and 18 months intervals. Clinically there was no recurrence of the condition after 18 months follow-up.

DISCUSSION:

The etiologic agents implicated in generalized gingival fibromatosis include inflammation, use of medications such as phenytoin, nifedipine or cyclosporine [15], Wegener granulomatosis, acanthosis nigricans and leukemic infiltration. Hereditary gingival fibromatosis generally manifests as dominant form and recessive forms are usually associated with certain syndromes like Ramon syndrome (Cherubism, hypertrichosis, mental deficiency, epilepsy and stunted growth), Cross syndrome (microphthalmia, athetosis, mental retardation and hypopigmentation) , Murray-Puretic-Drescher syndrome (multiple hyaline fibromas) [7].

In this present case. the patient did not present with any other feature suggesting that the condition was part of any syndrome. The pedigree analysis gives strong evidence that the condition is most likely to have an autosomal recessive mode of inheritance.

The esthetic and functional problems associated with gingival enlargement include delayed eruption, malpositioning of teeth, cross/open bites, prominent lips and incompetent lips. Extensive enlargement can result in decreased tongue space, difficulty in phonetics and mastication. The enlargement can also result in increased plaque accumulation resulting in periodontitis. The patient

presented in this case report had incompetent lips and was unhappy regarding gingival esthetics.

HGF usually presents with normal colour of gingiva with stippling and the clinical manifestation in the present case is consistent with this description. It usually becomes evident at the time of eruption of permanent teeth. Presence of teeth appears to be necessary for this condition to occur as HGF is not seen before teeth eruption and recedes after removal of teeth [13]. However in rare cases, it has been reported at birth. HGF progresses rapidly during active eruption and decreases with the end if this stage [1].

HGF is benign histologically, with accumulation of mature collagenous bundles running in various directions in a relatively avascular connective tissue [16]. Epithelial hyperplasia with long thin papillae extending deep into the connective tissue is also seen [17]. The histopathologic findings in this patient is consistent with previously reported cases of HGF.

Treatment of HGF varies with the degree of severity of enlargement. If the enlargement is minimal, a thorough oral prophylaxis with good oral hygiene maintenance could be adequate for maintaining good oral health. Excessive tissue associated with esthetic, phonetic or mastication related problems indicates a necessity for surgical intervention.

Suggested surgical modality for gingival fibromatosis is gingivectomy with either external or internal bevel incisions. Electrocautery, apically positioned flap and carbon dioxide laser have been tried for the excision of the enlarged gingival tissue [18]. We have used an internal bevel incision because the extensive gingival enlargement would have resulted in exposure of a wide area of connective tissue if external bevel incisions were used.

Recurrence of HGF has been reported by some authors, especially in children and teenagers. In severe cases, full mouth extraction has been advocated, as it has been suggested that the condition does not recur after tooth extraction [13]. Correct physiologic contour of free gingiva and good oral hygiene maintenance are important to prevent recurrence. Even if the cosmetic improvement is temporary, the psychological benefits should not be underestimated and may outweigh the probability of recurrence.

CONCLUSION:

Gingival Fibromatosis can be inherited condition in the progeny of consanguineous marriage. Genetic counselling making couples aware of this possibility could be of importance and is highly recommended.

REFERENCES:

