

CASE REPORT**RARE CASE OF IDIOPATHIC GINGIVAL FIBROMATOSIS AFFECTING PRIMARY DENTITION****Sanaa Ahmed, Zahid Ali**

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Gingival fibromatosis (GF) is a rare condition with an estimated incidence of 1:750,000 in autosomal dominant cases and is supposedly genetic in origin. Gingival fibromatosis (GF) may occur as an isolated finding (Idiopathic gingival fibromatosis or IGF) or in combination with additional clinical problems that is, giving rise to syndromic forms of the disease. It usually is triggered when permanent dentition starts to erupt and can cover crowns of the teeth. Gingival fibromatosis occasionally manifests at birth or affect primary dentition. This enlargement may cause mal-position and diastema. Surgical excision of excessive fibrous tissue is the only treatment but it recurs. We are presenting here a case of 5 year old patient presenting with severe idiopathic gingival fibromatosis covering crowns of primary teeth and causing functional impairment.

Keywords: Idiopathic fibromatosis, diastema, fibromatosis, gingival, hypertrophy, incidence, pseudoanodontia, deciduous

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INTRODUCTION

Also known as gingivomatosis elephantiasis, familial elephantiasis, juvenile hyaline fibromatosis, congenital familial fibromatosis, idiopathic fibromatosis and hereditary gingival hyperplasia¹, Idiopathic Gingival Fibromatosis is a rare, benign, non-hemorrhagic fibrous enlargement of gingival tissues. It has a phenotype frequency of 1:750,000 and the gene frequency is 1 in 350,000² and affects males and females equally³. Commonly seen in children in mixed dentition stage and teenagers³.

Gingival enlargement starts with eruption of permanent teeth but may affect primary dentition. It is not usually present at birth or affect edentulous arch and recedes after extraction. Clinical appearance is varied. Aetiology is still unknown but it is proposed that genetics, local factors and hormones all contribute. We are presenting here a rare and interesting case of idiopathic gingival fibromatosis in a five year old boy.

CASE REPORT

A five year old patient reported to Oral and maxillofacial surgery department of Abbasi Shaheed Hospital with complain of growth covering crowns of teeth showing only the occlusal surface since past 3 years. History of delayed eruption of primary dentition was there. Gradually the fibrous growth covered the crowns of all teeth causing in functional and aesthetic impairment. No history of bleeding or pain in gums. Negative drug and family history with no co-morbid conditions. On general examination no associated abnormalities were found. Mental and physical growth of the patient was normal. On oral examination upper lip incompetency (Figure-1) leading to constantly opened mouth was present. Intra-orally large masses of firm,

dense, resilient, growth covering maxillary (Figure-2) and mandibular (Figure-3) teeth symmetrically was observed leaving only the occlusal surface.

Orthopantomogram (OPG) (Figure-4) was advised which showed primary dentition present and absence of periodontal involvement. All baseline investigations were done which were within normal limit. Since, the gingival hyperplasia was symmetrical, differential diagnosis included drug induced Gingival Hyperplasia⁵, Gingival Fibromatosis⁵, Scurvy⁴, Sarcoidosis⁵, Crohn's disease⁵, Amyloidosis⁵, Orofacial Granulomatosis⁵, Leukaemia infiltrates⁵ and Wegener's Granulomatosis⁵.

Surgical excision with electrocautery under general anaesthesia was done. Post excision (Figure-5) showed primary teeth. Patient was kept on 5 day antibiotic regimen. After observing satisfactory healing for three days, he was discharged and called after one week. Satisfactory healing was observed after 1 week.

Table-1: Syndromes associated with hereditary gingival fibromatosis.

Syndrome	Features
Zimmer-man Laband syndrome	Hypoplastic distal phalanges, hepatosplenomegaly, epilepsy, hypertrichosis, and mental retardation.
Jones Syndrome	Progressive neural deafness.
Klippel-Trenaunay Syndrome	Hemihypertrophy, nevus flammeus, hemangioma, hypertelorism and macrocephaly.
Ramon syndrome	Hypertrichosis, mental retardation, epilepsy, rheumatoid arthritis and diabetes mellitus.
Rutherford syndrome	Unerrupted teeth, corneal dystrophy and mental retardation.
Cross syndrome	Nanophthalmos, micro-cornea, and severe mental retardation.



