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CASE REPORT

Hereditary Gingival Fibromatosis with Distinctive Facies

Sunkara Shree Ramalinga Prasad, Chitturi Radharani, Soumya Sinha, SV Kiran Kumar

ABSTRACT

Hereditary gingival enlargement also known as gingivitis or familial elephantiasis is a rare type of gingival enlargement. It appears as an isolated autosomal dominant disorder or maybe associated with other conditions. Oral manifestations may vary from minimal involvement of only tuberosity area and the buccal gingiva around the lower molars to a generalized enlargement inhibiting eruption of the teeth. This paper discusses the case of a 13-year-old female patient with distinctive facial characteristics who presented to the department with a chief complaint of swollen gums since 1 year. She had severe diffuse gingival enlargement of the maxilla and mandible. Diagnosis was made based upon clinical examination and family history. Quadrant wise internal bevel gingivectomy procedure was done for the patient to restore her functional and esthetic needs.

Keywords: Hereditary gingival fibromatosis, Gingivectomy, Synophrys.

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BACKGROUND

Hereditary gingival fibromatosis (HGF) (gingivostomatitis/diffuse fibroma/idiopathic gingival enlargement/idiopathic fibromatosis/familial elephantiasis)[1] is defined as a rare, benign , asymptomatic , nonhemorrhagic and nonexudative proliferative fibrous lesion of the gingival tissues.[2] It was first reported by Goddard and Gross in 1856.[3]

HGF can be generalized or localized in nature with males and females being equally affected at a phenotype frequency of 1: 1,75,000. The gingival enlargement usually begins at the time of eruption of the permanent dentition but can develop with the eruption of the deciduous teeth and is rarely present at birth. The presence of teeth seems to be necessary for HGF to occur because the condition disappears or recedes with the loss of the teeth. The growth may worsen throughout adolescence suggesting the role of sex hormones.[4]

HGF is traditionally considered as an autosomal dominant disease. The familial variations may occur as an isolated finding or in association with one of several hereditary syndromes, for example, Zimmermann-Laband, Murray-Puretic-Drescher, Rutherfurd, multiple hamartoma and Cowden syndrome. In modern times, a mutation in the son of sevenless-1 (*SOS-1*) gene has been suggested as a possible cause of isolated (nonsyndromic) gingival fibromatosis. However, no definite linkage has been established.[3] Hart et al[5] in an extensive review have enumerated 18 syndromic conditions associated with gingival fibromatosis. Also, some isolated generalized gingival lesions, such as plasma cell gingivitis, plaque-induced gingival hyperplasia and scurvy may show generalized symmetric gingival enlargements. These conditions can be differentiated from HGF on the basis of medical history, clinical examination and histopathological findings. For example, in plasma cell gingivitis, the gingival enlargement is red in color and can be symptomatic and histopathological examination reveals dense infiltrate of plasma cells separated into aggregates by strands of collagen. Orofacial granulomatosis can be demarcated from HGF by observing the features of orofacial granulomatosis like lip swelling, cobblestoning, in addition to gingival enlargement which are absent in HGF. In Crohn's disease, the orofacial features are similar to that of orofacial granulomatosis and there are additional GIT features of inflammation of ileocecal region. Plaque-induced gingival hyperplasia usually presents with reddish swollen gums that may show bleeding. In scurvy, in addition to the gingival enlargement, there can be gingival ulceration along with moderate to severe bleeding of the gums which is absent in HGF. In sarcoidosis the gingival enlargement is granular in appearance which is associated with other intraoral features, such as oral ulceration, split nodules on the palate, bone lytic lesions and jaw bone overgrowth, xerostomia. Wegener's granulomatosis is a vasculitides in which the

gingival enlargement takes a specific texture and color of a strawberry and is thus known as strawberry like gingivitis.[6]

The exact pathogenesis of the disease is unknown. However, the results of various morphological and molecular analysis suggest as follows:[7]

- The proliferation of the HGF fibroblasts is significantly higher and more cells are found in G2/ M and S phases of the cell cycle.
- Fibroblasts from HGF produce 30 to 50% more collagen than normal fibroblasts.
- There is a decreased level of extracellular matrix degradation due to the decreased level of expression of MMP-1 and MMP-2.
- Increased levels of transforming growth factor B1 stimulate synthesis and reduce degradation of the extracellular matrix.[8]
- There is a marked expression of ALPase in gingival fibromatosis-derived cells. A hypothesis has been put forward suggesting a positive correlation between the presence of ALPase and the clinical appearance of bulbous bony contours commonly seen in patients with HGF.[9]

Gingival enlargement results in both esthetic and functional problems for the affected individual. The most common effects are diastemas, malpositioning of teeth, prolonged retention of the primary dentition, delayed eruption, crossbites and open bites, prominent lip and open lip postures. Although gingival enlargement does not directly affect the alveolar bone, the gingival swelling may increase the bacterial plaque accumulation inducing periodontitis with bone resorption and halitosis. Michael Fritz[10] reported a case of idiopathic gingival fibromatosis with extensive osseous involvement in 12-year-old boy.