1. Fletcher JP. Gingival abnormalities of genetic origin: preliminary communication with special reference to hereditary gingival fibromatosis. J Dent Res. 1966; 45(3):597-612.
2. Jorgenson RJ, Cocker ME. Variation in the inheritance and expression of gingival fibromatosis. *Journal of Periodontology*. 1974; 45(7):472–477.
3. Haytac MC, Ozcelik O, Turkey A. The phenotypic overlap of syndromes associated with hereditary gingival fibromatosis: Follow-up of a family for five years. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2007; 103:521-7.
4. Ramakrishnan T, Kaur M. Multispeciality approach in the management of patient with hereditary gingival fibromatosis: 1-year followup: A case report. Int J Dent. 2010: 575979.
5. Bozzo L, Almeida OP, Scully C, Aldred MJ. Hereditary gingival fibromatosis. Report of an extensive four-generation pedigree. Oral Surg Oral Med Oral Pathol. 1994;78: 452-4.
6. Coletta RD, Graner E. Hereditary gingival fibromatosis: A systematic review. J Periodontol 2006;77:753-64
7. Ricardo D.C, Edgard G. Hereditary gingival fibromatosis: A systematic review. Journal of Periodontology. 2006; 77(5):753-764.
8. Butchi KB, Pavankumar K, Anuradha BR, Arora N. Hereditary gingival fibromatosis—a case report and management using a novel surgical technique. Revista Sul-Brasileira de Odontologia. 2011; 8(4):453–458.
9. Blackwell CC, Weir DM, James VS, Cartwright KA, Stuart JM, Jones DM. The Stonehouse study: Secretor status and carriage of Neisseria species. Epidemiol Infect. 1989;102:1–10
10. Hart TC, Pallos D, Bowden DW, Bolyard J, Pettenati MJ, Cortelli JR. Genetic linkage of hereditary gingival fibromatosis to chromosome 2p21. *American Journal of Human Genetics*. 1998; 62(4):876–883.
11. Xiao S, Bu L, Zhu L, Zheng G, Yang M, Qian M, et al. A new locus for hereditary gingival fibromatosis (GINGF2) maps to 5q13-q22. Genomics. 2001; 74:180–5.
12. Bakaeen G, Scully C. Hereditary gingival fibromatosis in a family of Zimmerman-Laband syndrome. J Oral Pathol Med. 1991;20:457–9
13. Kharbanda OP, Sidhu SS, Panda SK, Deshmuck R. Gingival fibromatosis: study of three generations with consanguinity. Quintessence Int 1993; 24(3):161-4.
14. Robert L N, Roderick R M, Huntington FW. Genetics in Medicine 7th ed. Philadelphia, Saunders, 2007.
15. Ramer M, Marrone J, Stahl B, Burakoff R. Hereditary gingival fibromatosis: Identification, treatment, control. J Am Dent Assoc. 1996; 127:493–5.
16. Sakamoto R, Nitta T, Kamikawa Y, Kono S, Kamikawa Y, Sugihara K, et al. Histochemical, immunohistochemical, and ultrastructural studies of gingival fibromatosis: A case report. Med Electron Microsc. 2002; 35:248–54.
17. Martelli-Júnior H, Bonan PRF, Dos Santos LA, Santos SM, Cavalcanti MG, Coletta RD. Case reports of a new syndrome associating gingival fibromatosis and dental abnormalities in a consanguineous

family. J Periodontol. 2008; 79:1287-96.

18. Pecaro BC, Garehime WJ. The CO 2 laser in oral and maxillofacial surgery. J Oral Maxillofac Surg. 1983;41:725–8

FIGURES:

