Hereditary Fibromatosis in a Child-A Case Report

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ABSTRACT

Enlargement of keratinized gingival tissues can be due to local irritating factors, drug-induced or hereditary. Hereditary gingival fibromatosis is attributed to mutation in son-of-sevenless gene (SOS1). We reported a case of a 9-year-old male with a major complaint of gingival enlargement and its associated unaesthetic appearance. A similar gingival enlargement was also found in a sibling without any associated drug history or syndromic conditions. Gingivectomy was done and he was followed for 6 months during which there was no recurrence. We recommend early intervention to ensure the prevention of malocclusion and aesthetic complications.

Key words: Family history, Hereditary gingival hyperplasia/fibromatosis, Gingivectomy, Early intervention

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INTRODUCTION

Hereditary gingival fibromatosis (HGF) is a progressive benign enlargement of the free and attached gingiva.[1] It is a rare gingival condition with incidence of 1 in 750,000 people.[2] It may occur as a non-syndromic or as a part of a syndromic condition.[2,3] Gingival fibromatosis can be hereditary or idiopathic.[3] The hereditary type may be isolated or can occur with hereditary syndromes such as Rutherfurd syndrome, Zimmermann-Laband syndrome, Murray-Puretic-Drescher syndrome, Jones syndrome, Ramon syndrome, Juvenile hyaline

fibromatosis, multiple hamartoma, and Cowden syndrome.[3-5] Autosomal dominant inheritance associated with chromosome 2p21-p22 and 5q13-q22 or recessive mode of inheritance has been reported.[6-8] A positive family history is always present in HGF;[6] while mutation of the son of sevenless-1 (SOS-1) gene and inadequate growth hormone release factor are the possible aetiology of isolated gingival fibromatosis.[3,6,7] The pathogenesis of gingival fibromatosis is attributed to an increase in the proliferation of gingival fibroblasts or decreased levels of collagenase activity.[1,2,7,8]

There are two clinical forms of gingival fibromatosis; the symmetric and nodular form.[8] The symmetric form which is the most common type appears as a uniform enlargement of the gingiva, while the localized nodular form is characterized by multiple enlargements of the gingiva.[7,8] The gingival enlargement varies from mild, moderate to severe form. Gingival fibromatosis can either begin with the eruption of the primary or permanent dentition.[8,9] However, it is rarely present at birth.[8] The condition is usually associated with teeth malpositioning, prolonged retention of primary dentition, crossbite, open-bite, prominent lips, altered lip, functional and esthetic problems.[8-11]

Hereditary gingival fibromatosis appears histologically as dense, avascular, collagenous connective tissue with a small number of chronic inflammatory cells underlining the surface epithelium.[9,11] Elongated rete processes may be observed at the attached gingival epithelium.[10]

This report aimed to present the management of a 9-year-old with HGF; to add to literature regarding this rare condition and to emphasize on the exigency for early intervention to avoid various aesthetic and functional problems.

CASE REPORT

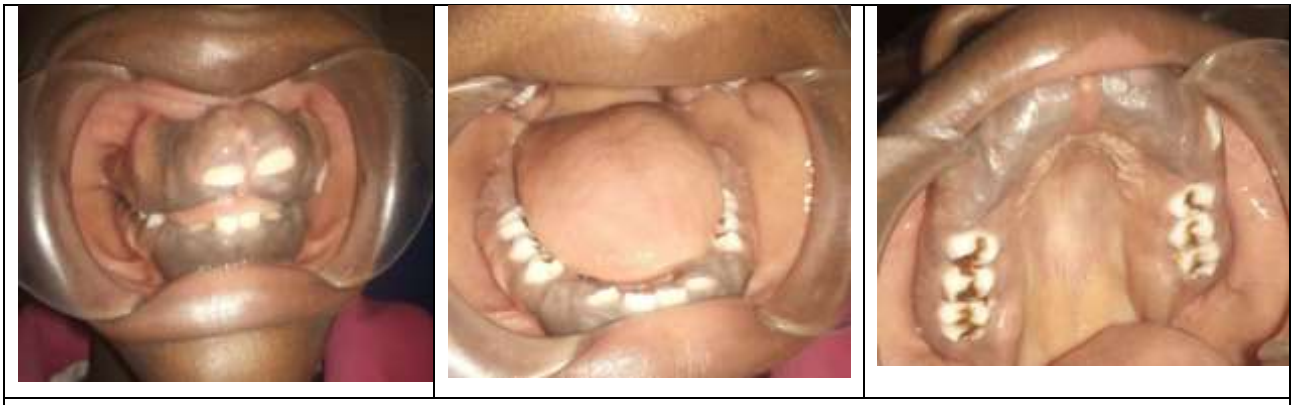
This is a case of a 9-year-old male patient who reported to the Paediatric Dental Clinic of the Dental Center of University of Port Harcourt Teaching Hospital with the complaint of enlargement of gums in all quadrant from birth. The swelling had slowly progressed with age. Swelling was not associated with discomfort and there was no disturbance with mastication. Patient was concerned about the unaesthetic appearance of the gingiva and the resultant pressure from his peers. There was a history of delayed eruption of the primary teeth. He did not have any associated medical conditions, recent hospitalization or syndromic features. There was no history of prolonged use of any hormonal or drug-induced gingival hyperplasia medications and he was apparently healthy. He was referred to the periodontics outpatient clinic for review and subsequent management. After a detailed history, it was found that the patient's younger brother also has the same problem. However, this type of gingival enlargement could not be confirmed in the patient's parent, uncle and grandfather. Also, there were no consanguineous marriages in the family.