This case report describes the dental management of an unusual case of gingival fibromatosis with distinctive facies in a 13-year-old female patient.

CASE DESCRIPTION

A 13-year-old female patient reported to the Department of Pediatric and Preventive Dentistry, PMNM Dental College and Hospital, Bagalkot, Karnataka with the chief complaint of swollen gums from the past 1 year. The swelling had been insidious in onset and had progressed slowly to the present size. There was no history of fever or bleeding on brushing.

The medical history did not reveal any signs of epilepsy or any other metabolic disorder though they do report of an operation that the patient underwent at the age of 1 month for pyloric stenosis. The family history was important because it indicated a consanguineous marriage of the parents. Also patient's maternal aunt had a similar clinical case presentation.

General examination of the child revealed the child to be of a short stature, with normal motor coordination and good personal hygiene. She had stubby fingers and a hoarse voice. Patient presented with a peculiar facies: Bushy eyebrows with synophrys, flat nasal bridge, cupid bow mouth, (Fig. 1), bimaxillary protrusion, convex profile (Fig. 2) and high-arched palate (Fig. 3). Intraoral examination revealed generalized, diffuse overgrowth of gingiva (Fig. 4). Overlying mucosa appears smooth. It is nontender, firm, no tendency to bleed, nonfluctuant and noncompressible (Fig. 5).

An orthopantomogram was done for the patient that showed no disharmony between dental, skeletal and chronologic ages or any gross bony deformity. Various blood and urine investigations were done for the patient. All parameters were in the normal range except for serum alkaline phosphatase levels which were considerably raised.

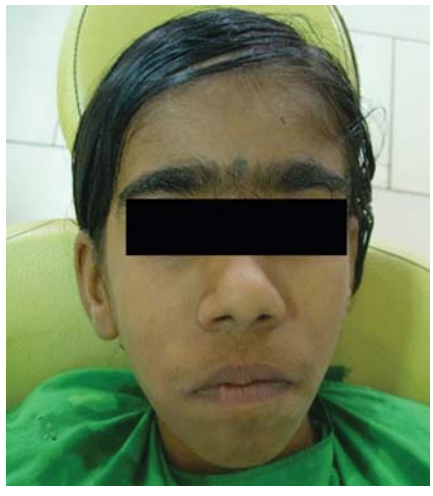

Fig. 1: Front view of the patient's face. Notice the bushy eyebrows, synophrys and cupid bow mouth

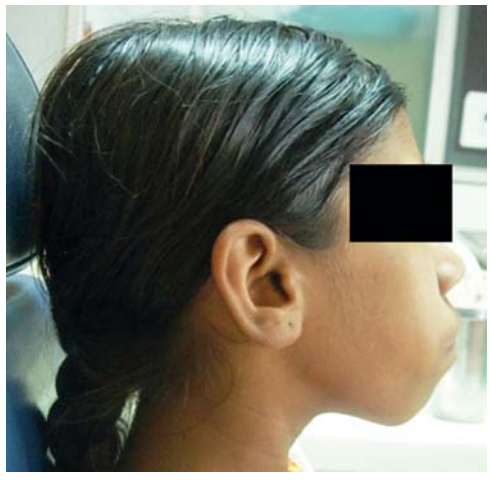

Fig. 2: Profile view of the patient. Appreciate the convex profile, flat nasal bridge and the bimaxillary protrusion

