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Case Report

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Hereditary gingival fibromatosis: A family case series.

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Abstract

Hereditary gingival fibromatosis is a rare disease characterised by severe gingival enlargement, which could result in serious aesthetic, functional impairment and emotional problems. Hereditary gingival fibromatosis can occur as an isolated disease or as part of a chromosomal abnormality or syndrome. Here we report a case of a 12-year-old female patient with generalized severe gingival enlargement covering almost all of the teeth and diagnosed as HGF. She had severe diffuse gingival enlargement of the maxilla and mandible. Diagnosis was made based upon family history and clinical examination. Her family history was of significance, since her father, her 3 year old sister and her 19 year old brother suffered from the same symptoms.

Key Words: Hereditary gingival enlargement, gingivectomy, recurrence, gingival fibromatosis

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Introduction

Hereditary gingival enlargement (HGF) is a rare condition with incidence of 1 in 750, 000 people [1]. According to various studies the pedigree analyses of HGF families confirm the autosomal dominant inheritance of HGF trait, although autosomal recessive or even as X linked inherited cases have also been reported in some literature [1, 2]. Mutation in SOS-1 or son-of-sevenless gene is responsible for this disease; this has been reported by several authors (2-5). Autosomal-dominant forms of gingival fibromatosis, which are usually non-syndromic, have been genetically linked to the chromosomes 2p21-p22 and 5q13-q22 [1–5]. The condition does not manifest itself until after eruption of permanent teeth and is most commonly seen associated with the permanent teeth. This gingival overgrowth is due to excessive production of collagen in the gingival corium (7). HGF is characterized by diffuse enlargement of gingiva that is slowly progressive, sometimes covering the major parts of, or the total, tooth surfaces. This enlargement

may project into the vestibule and floor of the mouth, interfere with normal mastication and even lip closure that make the speech difficult. The enlarged gingival tissue appears firm and pink with exaggerated stippling (7,8). The gingival hyperplasia may be generalized (symmetric) or localized (nodular) can involve the buccal and lingual tissues of both maxillary and mandibular arches. Local involvement mainly affects the maxillary tuberosities and lingual surfaces of lower molars and is typically characterized by the presence of multiple large masses. The symmetric form, which is the most common type of disorder, results in uniform enlargement of the gingiva that is firm, dense, resilient, insensitive fibrous tissue that covers the alveolar ridges and extends over the teeth resulting in extensive pseudo pockets. The degree of enlargement may vary from mild to severe and may be the same between the individuals of the same family (8-10).

We are here presenting a rare case series of a family suffering from hereditary gingival fibromatosis.

Case Presentation

12 years old girl refered to our clinic with her father. Her chief complaint was overgrowth of gingiva surrounding all of her teeth. Family history revealed that similar presentations were seen between siblings and father. Past dental history of father reveals that he got surgical treatment (gingivectomy) before but the condition had reoccurred. Intraoral examination revealed firm enlargement of the gingiva on both buccal and lingual/palatal sides with pinkish red, fibrous inconsistency and absence of stippling (Figure 1,2,3). Gingival enlargement enclosed the major surface of the teeth present except the incisal/occlusal surfaces. The teeth were displaced from their normal position. She also had multiple grossly decayed teeth. Her family members were requested to report for clinical examination. Her younger sister was 8 years of age. She also had bilateral, symmetrical gingival enlargements in the anterior, posterior region of maxillary and mandibular gingiva (Figure 4). The teeth were displaced from their normal position. Her elder brother was 19 years of age. He also had bilateral, symmetrical enlargements, more prominent in the mandibular molar region (Figure 5,6,7). Her father was 52 years old. He gave a history of similar enlargements in his gingiva and underwent gingival surgery 20 years ago for the same reason. Clinical examination revealed recurrence of gingival enlargements in the anterior, posterior region of maxillary and mandibular gingiva (Figure 8,9).

Histopathological Report*:* The sections revealed moderately dense collagenous connective tissue with collagen bundles arranged in a haphazard manner. Connective tissue was relatively avascular along with scanty inflammatory cell infiltrate showing dense wavy bundles of collagen fibers containing numerous fibrocytes and fibroblasts. The overlying epithelium was hyperplastic with elongated rete ridges.

After correlating histopathological results with the family history and clinical presentations, it was diagnosed as hereditary non-syndromic gingival fibromatosis. The patients were advised for gingivectomy and regular

oral hygiene maintenance with follow up as treatment plan.

