Case Report

Hereditary gingival fibromatosis

Akhilanand Chaurasia

Department of Oral Medicine and Radiology, Faculty of Dental Sciences, King George Medical University, Lucknow, Uttar Pradesh, India

Address for correspondence:
Dr. Akhilanand Chaurasia,
C/O Department of Oral Medicine and Radiology, Faculty of Dental Sciences, King George Medical University, Lucknow - 226003, Uttar Pradesh, India.
E-mail: chaurasiaakhilanand49@gmail.com

ABSTRACT

Hereditary gingival fibromatosis (HGF) is a rare condition that can occur as an isolated disease, chromosomal abnormality, or as part of a number of syndromes. The gingival enlargement in HGF can be so severe that it can cover the crowns of teeth completely, causing severe functional derangement and facial disfigurement. Through review of literature revealed that HGF is usually an autosomal dominant condition, however the recessive forms are also reported. The hyperplastic gingiva is firm on palpation and has normal color with abundant stippling on the adjacent gingiva. The buccal and lingual gingiva may be involved in both the mandible and maxilla. The degrees of gingival enlargement show both intra- and interindividual variations. Here, an interesting case report of massive HGF has been presented.

Key words: Gingival enlargement, gingival fibromatosis, gingivectomy

Introduction

The hereditary or idiopathic gingival fibromatosis is a rare condition of the gingival tissues characterized by enlargement of the free and attached gingivae. The hereditary gingival enlargement is also known as gingivomatosis,[1] diffuse fibroma,[2] idiopathic fibromatosis,[3] hereditary gingival fibromatosis (HGF),[3] and familial elephantiasis.[3] The HGF is a rare disease affecting only 1 in 750,000 people.[4] Usually it is an isolated disorder, but it may be associated with epilepsy, hypertrichosis, and mental retardation[5,6] or it can develop as a part of syndromes like Cowden's syndrome,[7] Zimmerman-Laband syndrome,[8] and Murray-Puretic syndrome.[7]

The HGF has a variable clinical presentation. In some cases it shows only minimal involvement characterized by enlargement of the tuberosity area and buccal gingiva around the mandibular molars; however, in severe form it can involve both maxillary and mandibular gingiva. The enlargement in severe form can be so massive that it covers crown of both primary or permanent teeth completely. The HGF appears as generalized nodular hyperplastic gingiva which is firm to bony hard in consistency. Usually the hyperplastic gingiva is pale pink to red and inflamed. It has characteristic smooth to pebbled surface with little tendency to bleed. The enlargement is usually painless and may extend up to the mucogingival junction but does not affect the alveolar mucosa[9,10] or alveolar bone. Excessive gingival growth results in pseudo pocketing and periodontal problems due to difficulties in daily oral hygiene. The HGF may also result in diastemas, delayed tooth eruption, and facial disfigurement due to lip protrusion. Severe hperplasia can result in restricted movements of the tongue, speech impediments, difficulty with mastication, and can prevent normal closure of lips.[11]

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Case Report

A 10-year-old girl [Figure 1] reported to outpatient department of Department of Oral Medicine and Radiology, Faculty of Dental Sciences, King George Medical University with presenting complaint of growth involving upper jaw and lower jaw, difficulty in eating food and speech impairment, and noneruption of teeth. The patient noted the growth 3 years back, since then it was progressively increasing in size causing inadequate lip apposition and poor esthetic. The family and postnatal history was noncontributory. She had no history of epilepsy or mental disorder. Her past medical and dental history was not significant. Further history revealed that neither of her family members was affected with similar condition. There was no history of consanguinity. She was malnourished and belonged to a

very poor family. Routine medical history and physical examination revealed no systemic disease.

Extraoral examination [Figure 2] showed a massive, generalized multinodular growth covering both maxillary and mandibular arch completely causing incompetent everted lips and a convex profile. The surface of maxillary growth was granular, mildly pigmented, and had multiple fissuration extending between corners of mouth and covering all maxillary anterior teeth completely; while mandibular growth was larger and extended beyond submental region of mandible giving an appearance of hanging mass attached to mandible. The growth was multilobular, pebbled, and had smooth surface extending between corners of mouth. The mandibular anterior teeth were not visible. The growth was pink to pinkish white in color. On palpation, it was found to be firm and nontender in consistency. There were no fluctuancy, no compressibility, no translucency, and no associated bleeding noted. There was mild pain associated with growth.