General body examination showed no medical abnormalities. Extra oral findings showed a symmetric facial profile with incompetent lip seal. Overjet and overbite were increased. Intraoral examination revealed fair oral hygiene index simplified with uniform gingival enlargement on both labial and palatal sides of the teeth (Figure 1). The swollen gingival was dark in colour, fibrous and firm in consistency with absence of stippling. The contour of the gingival showed the absence of knife edge margins and scalloping. There was no bleeding on probing. The patient had an upper marginal labial frenum and generalized false pockets in the posterior areas of all quadrants. Majority of the teeth surfaces were covered by the gingival enlargement except the incisal/occlusal surfaces. The teeth were normal in appearance and immobile. Clinical examination revealed that mandibular canines, maxillary lateral incisors and right canine were absent. Based on the above findings, we made a provisional diagnosis of generalized hereditary gingival fibromatosis. Panoramic and lateral cephalogram radiographs were advised. The orthopantomogram showed normal architecture of bone with all permanent teeth erupting. Also, blood investigations were within normal limits.

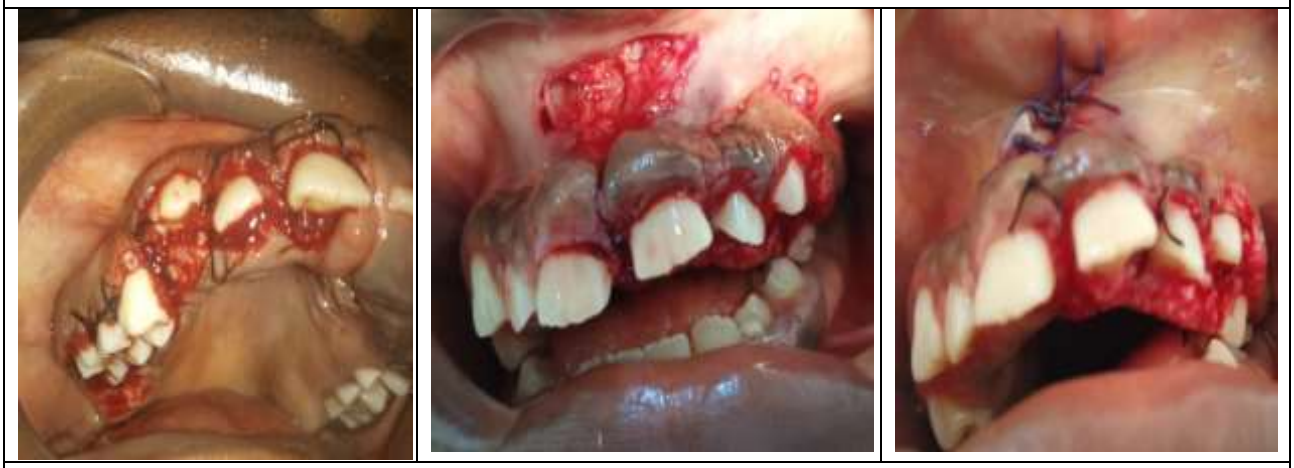
Multidisciplinary approach for treatment was considered. Treatment decided was quadrant-wise gingivectomy/gingivoplasty, frenectomy and orthodontic fixed appliance. The complete treatment procedures (oral prophylaxis and gingivectomy) were explained to the patient and his parents, and before proceeding with the treatment; written informed consent was obtained from the parents and assent from the patient. Full mouth scaling was done after which an external bevel gingivectomy was done in all four quadrants under local anaesthesia. Surgery was planned in three stages after considering the age of the patient and duration of surgery. In the first stage, excessive gingivae in both quadrants of the mandibular arch were excised and the right quadrant of the maxillary

arch was excised 2 weeks later. The left quadrant of the maxillary arch was operated in addition to a frenectomy of the upper labial frenum after 4 weeks of the initial surgery (Figure 2). Postoperative periodontal dressing was applied after each surgery. The patient was placed on antibiotics and analgesics postoperatively; and healing was uneventful. Patient .

