A Rare Case of non Syndromic Congenital Idiopathic Gingival Fibromatosis: Electrosurgical Management

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Idiopathic gingival fibromatosis (IGF) is a rare, genetically heterogeneous condition that is usually a part of syndrome or, rarely, an isolated disorder. It is characterized by a slowly progressive, non hemorrhagic, fibrous enlargement of keratinized gingiva which usually begins at the time of eruption of permanent dentition, however very few cases involving the primary teeth have been described in literature. Congenital gingival fibromatosis is very rare condition in which the gingival tissues become thickened and erupting teeth remain submerged beneath hyperplastic tissue masses. This case report discusses the rare case of congenital non syndromic idiopathic gingival fibromatosis in a two year old boy who reported with absence of teeth and incompetent lips. Gingivectomy was done using modified microdissection electrocautery needle to remove the excess gingival tissues. Excised tissue has been examined histologically. The patient was followed up for a period of one year and no recurrence was observed.

Keywords- fibromatosis, congenital, electrocautery

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INTRODUCTION

Idiopathic Gingival Fibromatosis (IGF) is an uncommon, benign condition with no specific cause. It is characterized by a gradually progressing, non hemorrhagic, fibrous enlargement of maxillary and mandibular keratinized gingiva. The affected gingival tissue is usually pink in color with exaggerated stippling and is firm and fibrous on palpation. It covers the teeth partially or totally and can be localized or generalized, with a variable degree of severity, and extends over the teeth resulting in extensive pseudo pockets but does not affect the bone[1,2.] This enlargement may extend into the vestibule and floor of mouth, interferes with lip closure, speech and chewing but above all, at the ages at which it appears, it can become a psychological burden and affect the patient's self-esteem[3]. Gingival Fibromatosis (GF) may be hereditary or idiopathic. Although, this hereditary condition exhibits autosomal dominant mode of transmission, an autosomal recessive inheritance has also been reported [4]. It occurs either as an isolated disease or combined with some rare syndromes or chromosomal disorders.

The GF usually begins at the time of eruption of permanent dentition but it's manifestation since birth is a very rare condition in which the gingival tissues become thickened and erupting teeth remain submerged beneath the hyperplastic tissue masses. This case report discusses a rare case of congenital non syndromic idiopathic gingival fibromatosis in a two year old boy.

CASE REPORT

A two year old boy was referred to the Pediatric dentistry department of Post Graduate Institute of Dental Sciences, Rohtak from the department of pediatrics due to unusually swollen gum pads and inability to close lips. Clinical examination revealed bulbous

enlarged maxillary alveolar ridge protruding five millimeter below the upper lip and extending into vestibular area. The lower alveolar ridge was also thicker and enlarged (Figure 1a). In addition, palatal tissue was also enlarged and deep fissures were evident on palate due to overgrown tissue (Figure 1b). The enlarged gingival tissue was non hemorrhagic and firm to hard in consistency. The overgrowth was most severe in maxillary region which resulted in incompetent lips. No primary teeth were visible in the oral cavity. The medical history, family history and drug history was non remarkable. The patient's mother gave history of bulbous gingival tissue since birth. The child weighed five pounds at birth following a full term delivery. The patient's speech was affected and he had not learnt speaking yet although he could masticate semi solid food. No significant inflammation was present around the tissue and the tissue was non tender on palpation. Radiographically, the anterior maxillary teeth were visible on intraoral periapical radiograph (Figure 2a and 2b). The primary diagnosis of congenital idiopathic gingival fibromatosis was made and excisional biopsy was planned to expose the deciduous teeth in both arches. Blood investigations were done and complete hemogram values were within the normal limits. A general physical examination revealed no abnormal findings. The consulting pediatrician was also contacted and did not report any significant findings. Quadrant wise gingivectomy was planned with Electrocautery under General Anesthesia. Written informed consent of parents was obtained. External bevel gingivectomy was done and the enlarged tissue was removed in each quadrant. Excised tissue was sent for histopathological examination. The surgery was performed with Modified Micro-Dissection electrocautery Needle (MMDN). A 21 gauge needle was modified and attached to monopolar electrocautery tip. The tissue to be excised was marked with marker on both the arches (Figure 3a and 3b). The teeth were exposed in both the arches and there was no bleeding as the tissue appeared to be largely fibrous. Wedge of the tissue was excised from palatal aspect of alveolar ridges also. The crowns of incisors, canines and first molars were exposed and spacing was seen between teeth due to excess tissue present around teeth. Patient was kept under observation for three days and the operated site was irrigated with saline thrice daily. Antibiotics and analgesics were prescribed for five days and patient was advised to take liquid diet for first two days and then semisolid diet for another seven days. Chlorhexidine mouthwash was prescribed and proper oral hygiene instructions were given.