Intraoral examination of maxilla [Figure 3] revealed a gross generalized growth of gingiva completely covering the maxillary teeth extending from right maxillary tuberosity to left tuberosity region. The mass had covered the anterior palatine region completely, while posteriorly it was extending bilaterally towards midline of palate without fusion. The mass in anterior palatine region had granular, mildly pigmented surface having pinkish white color. The surface had multiple fissurations. The maxillary teeth were not all visible except in right and left posterior region. There was no associated bleeding or pain noted with growth. On palpation, the mass was found to be firm and nontender.

However, intraoral examination of mandible [Figure 4] revealed a gross generalized growth of gingiva completely covering the mandibular teeth extending from right mandibular tuberosity to left tuberosity region. The mass was extending lingually bilaterally towards midline and fuses from anterior to posterior region along midline,

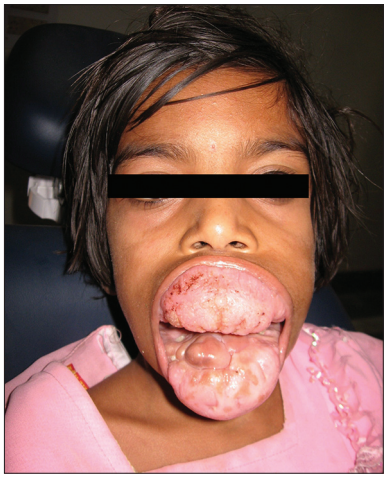

Figure 1: Patient photograph

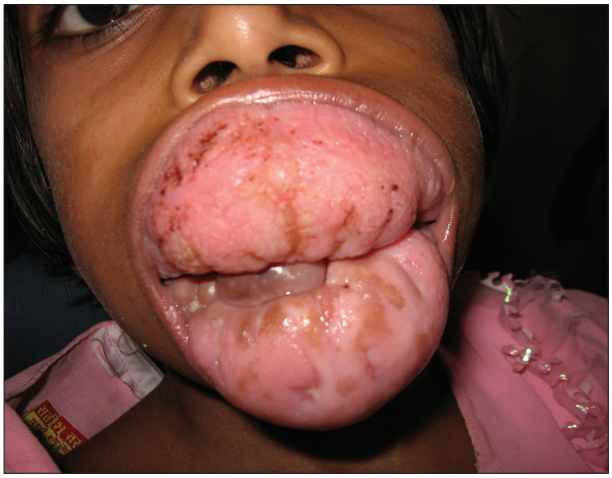

Figure 2: Extraoral view showing a massive, generalized multinodular growth covering both maxillary and mandibular arch completely

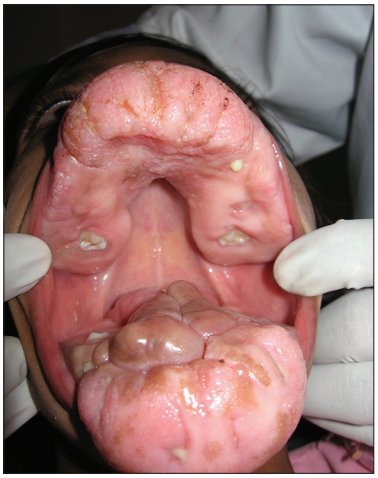

Figure 3: Intraoral view of maxillary arch showing a gross generalized growth of gingiva completely covering the maxillary teeth extending from right maxillary tuberosity to left tuberosity region

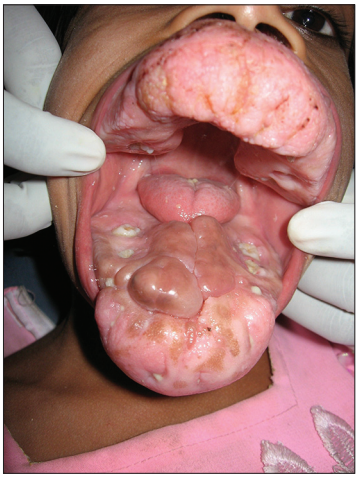

Figure 4: Intraoral view of mandibular arch showing a gross generalized growth of gingiva completely covering the mandibular teeth extending from right mandibular tuberosity to left tuberosity region

reducing the normal tongue space and covering the floor of mouth completely [Figure 4]. The tongue was confined in a small space posterior to fused mass. The mass was exuberant in mandibular anterior region obliterating the chin area and submental region [Figure 4]. The mandibular teeth were not at all visible except in right and left posterior region. The growth was pink to pinkish white in color. The surface was smooth and lobulated. The fusion line was well-demarcated along midline. The tongue was pushed back. On palpation, mass was found to be firm and nontender. There was no associated pain and bleeding noted. There was no associated lymphadenopathy noted.

