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Case Report

HEREDITARY GINGIVAL FIBROMATOSIS- A RARE AUTOSOMAL DOMINANT DISORDER

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ABSTRACT

Hereditary gingival fibromatosis is a rare autosomal dominant disorder occurring either as an isolated trait or as a manifestation of a systemic disorder or syndrome which is characterized by a generalized diffused involvement of the gingiva covering the entire surfaces of the crowns. The pathogenesis of the disease is still completely unclear, however mutations in the son of sevenless gene-1 (SOS-1) has been attributed for its occurrence in few forms of the disease. This paper discusses a case of a 13 year old female patient with generalized diffused gingival enlargement and a positive familial history. Though definitive treatment for the management of the condition is not available surgical gingivectomy with external bevel incision was performed and maintained for a period of 2yrs

Keywords: fibromatosis

INTRODUCTION:

Hereditary gingival fibromatosis also called as gingivostomatitis/ diffused fibroma/ idiopathic gingival fibromatosis/ familial elephantiasis[1]is a rare autosomal dominant disorder,[2]although sporadic cases are common and autosomal recessive inheritance has been reported.[3]It is found to occur in 1in 750,000 individuals and is characterized by a slow, progressive increase in the size of gingiva[4].It involves the overgrowth of the keratinized gingival tissues with varying degree and can be localized or generalized. Most recently, mutation in the son of sevenless-1 gene has been concentrated as a genetic factor responsible for hereditary gingival fibromatosis.[5]Linkage studies havelocalized loci for autosomal dominant non-syndromic formsof gingival fibromatosis (adHGF) to chromosomes 2p21– p22,[6,7] 2p22.3–p23.3,[8] and 5q13–q22.[9] The enlargement can occur as an individual manifestation or in association with various syndromes such as Zimmerman laband syndrome, Rutherford syndrome, Jones syndrome, Cross syndrome, Murray Puretic Dreshers syndrome, Ramon syndrome etc.[10,11]

The enlargement can occur either during the pre-eruptive period, eruptive period of deciduous or permanent dentition or after complete eruption of the teeth. Maximal enlargement happens either during the loss of deciduous teeth or in the early stages of eruption of permanent teeth. It progresses fast during the stages of active eruption and decrease by the end of this stage. The clinical features of HGF include generalized firm and fibrous gingival overgrowth, covering the entire or partial surfaces of crowns of the teeth. The enlarged gingivae appear normal in colour and feel firm and nodular on palpation. Exaggerated stippling may be present.The overgrowth may also result in functional and esthetic concerns, create diastemas, impede or delay tooth eruption, and

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create changes in facial appearance as a result of lip protrusion. Severe overgrowth can result in crowding of the tongue, speech impediments, and difficulty with mastication, and can prevent normal closure of lips[12]

The typical histologic appearance of the affected tissue includes hyperplastic epithelium with elongated rete ridges extending deeply into the underlying connective tissue. Coarse and fine dense bundles of collagen, oriented in all directions, and a few "plump" fibroblasts have been described as making up the connective tissue layer. A more cellular specimen with large fibroblasts, small calcified particles and small foci of bone has also been described.[13,14] The histologic features are nonspecific, and a definitive diagnosis of hereditary gingival fibromatosis can be made only in the presence of an adequate history and clinical examination.

Case report

A 13 year old female patient reported to the department of periodontics with the chief complaint of non-symptomatic gingival overgrowth covering almost the entire length of the crowns since the past 4-5 years and also a positive familial history where the patients mother also had a similar complaint which was treated by surgical intervention. The swelling was noticed during the age of eruption of the permanent dentition and has gradually increased covering the entire dentition. The patient's medical history revealed no contributory findings except for a slight hearing deficit which was noticed since her childhood. No mental retardation, skeletal deformities, or defects of skin or finger nails were observed. On general examination, the patient was found to be reasonably active and of normal built and gait. The patient is not under any medication for any kind of illness. Extra-oral examination revealed bilateral symmetrical face, with convex profile and bimaxillary protrusion.