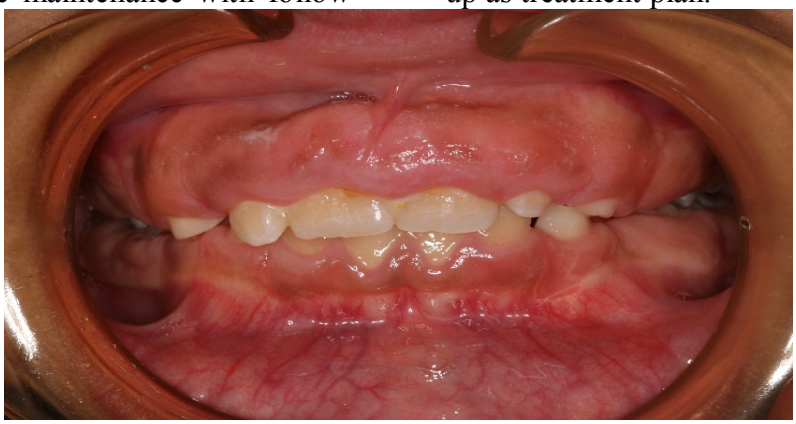

Figure 1: Clinical picture of 12-year-old female with gingival enlargement.

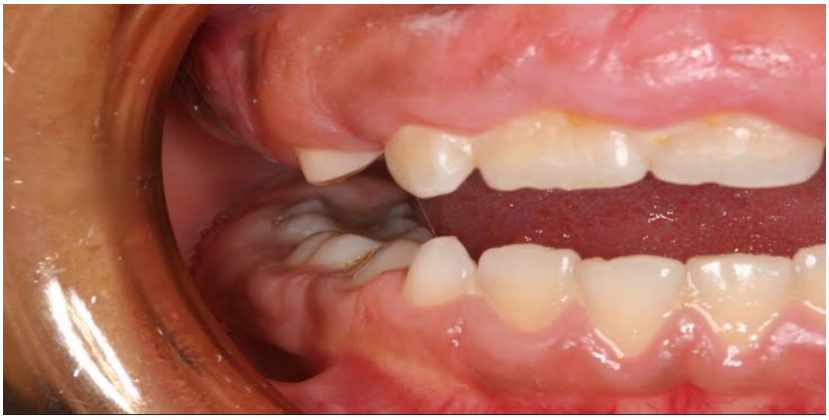

Figure 2: Clinical picture showing prominent enlargement in the posterior region.

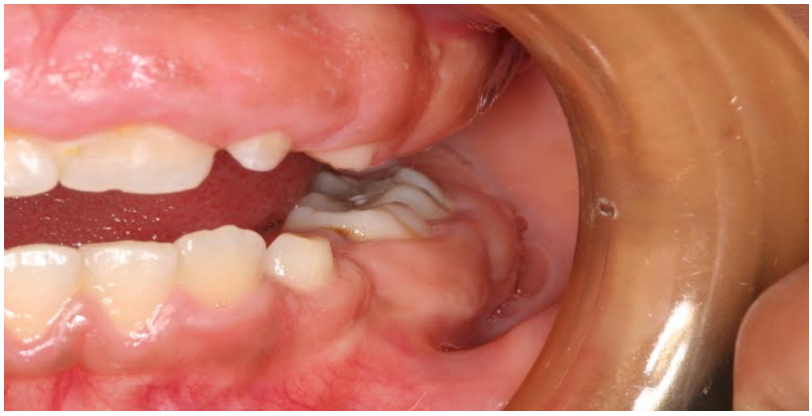

Figure 3: Clinical picture showing prominent enlargement in the posterior region.

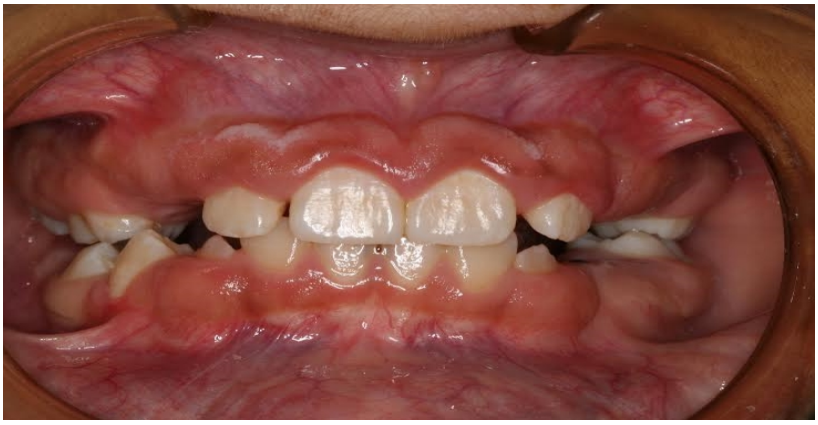

Figure 4: Clinical picture of 8-year-old sister with gingival enlargement.