Postoperative course was uneventful and healing was satisfactory. Gingival healing was very well appreciated after two weeks of surgery (Figure 4). Regular follow up till one year was done and no recurrence of fibromatous tissue was noted (Figure 5a and 5b). Mastication was also improved and normal age appropriate speech was developing as told by patient's parents.

Histopathological examination of the tissue revealed dense fibrous tissue with mild epithelial hyperplasia, chronic inflammatory cell infiltration around the epithelial region near surface and the rest of the tissue did not exhibit any inflammatory infiltrate (Figure 6).

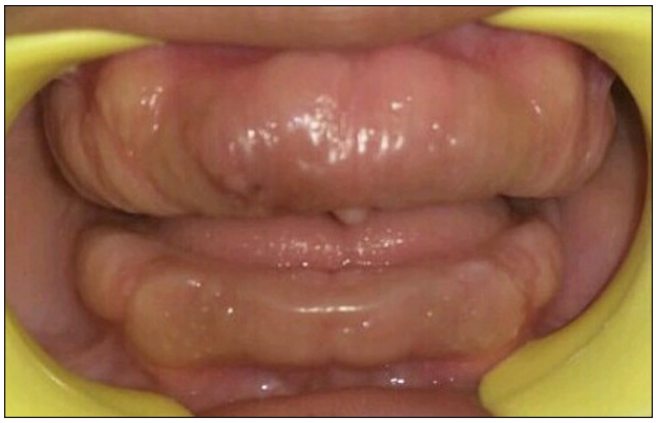

Figure 1A: Preoperative intraoral picture (frontal view) of congenital idiopathic gingival fibromatosis

Figure 1b: Preoperative intraoral picture (maxillary occlusal view) of congenital idiopathic gingival fibromatosis

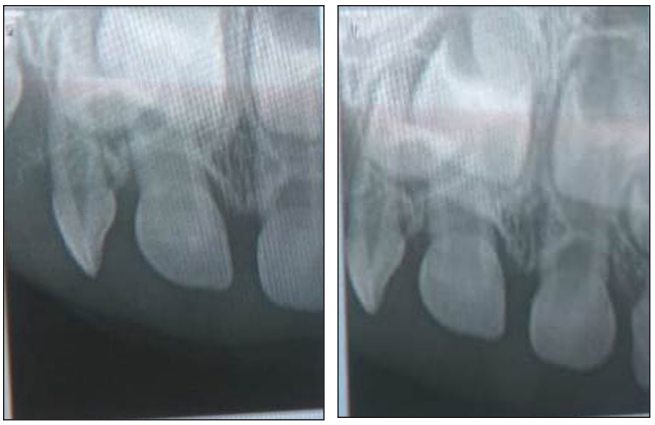

Figure 2: Intraoral periapical radiographS of maxillary central and lateral incisors impacted within gingiva.