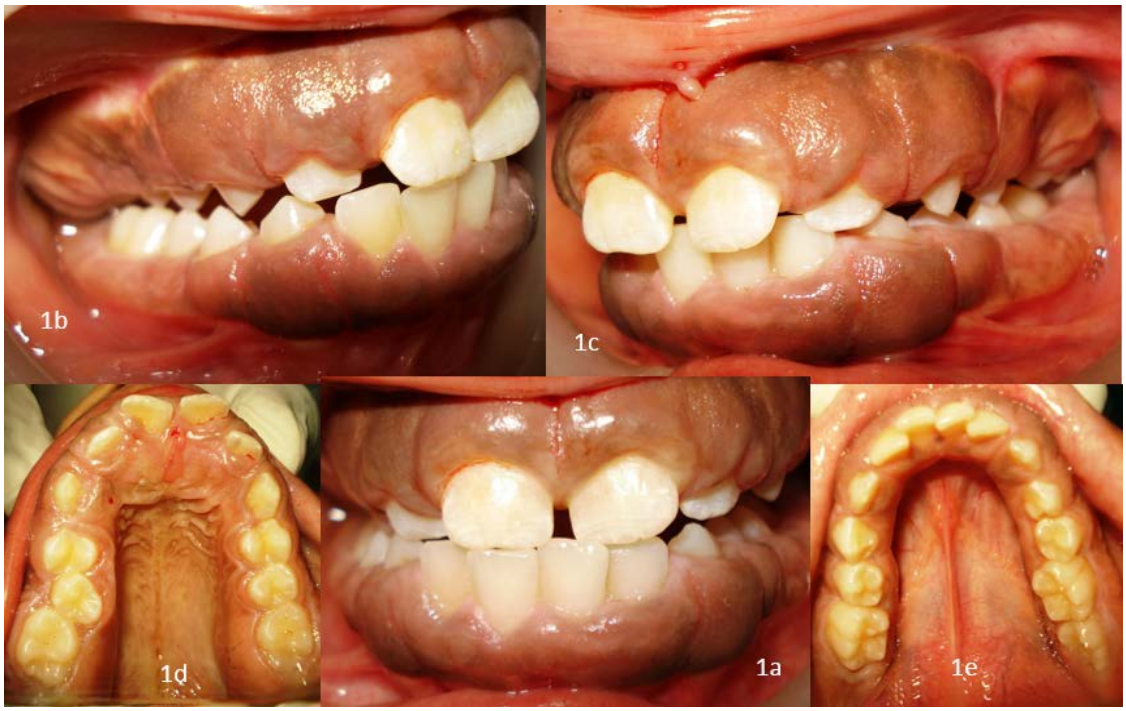

Figure 1: Pre-operative Figures Showing Generalized diffused Gingival overgrowth

On Intra-oral examination, oral mucosa, buccal mucosa, labial mucosa, palate, tongue, vestibule, floor of the mouth revealed no abnormality. Gingival examination showed diffused gingival enlargement, on both buccal and lingual surfaces and covered more than two-thirds of the clinical crown and un-erupted maxillary and mandibular second molars(figure 1). An orthopantamograph was advised which revealed the presence of complete set of permanent dentition, with four impacted third molars and incompletely erupted second molars in all the four quadrants (figure 2).

Complete blood investigations were advised which revealed no abnormal findings.