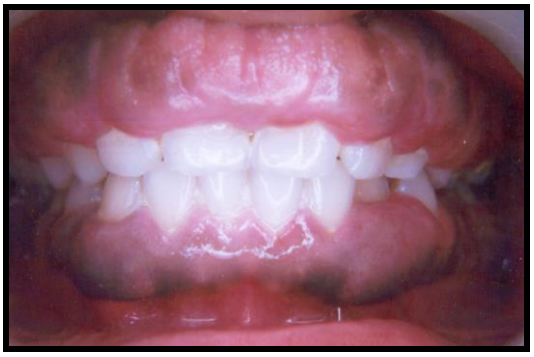

Figure 1: Clinical photograph at initial visit. Frontal view

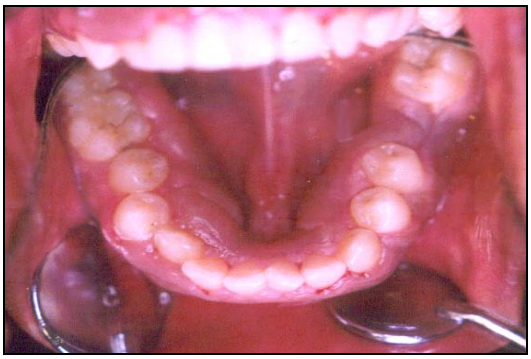

Figure 2: Clinical photograph at initial visit. Mandibular occlusal view

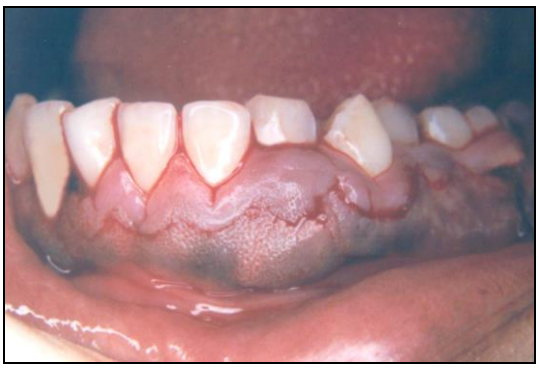

Figure 3: Clinical photograph during gingivectomy

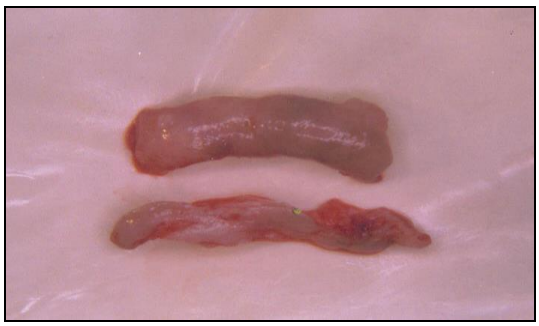

Figure 4: Excised tissue

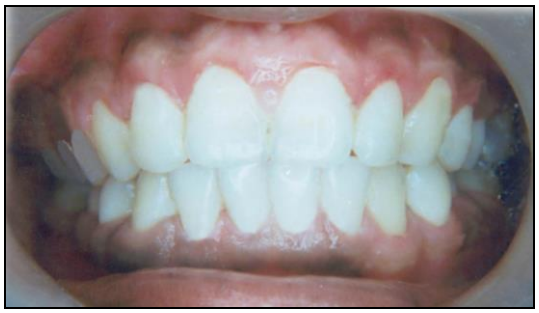

Figure 5: Post-operative photographs. Frontal view

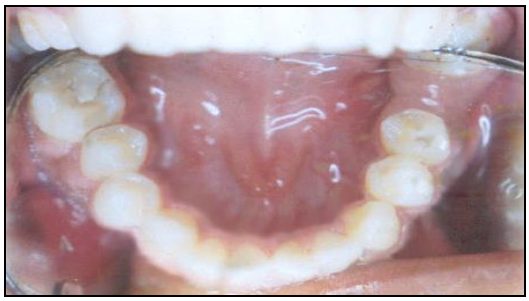

Figure 6: Post-operative photographs. Mandibular occlusal view

Figure 7: Photomicrograph showing pseudoepitheliomatous hyperplasia of the epithelium (H&E, x10).

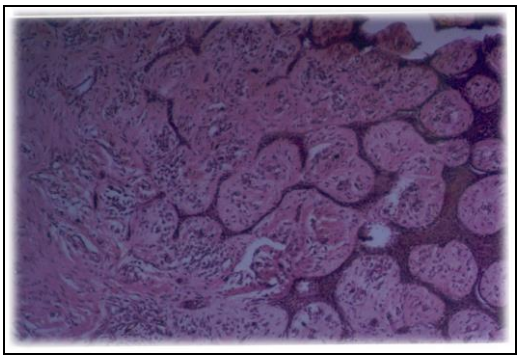

Figure 8: Photomicrograph showing densely collagenous, relatively avascular connective tissue (H&E, x10).