The patient was advised for panoramic radiograph. The panoramic radiograph [Figure 5] showed multiple retained primary and impacted permanent teeth. The mandible was relatively thinner in angle region bilaterally. On the basis of clinical and radiological findings, a provisional diagnosis of idiopathic gingival fibromatosis has been made. Patient was advised for incisional biopsy. Routine investigation was done and incisional biopsy performed. Histological examination revealed a fibroconnective tissue with extensive collagen bundles. The overlying surface of the epithelium exhibited hyperkeratosis, acanthosis, and elongation of rete ridges. The fibroconnective tissue consisting of densely arranged collagen fiber bundles, numerous fibroblasts, compressed blood vessels, and various degree of focal areas of chronic inflammatory cells [Figure 6a and b]. On the basis of clinical, radiological, and histopathological examination; a final diagnosis of HGF has been achieved. The patient is planned for surgical removal of lesion by CO_2 laser quadrant gingivectomy. But she absconded and so no surgical procedure and follow-up was done.

Discussion

The HGF is a rare condition of the gingival tissues characterized by enlargement of free and attached gingivae. The HGF is classified in two types, nodular type and symmetric type. The nodular type is characterized by presence of multiple tumors in dental papillae, while symmetric type is characterized by uniform enlargement of gingiva. The symmetric type is most common type of gingival enlargement, however there may be combination of both types.[9,10,12-14] The symmetric type is usually unilateral and has generalized or localized form. The localized form usually affects the maxillary molar and tuberosity area, particularly on the palatal surface. In severe involvement, teeth are almost completely covered.[15,16] The onset of gingival overgrowth usually coincides with the eruption of the permanent dentition or with the eruption of the primary dentition. Rarely, it can also be present at birth.[17] The age at onset is divided into the pre-eruptive period (<6 months), deciduous dentition period (6 months-6 years), mixed dentition period (6-12 years), permanent dentition period before adolescence (12-20 years), and permanent dentition period after adolescence (>20 years).[17] Maximal enlargement occurs either during the loss of deciduous teeth or in the early stages of the eruption of permanent teeth. It progresses rapidly during 'active' eruption and decreases with the end of this stage.[4] HGF has not been reported in edentulous patients. Hence, it was concluded that the presence of dentition is necessary for HGF to develop. HGF usually involves the marginal gingiva, however only the interdental papilla involvement is also reported.[18] No gingival overgrowth has been reported after completion of growth of the individual. However, slow enlargement is reported into adult life of few individuals.[10]

Gingival fibromatosis can be caused by number of factors including inflammation, leukemic infiltration, and use of medications such as phenytoin, cyclosporine or nifedipine,[14] and vigabatrin.[19] Gingival enlargement can be part of Laband, Rutherford, Ramon, or Cross syndrome.[20] It has been also suggested that HGF may be due to nutritional hormonal factors but it is not proven.[21] The genetic heterogeneity seems to play an important role in the development of the HGF. HGF can occur as an isolated disease affecting only gingiva, or as part of a syndrome or chromosomal abnormality, and

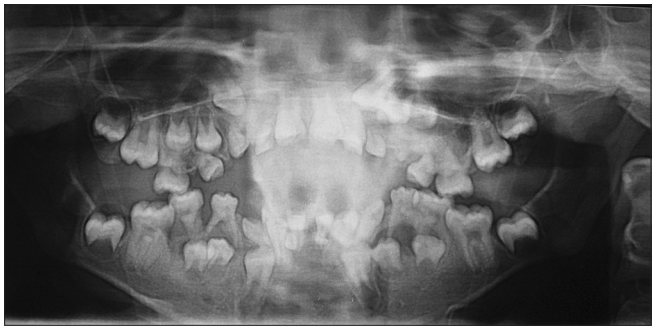

Figure 5: Panoramic radiograph showing multiple retained primary and impacted permanent teeth. The mandible was relatively thinner in angle region bilaterally