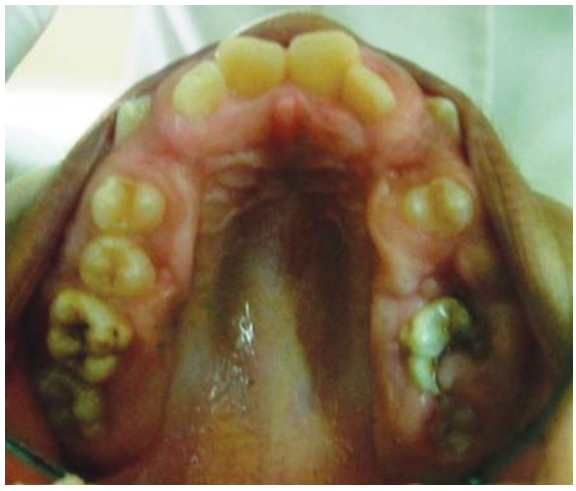

Fig. 3: Preoperative maxillary occlusal view

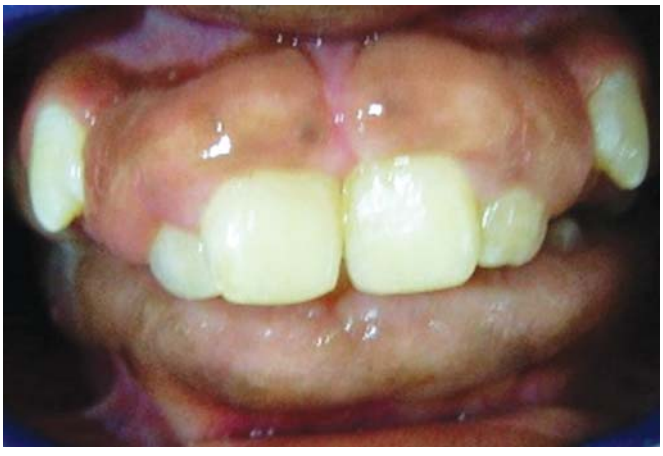

Fig. 4: Preoperative intraoral frontal view

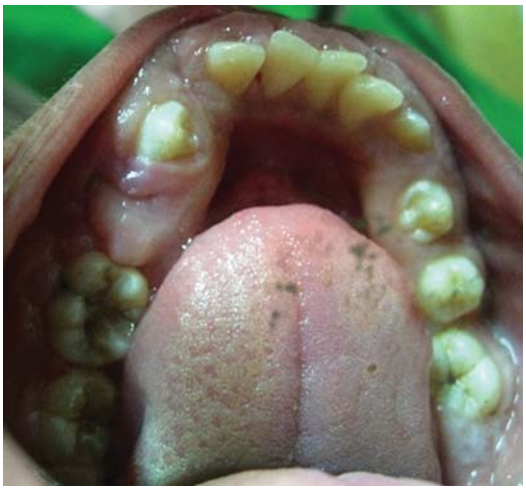

Fig. 5: Preoperative mandibular occlusal view

A diagnosis of 'HGF with unusual facies' was made based upon clinical presentation, family history and drug history.

After obtaining parental consent a phase 1 therapy comprising of supra- and subgingival scaling, restorations and extractions of over retained deciduous teeth was done. Internal bevel gingivectomy was planned to remove the excess gingival tissue. After using pocket markers, incisions at an angle of 45° were given using Bard Parker blade number 12 and 15. Full thickness flaps were elevated and then internally thinned. Bulbous bony contours and ledges were removed. Final gingival contours were created using electrocautery. Thinned flaps were sutured with interrupted suture technique using 3-0 silk sutures. Quadrant wise surgeries spaced out at a gap of 2 weeks were performed.

Postoperatively, the patient was put on antibiotics and analgesics for 5 days and 0.12% chlorhexidine mouthwash for 2 weeks. The patient tolerated the procedure well and the postoperative healing was uneventful (Figs 6 to 8).

The excised gingival tissue was sent for a histopathological evaluation. Microscopic finding showed parakeratinized stratified squamous epithelium with elongated rete pegs at some sites and some pseudoepitheliomatous hyperplasia at the same site. The underlying connective tissue showed dense fibrocellular proliferation with few blood capillaries. The overall