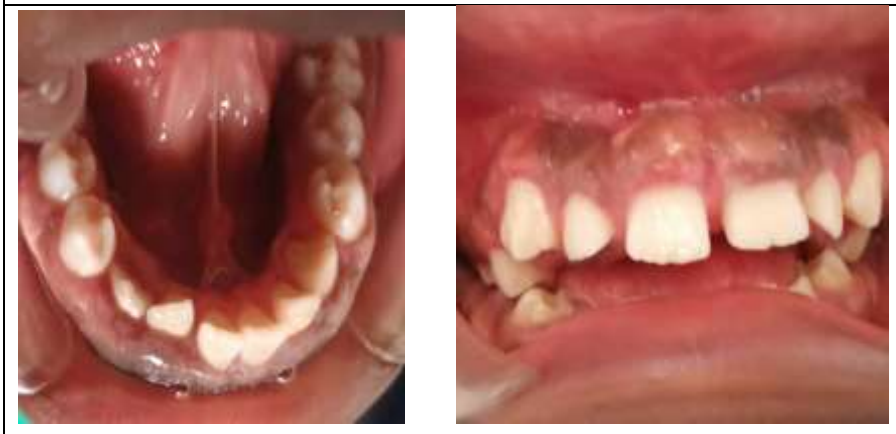
was reviewed after one month (Figure 3), 3 month and 6 months (Figure 4) during which there was no recurrence. Patient was instructed to ensure regular tooth brushing to minimize the effect of inflammation on the gingival tissue Patient is still under review while orthodontic fixed appliance is being planned



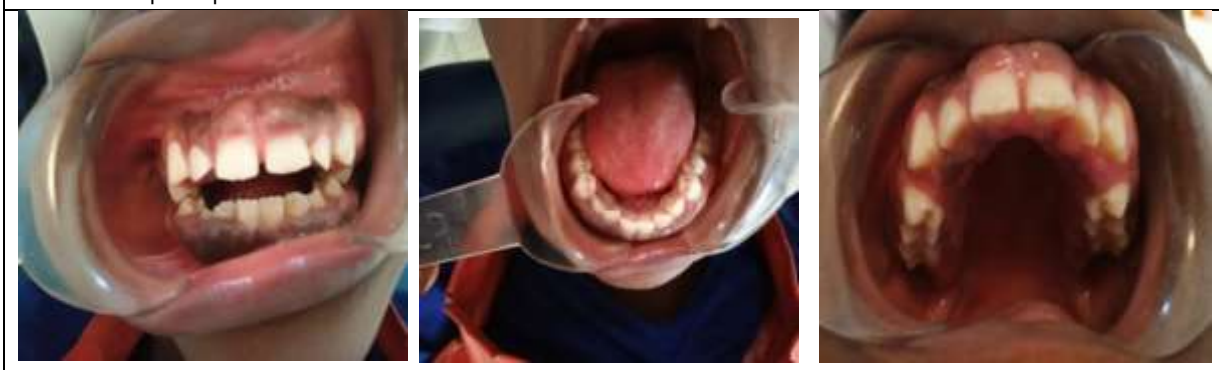
Preoperative photographs



Intraoperative photographs



One month postoperative



6 months post-operative

DISCUSSION

This is a case report of a 9-year-old with non-syndromic hereditary gingival fibromatosis. The diagnosis was based on the clinical presentation and the history of a similar features in a sibling. Literature majorly attribute the condition to genetic factor.[9] This case was probably autosomal recessive since it can manifest by skipping generations, as the affected patient was a child of unaffected carriers.[10-12] The gingival hyperplasia was not related to drug induced gingival enlargement medications in our patient. Likewise, history and examination did not reveal bone deformities, supernumerary teeth, hypertrichosis, and other syndromic features. The laboratory investigations did not reveal any systemic conditions like diabetes mellitus, leukemia and endocrine disorders.

Though gingival fibromatosis usually present before the eruption of the permanent dentition,[6,10,12] this present case started with the eruption of the primary dentition. This resulted in the delay of both primary and permanent dentition and teeth malpositioning. The progressive enlargement of the dense connective tissue can result in malocclusion with features of arch deformity, teeth displacement and migration of teeth.[12,13]

Minimal gingival enlargement had been managed with professional oral prophylaxis and self-maintenance of good oral health to prevent exacerbation of the swelling by dental plaque. Moderate to severe swelling however, dictate the need for removal of excess tissue.[6] Definitive treatment usually consists of surgical excision of the enlarged tissue using the conventional external bevel gingivectomy with gingivoplasty or carbon dioxide laser.[6,9-12] Also, a periodontal flap procedure can be used when there is an associated clinical attachment loss or alveolar bone defects.[13] The need to prevent further malpositioning of teeth necessitates the early intervention in this patient. However, James and Prasad[14] advised that there is a less likelihood of recurrence if surgical excision is delayed until the complete eruption of the permanent dentition.

CONCLUSION

We presented a case of non-syndromic hereditary gingival enlargement, that was managed by gingivectomy of all quadrants with appreciable improvement in aesthetics. Early intervention will ensure the prevention of malocclusion and aesthetic complications. After treatment, regular recall is needed to evaluate the stability of the gingiva and oral hygiene.

Source of Support

Nil.

Conflict of Interest

None declared

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