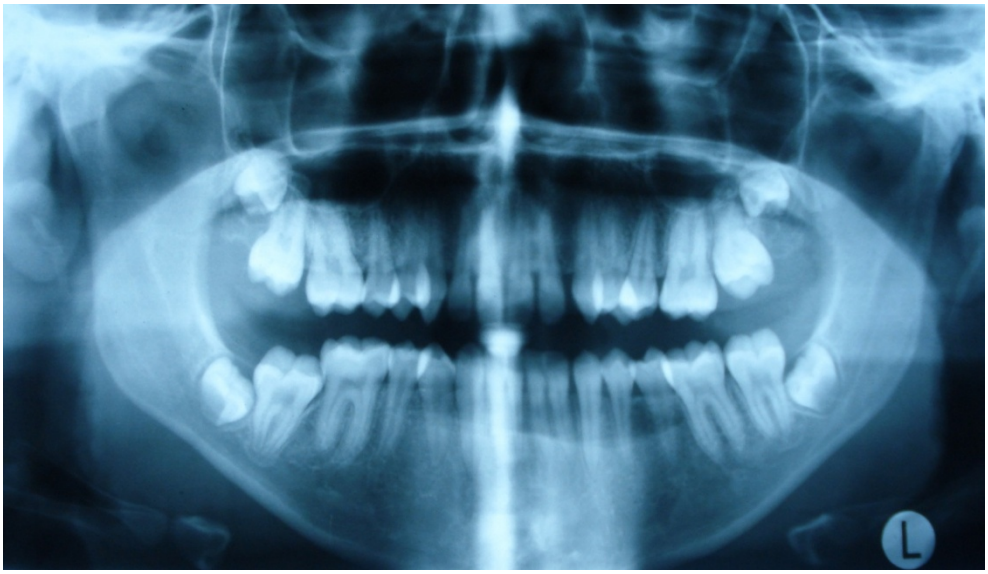

Figure 2: Orthopantamograph showing impacted permanent 3rd molars and partially erupted 2nd molars

After thorough investigations and scaling and root planing in two divided appointments to reduce the inflammation of the gingival tissues, surgical intervention was planned. Maxillary and mandibular external bevel gingivectomy was planned on both buccal and palatal aspects along with the surgical removal of the pericoronal flap covering the maxillary and mandibular second molars to facilitate complete eruption. The Gingivectomy was performed under local anaesthesia containing adrenaline in four divided appointments. A biopsy sample of the gingival tissues was obtained and submitted for histological examination(Figure 3). The wound area was protected from external environment using a tin foil and periodontal dressing. A twice daily use of 0.2% chlorhexidine gluconate mouth rinse was advised for a period of two weeks. Post-surgical healing was uneventful. Supportive periodontal management was done every 3 months during the first year and no recurrence has been observed (Figure 4).

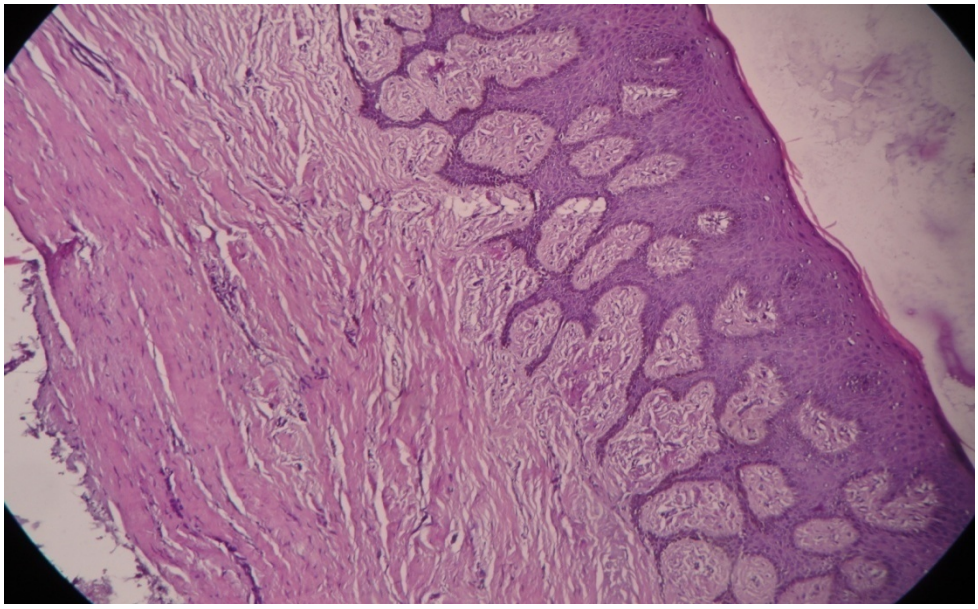

Figure 3: histological picture showing hyperplastic epithelium with elongated rete ridges and few plump fibroblasts

