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

Deepali Rasila[1], Kanwarjit Asi[2], Ajay Mahajan[3,*]

[1]Junior Resident, [2]Professor, [3]Assistant Professor, Himachal Pradesh Govt. Dental College & Hospital, Shimla

***Corresponding Author:**
Email: drajdent@yahoo.co.in

Abstract

Hereditary gingival fibromatosis (HGF) is an uncommon condition characterized by an accumulation of extracellular matrix resulting in a fibrotic enlargement of the gingival. 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.[1-3] It is also called as familial or idiopathic gingival fibromatosis, and is characterized by a slowly progressive, benign enlargement of keratinized gingival.[4,5] HGF first reported by Goddard and Gross in 1856.[6] HGF has an autosomal dominant mode of inheritance; however, an autosomal recessive mode of inheritance has also occasionally been reported.[7]

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.[8-9]

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 form 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)

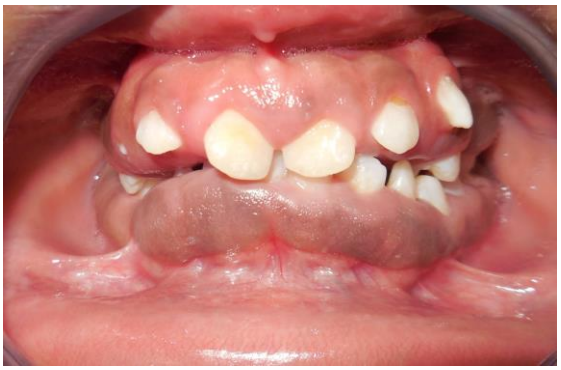

Fig. 1: Frontal View

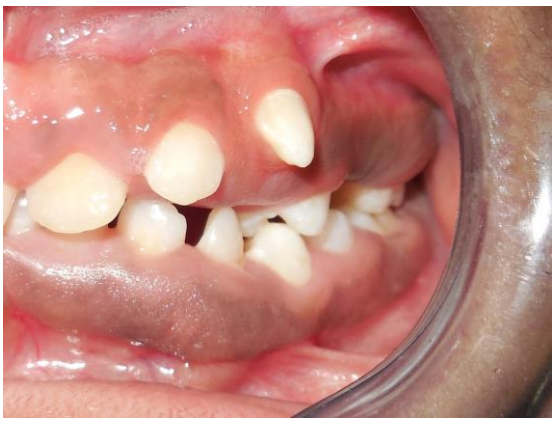

Fig. 2: Left lateral view

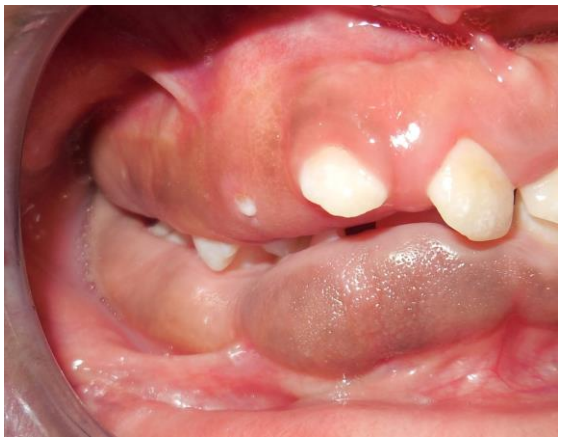

Fig. 3: Right lateral view

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.[10] 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,[11] mental retardation,[12] epilepsy,[13] progressive sensorineural hearing loss[14] and abnormalities of the extremities, particularly of fingers and toes.[15] 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.[16] 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 1 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.

References

1. Nibali L, Medlar A, Stanescu H, Kleta R, Darbar U, Donos N. Linkage analysis confirms heterogeneity of hereditary gingival fibromatosis. Oral Dis 2013;19:100-5.
2. Ramakrishnan T, Kaur M. Multispeciality approach in the management of patient with hereditary gingival fibromatosis: 1-year followup: a case report. Int J Dent 2010;2010:575979.
3. Dhadse PV, Yeltiwar RK, Pandilwar PK, Gosavi SR. Hereditary gingival fibromatosis. J Indian Soc Periodontol 2012;16:606-9.
4. Sengu¨n D, Hatipog˘lu H, Hatipog˘lu MG. Long-term uncontrolled hereditary gingival fibromatosis: A case report. J Contemp Dent Pract 2007;8:90-96.
5. Schulze C, Bensch M, Winterhoff N, Ansorge H, Teifke JP. Gingival fibromatosis (hereditary hyperplastic gingivitis) in a wild European red fox (Vulpes vulpes). Dtsch Tierarztl Wochenschr 2008;115:471-474.
6. Goddard WH, Gross SD. Case of hypertrophy of the gums. Dent Regist West. 1856;9:276–82.
7. Singer SL, Goldblatt J, Hallam LA, Winters JC. Hereditary gingival fibromatosis with a recessive mode of inheritance. Case reports. Aust Dent J 1993;38: 427-432.
8. Araujo CS, Graner E, Almeida OP, Sauk JJ, Coletta RD. Histomorphometric characteristics and expression of epidermal growth factor and its receptor by epithelial cells of normal gingiva and hereditary gingival fibromatosis. J Periodontal Res 2003;38:237- 241.
9. Martelli-Junior HBG, Graner E, Bozzo L, Coletta RD. Microscopic and proliferative comparison of gingival fibroblasts from patients with normal gingiva and with hereditary gingival fibromatosis. Pesqui Odontol Bras 2000;14:123-129.
10. Bozzo L, Machado MA, de Almeida OP, Lopes MA, Coletta RD. Hereditary gingival fibromatosis: Report of three cases. J Clin Pediatr Dent 2000;25:41-46.
11. Horning GM, Fisher JG, Barker BF, Killoy WJ, Lowe JW. Gingival fibromatosis with hypertrichosis. A case report. J Periodontol 1985;56:344-347.
12. Araiche M, Brode H. A case of fibromatosis gingivae. Oral Surg Oral Med Oral Pathol 1959;12:1307-1310.
13. Ramon Y, Berman W, Bubis JJ. Gingival fibromatosis combined with cherubism. Oral Surg Oral Med Oral Pathol 1967;24:435-448.
14. Hartsfield JK Jr., Bixler D, Hazen RH. Gingival fibromatosis with sensorineural hearing loss: An autosomal dominant trait. Am J Med Genet 1985;22: 623-627.
15. Skrinjaric I, Bacic M. Hereditary gingival fibromatosis: Report on three families and dermatoglyphic analysis. J Periodontal Res 1989;24:303-309.
16. Hart TC, Zhang Y, Gorry MC, et al. A mutation in the SOS1 gene causes hereditary gingival fibromatosis type 1. Am J Hum Genet 2002;70:943-954.