Figure-1: Extra-oral picture showing incompetent lip due to fibrous growth.



Figure-2: Intra-oral picture showing maxillary teeth covered with gingiva.



Figure-3: Intra-oral picture of mandibular teeth covered with fibrous tissue.



Figure-4: OPG showing primary dentition.



Figure-5: Post-excision picture showing primary teeth of maxilla and mandible.

DISCUSSION

Gingival Fibromatosis is the benign, progressive generalized or nodular enlargement of otherwise normal gingiva. It can be further termed as idiopathic (negative family history) and as hereditary (positive family history). Idiopathic gingival fibromatosis is a rare disease affecting gingival tissues and is classified as localized or generalized affecting arches unilaterally or bilaterally showing nodular or symmetric form. Further the gingival fibromatosis has following forms:⁴

- Isolated HGF.
- Isolated IGF.(Rare in contrast to HGF)
- GF with hypertrichosis.
- GF with hypertrichosis and mental retardation and/or epilepsy.
- GF with mental retardation and/or epilepsy.
- GF with other diseases or part of other syndromes.

It is usually associated with a number of syndromes (Table-1) which were ruled out in our case as the patient didn't show any of the associated features.⁵

Histopathological features are increased IGF and collagen fibres in all directions, associated with young fibroblasts with scarce blood vessels. Inflammatory infiltrate are present in sub-epithelial connective tissue on occasions. Small areas of osseus metaplasia, ulceration of overlying mucosa, deposition of amyloid and odontogenic epithelium can also be present.

Problem starts with eruption of permanent teeth triggering gingiva to grow over the teeth completely resulting in delay in exfoliation of deciduous teeth and eruption of permanent teeth, diastemas

resulting in speech and mastication impairments. On rare occasions it affects the deciduous dentition so severely to require treatment. Case presented above is unique as the gingival enlargement severely affected the deciduous dentition to cause functional and aesthetic impairments.

Diagnosed usually on basis of characteristic appearance and positive family history but on extremely rare occasions sporadic cases are present. In current cases a new differential was made including systemic diseases which were ruled out on following basis:⁵

- Age of patient ruled out puberty induced hyperplasia.
- No history epilepsy or of drug intake like phenytoin, nifedipine, cyclosporine ruled drug induced gingival hyperplasia.
- All base-line investigations were within normal limits which excluded Leukaemia on basis of normal complete blood count report, diabetes on basis of normal random blood test, sarcoidosis on basis of urine detailed report, amyloidosis on basis of normal blood count report, liver function test, urine detail report, and Wegener` granulomatosis on basis of normal erythrocyte sedimentation rate and urine detail report.

After thorough family, drug and medical history, it was evident that it was a sporadic non familial, non-syndromic idiopathic case of gingival fibromatosis. On examination it was classified as of grade-3 severity on basis of following scale:⁶

Grade-0: No gingival enlargement.

Grade-1: Enlargement confined to interdental papilla.

Grade-2: Enlargement involving papilla and marginal gingiva.

Grade-3: Enlargement covering three quarter or more of crown.

Patient was having difficulty in mastication and in speech. Teeth were with diastemas, malpositioning and delayed eruption of deciduous dentition and the growth had displaced upper lip causing lip incompetency. The enlarged gingiva is usually normal in color, consistency and with a minimal tendency to bleed. It presents with nodular appearance and an abundance of stippling of the attached gingiva.

Treatment is usually varies according to severity of enlargement. In mild and moderate cases, it is delayed till after eruption of all of permanent dentition as higher recurrence before it.⁷⁻¹⁴ Minimum

enlargement is managed with scaling and maintenance of good oral hygiene. In moderate and severe cases surgical intervention is required to improve the aesthetic, speech, mastication and hygiene. Surgical removal of gingiva can be achieved by blades, surgical knives, laser or electrosurgery. Recurrence is slow, over 2-14 years. But the overall prognosis is good.¹⁵ We performed gingivectomy under general anaesthesia with electrocautery due to compliance of patient, severity of the case and to minimize bleeding. Patient was discharged 3rd post-operative day after initial satisfactory healing.

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