

## A rare case report of hereditary gingival fibromatosis with atrial septal defect

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### Abstract

Hereditary gingival fibromatosis (HGF) is an uncommon condition characterized by an accumulation of extracellular matrix resulting in a fibrotic enlargement of the gingiva. It results in various complications such as drifting of teeth, prolonged retention of primary dentition, diastemata, and poor plaque control. In this case report, we present a case of a 17 year old boy with the presence of HGF with a history of treated atrial septal defect.

**Keywords:** Hereditary gingival fibromatosis, Atrial septal defect, Autosomal dominant, Complications.

### Introduction

Hereditary gingival fibromatosis (HGF) is a rare benign gingival overgrowth with a prevalence of 1 in 750000 and equal distribution in both sexes.<sup>(1-3)</sup> It is also called as familial or idiopathic gingival fibromatosis, and is characterized by a slowly progressive, benign enlargement of keratinized gingiva.<sup>(4,5)</sup> HGF first reported by Goddard and Gross in 1856.<sup>(6)</sup> HGF has an autosomal dominant mode of inheritance; however, an autosomal recessive mode of inheritance has also occasionally been reported.<sup>(7)</sup>

Clinically it presents as a firm, dense, resilient, insensitive fibrous tissue that covers the alveolar ridges and extend over the teeth, resulting in extensive pseudo pockets. The color may be normal or erythematous if inflamed. Histologically, it is characterized by densely arranged collagen bundles, numerous fibroblasts, and connective tissue that is avascular along with well-structured epithelium with elongated and thin papillae inserted in fibrous connective tissue.<sup>(8-9)</sup>

In this case report, we present a case of a 17 year male boy who reported to us for the presence of swollen gums.

### Case Report

In this case, a 17 year old male boy along with her sister reported to the Department of Periodontology, Himachal Pradesh Government Dental College and Hospital, Shimla, H.P. with chief complaint of presence of swollen gums and also, for the progressive increase in size of the gums since the past 8 years. A thorough medical history was taken which revealed that the patient was operated upon for the presence of atrial septal defect 4 years back. Since then, the patient was taking no medications for the same. The present health condition and mental health condition was normal. Complete dental examination was performed. Intraoral examination detected a generalized enlargement of the gingival tissue. Gingival enlargement had normal color with a firm and fibrous consistency, and it was most evident in the mandibular and maxillary anterior region.

(Fig. 1) The maxillary lateral incisors were peg shaped and were in the erupting stage along with the maxillary canines. (Fig. 2, 3) Delayed eruption was seen in regard to various teeth. An OPG was advised to the patient to see the presence of full complement of teeth and for the presence of any bone defects. Radiographically, no bone defects were evident. (Fig. 4)



**Fig. 1: Frontal View**



**Fig. 2: Left lateral view**



**Fig. 3: Right lateral view**

Before proceeding for the treatment, clearance was taken with the department of cardiology. Thorough scaling and root planning was done. Mechanical and chemical plaque control instructions were given to the patient. It was decided to proceed for the surgical intervention as the presence of gingival enlargement interfered with normal functions like eating and speech dysfunction of the patient although the patient did not report for the same.

### Discussion

HGF is a rare condition characterized by varying degrees of gingival hyperplasia. It is mainly inherited in an autosomal-dominant manner, although autosomal-recessive inheritance has also been reported. Oral manifestations of HGF can vary from focal sites of gingival hyperplasia to generalized involvement, and the degree of enlargement may vary from mild to severe.<sup>(10)</sup> HGF has been reported as an isolated finding or in association with other features, as part of a syndrome. As such, HGF has been recorded in association with hypertrichosis,<sup>(11)</sup> mental retardation,<sup>(12)</sup> epilepsy,<sup>(13)</sup> progressive sensorineural hearing loss<sup>(14)</sup> and abnormalities of the extremities, particularly of fingers and toes.<sup>(15)</sup> None of the above mentioned symptoms were applicable in the present case report. The patient was diagnosed with the atrial septal defect 4 years back and was operated upon for the same at that time. It is suggested that a mutation in the *SOS1* gene, which encodes a guanine nucleotide exchange factor for ras proteins, is responsible for the etiology of HGF.<sup>(16)</sup> HGF results in many complications such as tooth migration, prolonged retention of primary teeth, diastema, late eruption of permanent teeth. It may cover the dental crowns resulting in abnormal occlusion, inability to close the lips, problems in eating, speech and esthetics. Therefore, owing to the presence of such complications, it necessitates to proceed for the surgical therapy following the phase I periodontal therapy.

Although in our case the surgical intervention was not performed as the patient did not report to us because

of his own personal reasons our intervention definitely helped him to maintain his oral hygiene and make him aware of the future treatment needs for him.

### Conclusion

Hereditary gingival enlargements must be diagnosed in view of the fact that many a times these enlargements are associated with other systemic conditions and hence treatment plan and further management should be done accordingly.

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