

Hereditary Gingival Fibromatosis: A Case Report with Review of Literature

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Abstract: Gingival fibromatosis is characterized by localized or generalized fibrous enlargement of the gingivae, mainly around permanent teeth. Gingival fibromatosis affects only the masticatory mucosa and does not extend beyond the muco-gingival junction. This article describes an unusual case of hereditary gingival fibromatosis with delayed eruption of permanent teeth in an 11 year-old girl and her younger sibling. The patient presented with severely enlarged gingival tissues affecting both arches and multiple retained deciduous. Most of the permanent teeth were not erupted. She had no associated symptoms to suggest any syndrome but there was family history of similar disorder with father and paternal aunt. The over retained primary teeth were extracted and gingivectomy as well as gingivoplasty were performed by exposing unerupted permanent teeth in all the quadrants. The excised gingival tissue was sent for histopathological examination to confirm the diagnosis. Two year postoperatively, follow up revealed no recurrence.

Keywords: Fibromatosis Gingivae, Hereditary Gingival Fibromatosis, Delayed Eruption, Gingivectomy, Gingivoplasty.

INTRODUCTION

Hereditary gingival fibromatosis (HGF), also known as elephantiasis gingiva, hereditary gingival hyperplasia is a rare condition (1:750000) [1] which can present as an isolated disorder or more rarely as a syndrome component [2, 3]. This condition is characterized by a slow, progressive enlargement of the maxillary and mandibular gingiva [2]. Males and females are equally affected [3]. Most cases of HGF appear to be inherited in an autosomal-dominant manner [4, 5], although autosomal recessive inheritance has been reported [6]. The hyperplastic gingiva presents with normal color, firm in consistency, and an abundance of stippling of the attached gingiva [6]. Buccal and lingual tissues may be involved in both the maxilla and mandible [2, 3].

The degree of enlargement may vary from mild to severe and may be the same between individuals within the same family [5, 6]. Most cases are seen from birth, but the condition may not be noticed until late childhood till the time of eruption of the deciduous or permanent teeth [2]. This condition may appear as an isolated disorder or associated with other conditions or syndromes such as Zimmerman-Laband, Murray-Puretic-Drescher, Cowden's syndrome, and Cross syndrome [2, 3].

CASE REPORT

An 11-year-old female child attended the Department of Pediatric dentistry with the chief complaint of generalized gingival overgrowth. The patient was present with several retained primary teeth. The permanent teeth were embedded in the overgrowth and left unexposed into the oral cavity. The patient complained about difficulty in swallowing due to lip incompetence as a result of severely enlarged gingiva and also disturbed phonetics. In the treatment, the over retained deciduous teeth were extracted and embedded permanent teeth were exposed by performing gingivectomy and contouring the gingiva with gingivoplasty.

Family History

The patient's family history revealed a hereditary pattern with father and paternal aunt presenting with similar condition and underwent treatment during adolescence. But the clinical condition and treatment details were not clear.

Medical History

There was no significant medical history suggesting any medication that could be associated with gingival hyperplasia and no signs of any other general health problems.

Clinical Examination

The extra oral examination did not reveal any abnormality except the incompetent lips. The intraoral examination revealed generalized, severe gingival hyperplasia involving both buccal and lingual in the maxillary and mandibular arches. The enlarged gingival tissues covered two-thirds or more of the clinical crowns of almost all teeth. The gingiva was pink and firm but the patient's oral hygiene was very poor [Figure 1].



Fig-1: hypertrophied gingiva

Radiographic Examination

Panoramic radiographic examination revealed multiple retained primary teeth and all permanent teeth which are nearing the completion of root formation and erupted through alveolar process but hidden under hyperplastic gingiva (Figure 2).



Fig-2: X-ray showing teeth

Histologic Examination

The sections revealed fibrous connective tissue and overlying stratified squamous epithelium. The connective tissue shows numerous collagen bundles arranged haphazardly with few interspersed fibroblasts.

Connective tissue also shows few endothelial lined vascular spaces and deeper down at one focus chronic inflammatory cells. The epithelium is of stratified squamous type showing acanthosis and elongated rete ridges [Figure 3].