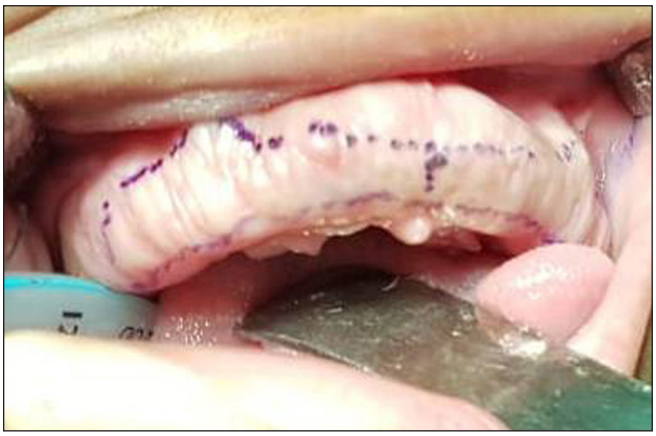

Figure 3a:Demarkated areas in maxillary arch for external bevel incision for gingivectomy

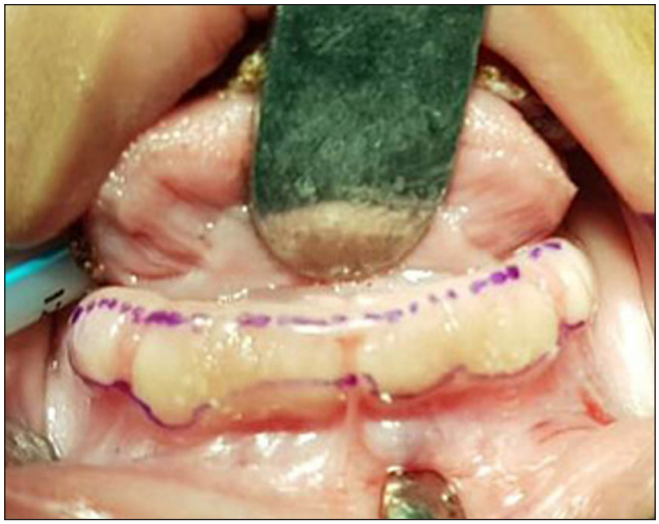

Figure 3b: Demarkated areas in mandibular arch for external bevel incision for gingivectomy

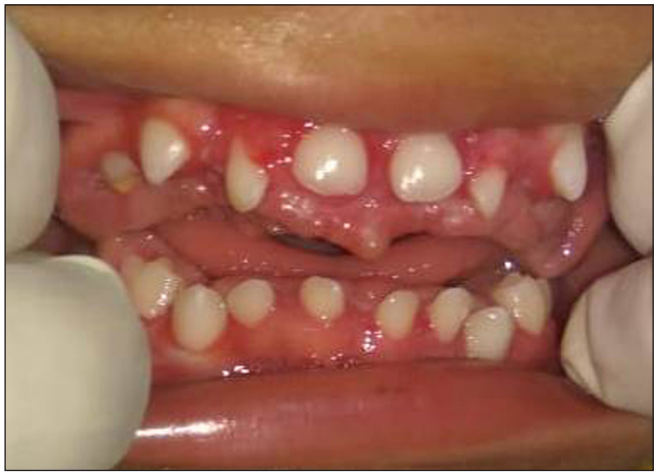

Figure 4: Postoperative intraoral picture after 1 week of surgery

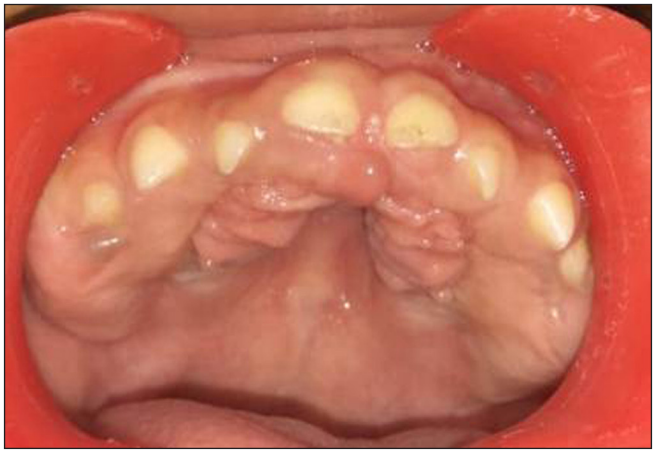

Figure 5a: Postoperative intraoral picture of maxillary arch after 1 year of surgery