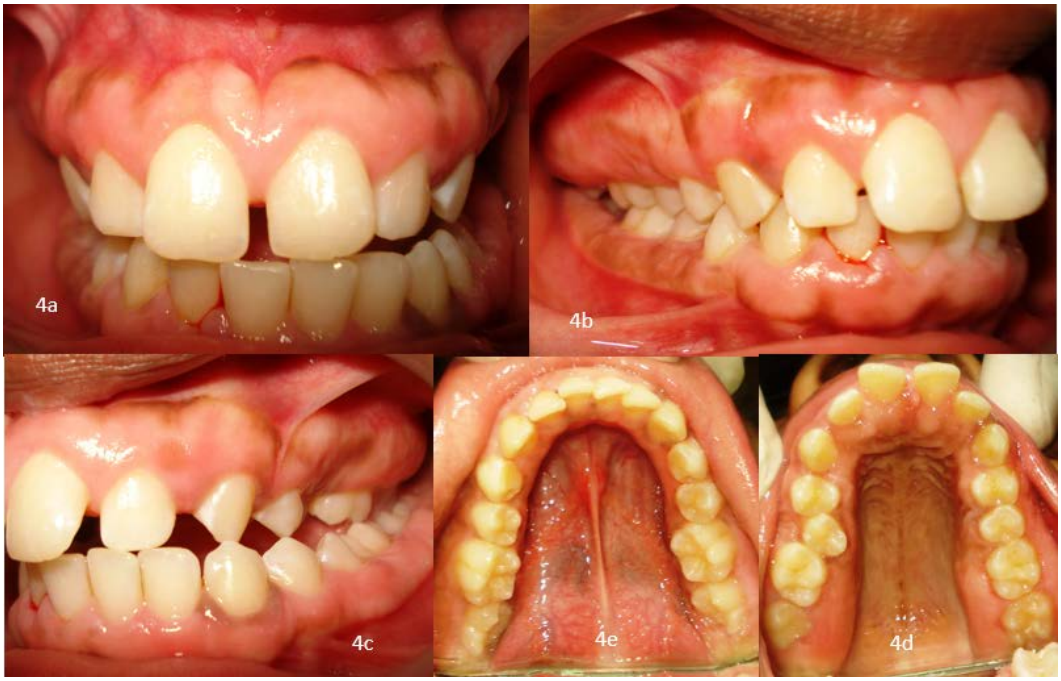

Figure 4: post operative figures after complete treatment with external bevel gingivectomy

Discussion

Hereditary gingival fibromatosis depicts the generalized gingival enlargement occurring during the stages of eruption of the permanent dentition. It usually manifests as an isolated condition or may be associated with other systemic conditions in the form of syndromes. Most of the associated syndromes may include epilepsy, deafness, hypertrichosis, mental retardation along with hereditary gingival fibromatosis. However the clinical manifestations in the current patient presents as an isolated condition with only a mild hearing deficit which is not remarkable. However non syndromic isolated generalized symmetric gingival enlargements can also be seen in other conditions such as Plasma cell gingivitis, Plaque induced gingival hyperplasia, Drug (phenytoin, cyclosporine, and calcium channel blockers) induced gingival enlargement, Tuberculous sclerosis, Scurvy,Leukemia (Acute myeloid leukemia), Oro-facial granulomatosis, Crohn's disease, Sarcoidosis, Wegener's granulomatosis, Amyloidosis etc.

The diagnosis of hereditary gingival fibromatosis can be made on the basis of medical history, clinical examination and histological examination. The present case shows a definitive genetic association with prevalence of a similar condition in the patients mother, and the development of the condition which has started at the age of eruption of the permanent dentition. The clinical examination reveals the presence of diffused nodular type of gingival enlargement with firm and fibrous type of gingiva and increase in the surface stippling. The increase in the surface stippling can be correlated to the histological picture which depicts an increase in the length of the rete ridges and an increase in the connective tissue component.Bozzo et al(1994)[2] reported a four generation family with 50 affected offspring from a total of 132 members suffering from HGF with all the subjects showing similar expression of disease. Similarly Hart TC (2000)[15] reported a four generation pedigree with variable expression of the disease. In the current report findings are similar to the documentation done by Bozzo (1994).

The radiographs also reveal the presence of incompletely erupted 2nd molars which are covered by a dense and firm connective tissue component, which could have hindered the process of eruption. Therefore during the surgical procedure the gingival tissue covering the unerupted third molars are removed to uncover the occlusal surfaces of the teeth and to provide a path for eruption. During the two year follow up period there is no evidence of recurrence of the

condition. As reported in the earlier case reports hereditary gingival fibromatosis is an uncurable condition with high affinity towards recurrence requiring long term follow-up of the patient. However effective supportive periodontal management could be beneficial in providing successful long term results and preventing the recurrence of the disease.

Conclusion:

Hereditary gingival fibromatosis is a rare familial disorder which requires development of new treatment strategies and approaches including gene therapy to modify the genetic susceptibility and to prevent the recurrence of the disease.

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