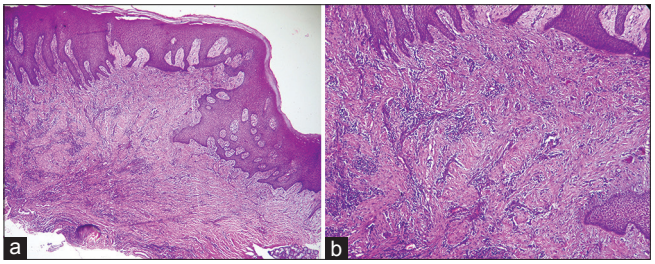

Figure 6: (a) Scanner view of hematoxylin and eosin stained section shows proliferated parakeratinized stratified squamous epithelium and fibroconnective tissue (original magnification, ×40). (b) Low power view of hematoxylin and eosin stained section shows fibroconnective tissue consisting of densely arranged collagen fiber bundles, numerous fibroblasts, compressed blood vessels, and focal areas of chronic inflammatory cells (original magnification, ×100)

both autosomal dominant and recessive forms of this disorder have been described in the literature.[7,22-24] In a study of few families affected with HGF, no linkage with human leukocyte antigen (HLA) was observed.[19] The condition is most frequently reported to be transmitted as an autosomal dominant trait, and recently at least two gene loci on the short arm of chromosome 2 that are responsible for gingival fibromatosis were identified in a Brazilian family. One locus was located in 2p21-2p22[25] and the other was located more proximally in the region of 2p13-p16.[15,26] Xiao *et al*.,[27] identified a new locus (GINGF2) located on chromosome 5q13-q22.

All the forms of HGF is not characterized histologically, especially some syndromic forms. HGF usually involves moderate hyperplasia of a dense, hyperkeratotic epithelium with elongated rete ridges.[28-30] Epithelial hyperplasia can also occur as a consequence of acanthosis, but this was found only in areas of chronic inflammation in HGF.[31,32] The connective tissue in HGF exhibits an accumulation of excess collagen, and elastic and oxytalan fibers; but has relatively few fibroblasts and blood vessels.[28,30] Enlarged fibroblasts appear to alternate with thin and thick collagen fibrils. Unlike in normal gingiva, the collagen fiber bundles are oriented mostly parallel to one another.[33,34] However, small osseous calcifications and abundant neurovascular bundles may also be present.[33,35] HGF does not usually involve inflammation, and local accumulation of inflammatory cells can be found only in cases where pseudo pocketing resulted in plaque accumulation.[11] Diagnosis of HGF is based on medical history and clinical examination, since there are currently no specific immunohistochemical markers available.

There are various procedures available for removal of HGF including surgery, electrocautery, and use of a carbon dioxide laser. The carbon dioxide laser is treatment of choice which has been used in a number of studies.[36-39] If no carbon dioxide laser is available, the conventional external bevel gingivectomy is the most effective method for removing large quantities of gingival tissue particularly when there is no attachment loss and false pocketing is present.[14,40] Ramer *et al*.,[14] advocated quadrant by quadrant gingivectomy with periodontal pack placement for 1 week, followed by 0.2% chlorhexidine oral rinse twice a day for 2 weeks after each surgery. The appropriate time for removal of HGF varies: Emerson[41] recommended the best times to be at the ages of 3, 6, and 12 years. Oral hygiene and the superimposition of plaque accumulation have a crucial effect on the prognosis of HGF.

Recurrence rate in HGF is very high after surgery, so the patient should be followed for considerable period of time and may require repeated surgery. The appropriate time of removal of recurrent gingival enlargement varies. Emerson recommended that the best time is when all the permanent teeth have erupted.[41]

Conclusion

Patients with gingival hyperplasia should be examined carefully. The routine blood investigation and drug history should be taken to exclude blood dyscrasias and drug-induced hyperplasia. While the gingiva may be the only tissue involved, some cases of gingival fibromatosis may be associated with hypertrichosis, and/or mental retardation, and/or epilepsy. Treatment is not required in all cases of idiopathic gingival hyperplasia. If functional impairment exists, surgical excision is indicated.

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