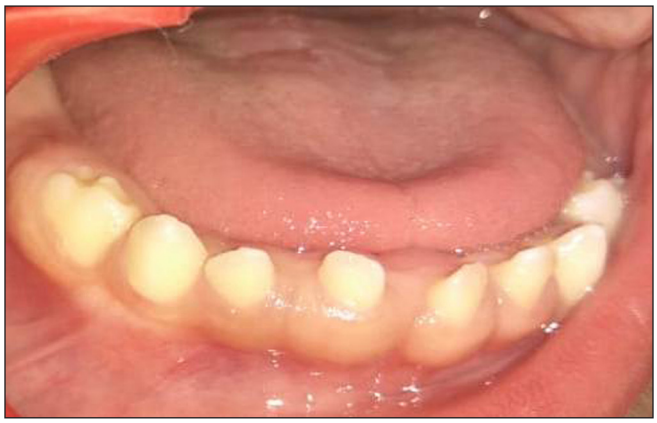

Figure 5b: Postoperative intraoral picture of mandibular arch after 1 year of surgery

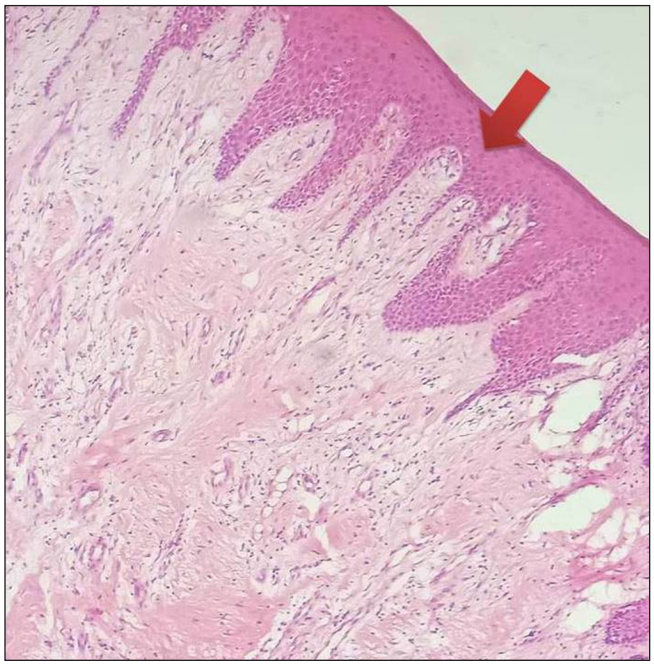

Figure 6: Histopathological image showing hyperplastic epithelium (red arrow) with fibrocollagenous connective tissue.

DISCUSSION

Gingival Fibromatosis (GF) is a discordant group of disorders characterized by progressive enlargement of the gingiva caused by escalation of submucosal connective tissue elements. It can be hereditary or idiopathic. Hereditary Gingival Fibromatosis (HGF) is a rare condition with the prevalence of one per 1,75,000 population and is found equally between males and females[5]. Most cases of hereditary gingival fibromatosis are found to be inherited in an autosomal-dominant manner, although autosomal recessive inheritance has also been reported. Autosomal-dominant form of gingival fibromatosis is usually non syndromic and has been genetically linked to the chromosomes 2p21-p22 and 5q13-q22[6,7]. In modern times, a mutation in the Son of sevenless- 1 (*SOS-1*) gene has been suggested as a possible etiology for isolated (non syndromic) gingival fibromatosis [6].