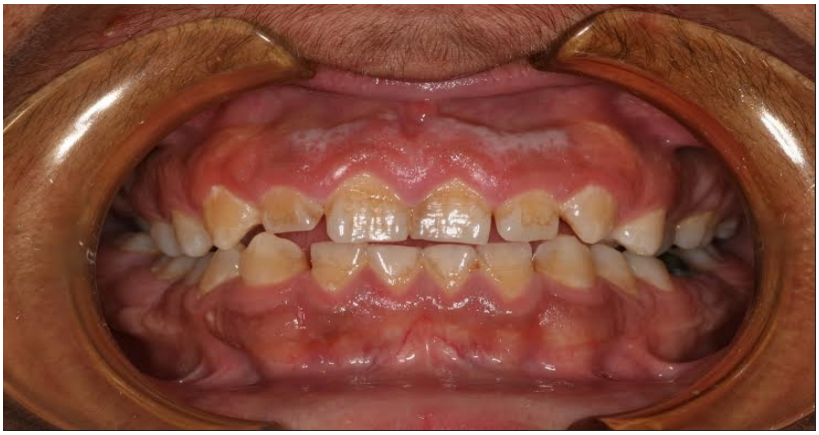

Figure 5: Clinical picture of 19-year-old brother with gingival enlargement.

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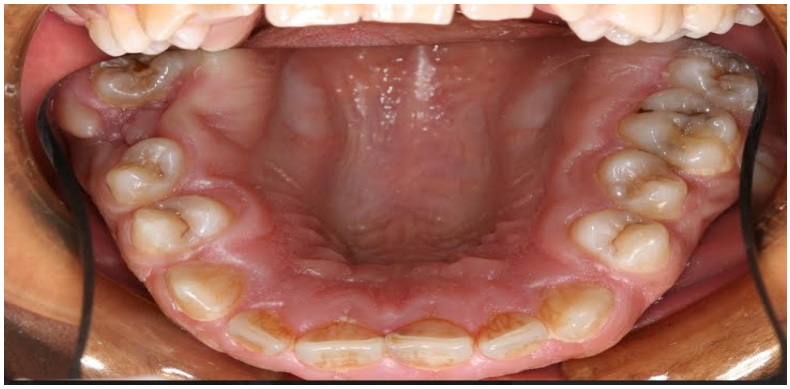

Figure 6: Clinical picture showing prominent enlargement in posterior region.

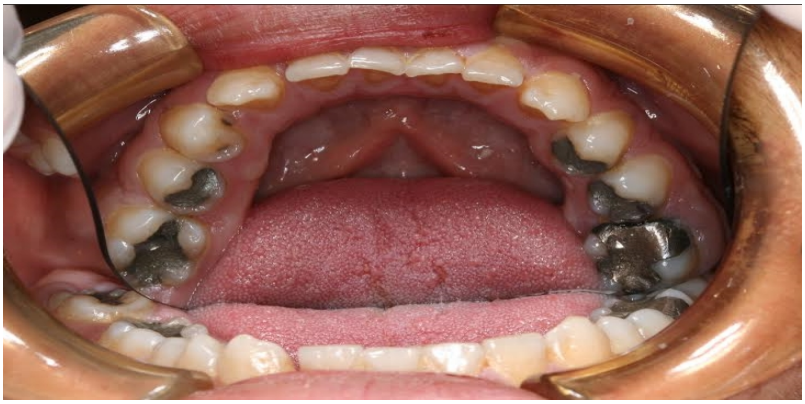

Figure 7: Clinical picture showing prominent enlargement in posterior region.