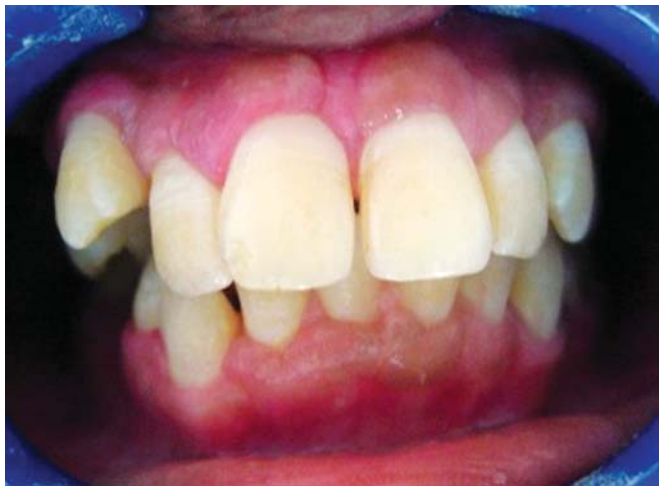

Fig. 6: Postoperative intraoral frontal view

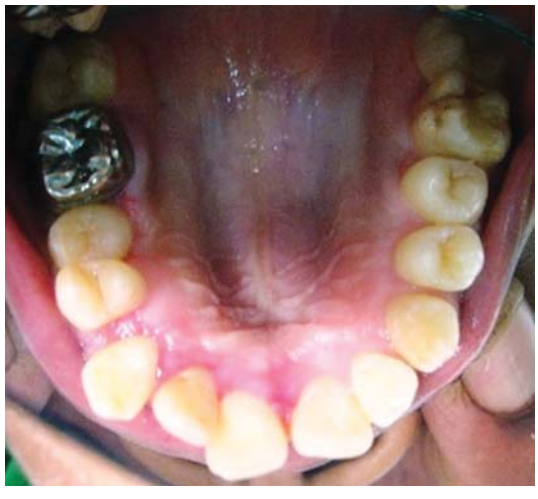

Fig. 7: Postoperative maxillary occlusal view

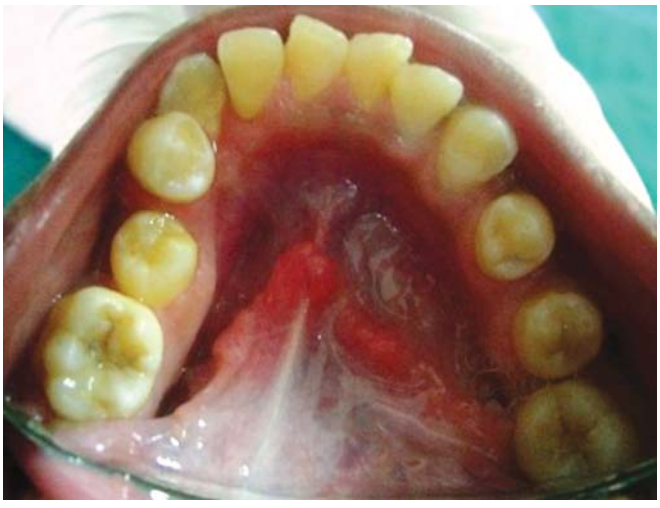

Fig. 8: Postoperative mandibular occlusal view

histopathological features correlated with the clinical features were suggestive of HGF.

Patient and the guardians were happy with the treatment outcome. Patient is on regular follow-up in the department to identify any early signs of recurrence.

DISCUSSION

Earlier authors recommended extraction of all the teeth with alveolar reduction as the treatment for HGF. However, this is not an acceptable form of therapy in children, where a more conservative approach is desirable. Many techniques have been used for the excision of gingival tissue, including external and internal bevel gingivectomy in association with gingivoplasty, an apically-positioned flap, electrocautery and carbon dioxide laser.

There is no general agreement as to the timing of surgical intervention. Emerson prefers to wait until the patient reaches the age at which eruption of all permanent teeth might normally be expected. Rushton[11] suggests waiting for 1 or 2 years past the time when the teeth should have erupted.

Recurrence is inevitable and has been reported to occur as soon as 3 months and as late as 14 years postoperatively, but is expected to be minimal if surgical excision is performed after the eruption of permanent teeth.[12] Emerson demonstrated that correct physiologic contours of the marginal gingival and maintenance of good oral hygiene are more important to prevent recurrence.

CLINICAL SIGNIFICANCE

The gingival enlargement results in multiple esthetic and functional problems for the affected individuals. Speech and mastication can be affected depending on extend and severity of the overgrowth. It often requires surgical intervention and excision of the overgrown tissue, even though recurrence cannot be predicted. Although it is a rare condition, it is possible to encounter it in our everyday practices. Correct diagnosis and treatment of the affected individuals is important.

CONCLUSION

Gingival enlargement is a prevalent disease in children, conferring to the pedodontist and periodontist an important role in the correct diagnosis and treatment of the affected patients. Esthetic and dental-associated alterations can considerably reduce quality of life. The recurrence of HGF cannot be predicted. Patients should be given every opportunity to undergo conservative surgical procedures. The aims are to minimize the serious emotional and social problems and functional problems associated with HGF patients.

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