The gingival enlargement may occur alone or may represent as one of the sign of syndromes like Zimmerman-Laband syndrome

(GF, hypoplastic distal phalanges, hepatosplenomegaly, epilepsy, hypertrichosis, and mental retardation), Jones syndrome (GF and progressive neural deafness), Klippel-Trenaunay syndrome (GF, hemi hypertrophy, Nevus flammeus, hemangioma, hypertelorism, and macrocephaly), Rutherfurd syndrome (GF, unerupted teeth, corneal dystrophy, and mental retardation), Ramon syndrome (GF, hypertrichosis, mental retardation, epilepsy, rheumatoid arthritis, and diabetes mellitus), and Cross syndrome (GF, nanophthalmos,microcornea, and severe mental retardation) [3]. Also, it can occur in association with medications like nifedipine, phenytoin, verapamil, and cyclosporine [8].

Gingival overgrowth can be observed varying in extent and severity. The excess gingival tissue may cover partial or whole crown, resulting in speech alteration, diastema in teeth and cessation of eruption of permanent teeth leading to difficulty in mastication, malocclusion, and unaesthetic appearance. The hyperplastic gingiva is usually pink in color, with firm consistency and exaggerated stippling.

In the present case, gingival enlargement has led to non-eruption of primary teeth, speech difficulty due excessively enlarged gingival tissues and an unaesthetic appearance. Enlargement usually begins with the eruption of the permanent dentition but can develop with the eruption of the deciduous dentition as was observed by Horning *et al* [9] in a three year old girl and rarely, it may present at birth or arise in adulthood. The congenital type of fibromatosis is extremely rare and we have found only four reported cases of congenital gingival fibromatosis after extensive search of literature[10-13]. In our case, the gingival enlargement was present right from the birth and was generalized involving both the arches. Patient had no history of any systemic disease, mental retardation, hypertrichosis, epilepsy, or medication which could contribute to gingival overgrowth. Moreover, family, prenatal, medical, and drug histories were non contributory. The clinical, histopathological features and systemic examination excluded the diagnosis of neoplastic enlargement, Wegener's granulomatosis and acanthosis nigricans. The attached and marginal gingiva was pink and firm in consistency and the absence of inflammation in the present cases ruled out the diagnosis of inflammatory gingival enlargement. Thus, the patient was diagnosed as a case of congenital IGF.

Management of gingival enlargement depends on the age of occurrence and cause of this condition. Various procedures are recommended for removal of fibromatosed gingiva like surgery, electrocautery and carbon dioxide laser. The most effective method for removing large quantities of gingival tissue is the conventional, external bevel gingivectomy especially when there is no attachment loss and all the pocketing is false. In 1941, this procedure was first advocated for drug-induced gingival enlargement[14.] Ramer *et al* [1] advocated quadrant wise gingivectomy with periodontal pack placement for one week, followed by 0.2% chlorhexidine oral rinse twice a day for two weeks after each surgery. In this case, the enlarged gingival tissue was excised with the help of MMDN. For this modification, a standard needle of 21 gauge is separated from its hub using wire twister and taking care not to cause needle stick injury. The shaft of the needle is secured to the monopolar electrocautery tip by adapting it using an insulating tube sleeve. The insulating tubes can be customized by cutting a plastic feeding tube of various lengths. The tube allows for a snug fit of the needle, which means a lesser loss of power and secure placement of the needle with the cautery tip. The needle length and shape could be adjusted for different sites of surgery and the convenience of the operator. It is very cost effective and works equally and efficiently for skin and scalp incisions. It works best at 10-15 Watt (W), lower power usually for infants and deeper tissue dissection. The needle shape and length could be adjusted according to the site of surgery and the convenience of the operator. The modification procedure requires approximately two to four minutes. Therefore authors suggest the use of MMDN considering its advantages and efficacy. The present case was followed up for a period of one year and no recurrence was observed during this period.

The recurrence rate of gingival fibromatosis after surgery is very high. Hence patient should be followed for a considerable period of time as he may require repeated surgeries. Therefore, counselling of a pediatric patient and parents is of paramount importance. Education and motivation of patient is of utmost importance for maintenance of proper oral hygiene and regular recall visits.

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