Fig-3: Histology showing dense fibrous tissue

Surgical Treatment

Gingivectomy was performed quadrant-wise. In the 1st quadrant 54 was extracted and 12, 13 and 14 were exposed by excising the overgrowth. In the 2nd quadrant 21, 22, 23 and 24 were exposed by excising the overgrowth. In the 3rd quadrant 74 was extracted followed by excision of overgrowth and gingivoplasty. In the 4th quadrant 84 was extracted and 41, 42 and 43 were exposed. The case was followed for 2^{1/2} years [Figure 4].



Fig-4: Post surgery

DISCUSSION

Gingival enlargement can be caused by inflammation, systemic diseases such as leukemic infiltration, and use of medication such as cyclosporine, phenytoin, or nifedipine [7]. HGF is rare condition and synonyms of GF include Elephantiasis gingivae, Congenital hypertrophy of gingiva, Fibromatosis gingivae, Gigantism of gingiva, Symmetric fibroma of

palate, Congenital macrogingivae, Hereditary gingiva hyperplasia and Hypertrophic gingiva [8, 9].

GF can occur as an isolated condition or be associated with other diseases and syndromes and the involvement can be generalized or localized. Accordingly they are classified [8]:

1. Isolated Hereditary GF(isolated HGF):a) Generalized b) Localized
2. Isolated Idiopathic GF(Isolated IGF):a) Generalized b) Localized
3. GF with hypertrichosis
4. GF with hypertrichosis and mental retardation and/or epilepsy.
5. GF with mental retardation and/or epilepsy.

GF associated with other diseases with formation of syndromes. Isolated HGF occurs more often in the younger age groups than isolated IGF. HGF has a tendency to occur more frequently as a generalized type in comparison to IGF. The ratio of generalized-to-localized types in HGF and IGF was 15.2:1 and 1.6:1 respectively. In this case a diagnosis of HGF was made because no medication was taken could be associated with gingival hyperplasia. The history of familial inheritance was clearly evident as father and paternal aunt were treated for similar condition. The younger sibling was also seen with mild form of gingival overgrowth. Though, there will not be any untoward sequelae with gingival hyperplasia, surgical intervention is required when enlarged tissues impair esthetics and function and act as a barrier to the eruption of teeth.

The syndromes associated with GF include Murray-Pruretic Drescher Syndrome (multiple hyaline fibromas); Rutherford's Syndrome (corneal dystrophy); Laband Syndrome (ear, nose, bone and nail defects with hepatosplenomegaly); Jones' Syndrome (progressive deafness); Cross Syndrome (microphthalmia, mental retardation, athetosis and hypopigmentation) [14]. Cornelia de Lange Syndrome (primordial growth deficiency, severe mental retardation, anomalies of the extremities and a characteristic face) [8] and Ramon's Syndrome (association with cherubism). A syndrome associated with hearing deficiencies, hypertelorism and supernumerary teeth has been reported by Wynne and colleagues [9]. Other associated problems include hypothyroidism, chondrodystrophia and diffuse osteofibromatosis (GF with osteofibrosis) [8].

Fletcher reported, that the enlargement seems to progress rapidly during active eruption and decreases with the end of this stage [1]. Bozzo *et al* [10] and Bittencourt *et al* [11] suggested and implemented a conservative approach in their cases based on the patient's condition and previous reports in the literature. However some authors recommended excision of the excess gingival tissue combined with removal of all teeth in severe cases and suggested that the condition

would not recur if the teeth are extracted [3]. The most widely used surgical approach is the gingivectomy/gingivoplasty [3, 10, 12]. Following surgical excision, the patient would have less postoperative discomfort with reverse bevel incision resulting in minimal cut tissue surface.

CONCLUSION

There is no consensus among authors regarding the timing that surgery should be accomplished in gingival fibromatosis cases. Emerson [13] suggested the best time is after complete eruption of the permanent teeth. But since the permanent teeth were locked under overgrown gingiva, gingivectomy was performed to expose them followed by gingivoplasty. The 2^{1/2} yrs follow up period did not show any recurrence or functional loss.

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