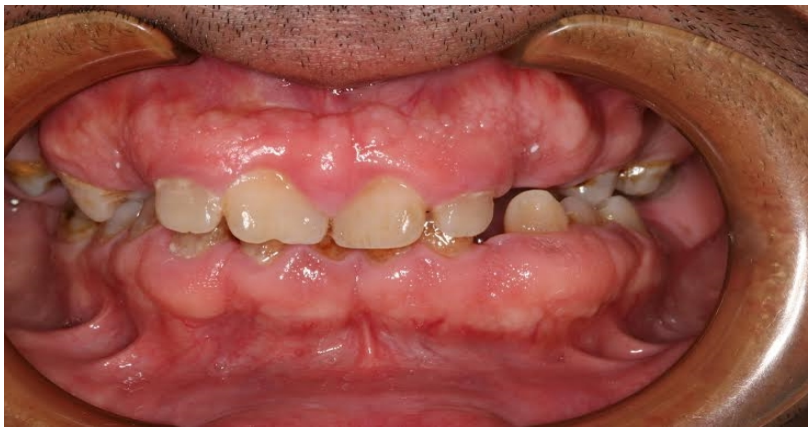

Figure 8: Clinical picture of 52-year-old father with missing teeth.

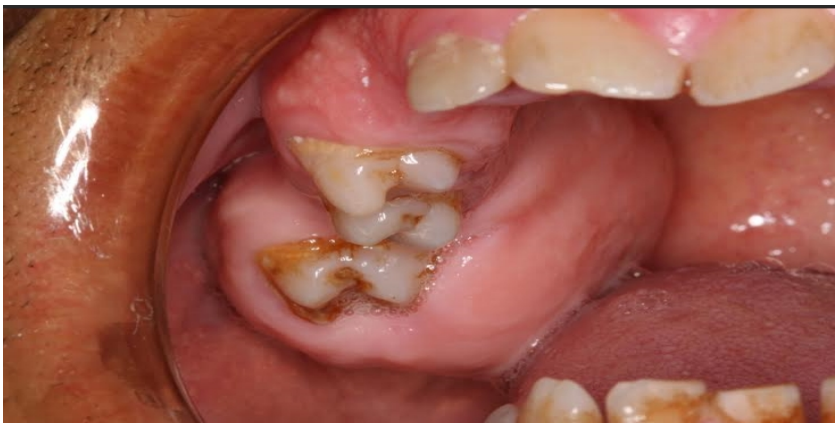

Figure 9: Clinical picture showing gingival enlargement in posterior region

Discussion

This paper reports case series of a

family with idiopathic gingival fibromatosis. HGF occurs due to congenital or hereditary causes. Gingival overgrowth varies from mild enlargement of isolated interdental papillae to segmental or uniform and marked enlargement affecting one or both jaws (7,10). Enlargement of gingiva can be due to use of medication like Phenytoin, cyclosporine, and calcium channel blockers (11), but no drug history was present in the present cases. Gingival fibromatosis can occur as an isolated nonsyndromic condition or be associated with other syndromes. The syndromes associated with GF include Jones hartsfield, Murray- Puretic-Drescher syndrome, Zimmermann-Laband, Rutherfurd, Cross, Ramon, Prune-belly syndrome associated with hearing deficiencies, hypertelorism, and supernumerary teeth (7). In the present cases, family members had no history of any systemic disease, mental retardation, epilepsy, or medication that could contribute to gingival overgrowth. The histologic features observed in the present case had the typical appearance of gingival fibromatosis; hyperplasic dense fibrous connective tissue with acanthotic gingival epithelium (7). As the family history contributes to this case with no systemic and drug history and with no clinical features fulfilling these possible syndromes, diagnosis of isolated generalized hereditary gingival fibromatosis was made. The mechanism of HGF is unknown but could be confined to the fibroblasts in the gingiva. Some authors report an increase in the proliferation of gingival fibroblasts, whereas others report slowerthan normal growth. Increased collagen synthesis rather than decreased levels of collagenase activity may be involved (7,8). Histopathology of affected tissue reveals dense connective tissue rich in coarse connective tissue fibres, with young fibroblasts. The overlying epithelium is of variable thickness and can show hyperkeratosis with elongated rete ridges.

Treatment depends upon severity of enlargement. Severe enlargement needs full mouth gingivectomy. The most preferred method for excision is external bevel gingivectomy especially when there is no bone loss and only false pockets (9). The literature reports high recurrence rate after surgery and required a close follow up. However, in several reported cases there was no recurrence in a period of 2 years, 3 years, or even a 14-year follow-up. There is debate regarding the time of surgery. Eruption of complete set of permanent teeth is the recommended time for surgery (7). The father had previously undergone gingivectomy and now recurrence occurred again. The treatment plan advised for the affected patients is gingivectomy with regular oral hygiene measures and frequent followup.

To conclude, herewith we report the case series of HGF affecting the father and all his children. A multidisciplinary management is advised to completely rehabilitate the patient.

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