Non syndromic gingival fibromatosis in a mild mental retardation child

MAHESH K. DUDDU, RADHIKA MUPPA, G. S. PRASAD REDDY1, P. VEERENDRA NATH REDDY2

Abstract

Gingival fibromatosis is a benign oral condition characterized by enlargement of gingival tissues. It usually develops as an isolated disorder but can be one of the features of a syndrome. This case report is of a 5-year-old male with severe gingival hyperplasia and mild mental retardation which was complicated by open bite, abnormal occlusion, open lip posture, and disabilities associated with mastication and speech. Full mouth gingivectomy in single sitting under general anesthesia was done with electrocautery.

Keywords: Electrocautery, idiopathic gingival fibromatosis, mental retardation, SOS-1 gene

Introduction

Gingival fibromatosis (GF) is a heterogeneous group of disorders characterized by progressive enlargement of the gingiva caused by an increase in sub-mucosal connective tissue elements.[1] Idiopathic gingival fibromatosis (IGF) is a rare, benign, asymptomatic, nonhemorrhagic, and nonexudative proliferative fibrous lesion of the gingival tissue. [1] It occurs either as an isolated disease or in combination with rare syndromes or chromosome disorders. Associated syndromes are Zimmerman-Laband syndrome, Murray-Puretic-Drescher syndrome, Rutherford syndrome, Cowden syndrome, and Cross syndrome.[2] The condition is also reported to be associated with deficiency of growth hormone due to lack of growth hormone release factor.[3] Synonyms of IGF include elephantiasis gingivae, congenital hypertrophy of the gingiva, fibromatosis gingivae, gigantism of the gingiva, symmetric fibroma of the palate, congenital macrogingivae, hereditary gingival hyperplasia, and hypertrophic gingiva. [1] It has recently been found that there are qualitative and quantitative differences in transforming growth factor beta isoform (TGF-β) and receptor expression by fibroblasts in gingival overgrowth and this may contribute to the pathogenesis of the disease. This rare case report describes a 5-year-old male child with mild mental retardation (MR) associated with gingival fibromatosis which was affecting his mastication and speech.

Case Report

A 5-year-old boy with mild MR reported to the Department of Pedodontics with the chief complaint of painful and swollen gums. Patient’s mother also complained of teeth not seen due to overgrowth of gums. Family and postnatal history was non-contributory and the patient did not have any history of epilepsy and drug intake. Developmental milestones of the child were delayed. On general examination there were no signs of hypertrichosis. Extra oral examination revealed incompetent lips [Figure 1]. Intraoral examination revealed generalized enlargement of the gingiva involving both the maxillary and mandibular arches [Figure 2]. The other associated problems were difficulty in speech, mastication, and swallowing. Based on history and clinical features, the case was provisionally diagnosed as IGF.

Investigations

Panoramic radiograph revealed a full complement of teeth and migration of lateral incisors in the maxillary anterior region [Figure 3]. Results of complete hemogram and thyroid tests (T3, T4, TSH) were within the physiological limits.

Treatment done

Consent was obtained. Full mouth gingivectomy by electrocautery was performed under general anesthesia to restore the patient’s esthetics and functional/masticatory needs [Figures 4 and 5]. The patient was prescribed a 0.12% chlorhexidine mouthwash for 2 weeks to maintain oral hygiene and was recalled after 15 days for a checkup. Post-operative examination revealed healthy gingiva which was in healing stage [Figures 6].
**Histopathological investigations**

Gingiva excised from the buccal and interdental areas using a B.P. blade was immediately fixed in a formalin solution and sent for histopathological examination. An excisional biopsy revealed keratinized stratified squamous epithelium exhibiting pseudoepitheliomatous interconnecting epithelial ridges [Figure 7]. The connective tissue was hyperplastic with extensive thick collagen bundles, few fibroblasts, and blood vessels. Inflammatory cells were not prominent except at the areas subjacent to epithelium. On the basis of the medical, family, drug histories and the clinical, investigative findings, the present case was diagnosed as IGF.

**Discussion**

IGF can be one feature of several multisystem syndromes, occasionally associated with severe medical problems and has a variety of psychosocial consequences for individuals. Hereditary gingival fibromatosis is genetically heterogeneous. It can occur either as autosomal dominant (common) or recessive form and usually a positive family history is always present. In IGF, no causative agent can be identified and a family history is lacking. If the inheritance is autosomal dominant, then the phenotypic frequency is 1 in 750,000 people and the gene frequency is 1 in 350,000. Although the cause of IGF is unknown, there appears to be a genetic predisposition. The condition may manifest as an autosomal dominant or less commonly as autosomal recessive. Autosomal dominant nonsyndromic forms have been genetically linked to the chromosome 2p21-p22 and Sq13-q22. It is possible that isolated GF may result from a single gene mutation while syndromic forms may result from alterations of multiple genes or perhaps the gene dosage effect. Recently, a mutation in son of sevenless-1 (SOS-1) gene has been held responsible for this rare hereditary condition. SOS-1 gene codes for a protein that activates the RAS pathway, which signals cell growth. The variability may be due to the expression of a common gene mutation, allelic mutations, or nonallelic mutations. HGF, as a part of a syndrome and not as an idiopathic form, is related to hypertrichosis and mental retardation. The coexistence of GF and mental retardation does not comprise a distinct syndrome, but provides direct evidence of genetic heterogeneity for HGF.

Our case presented with symptoms and clinical findings resembling GF and there was no family history. Hence, the case was diagnosed as IGF, which was probably caused by variable gene expression or a gene mutation. The gingival
Duddu, et al.: Non syndromic gingival fibromatosis in a mild MR child

Enlargement usually begins at the time of eruption of the permanent dentition, but can develop with the eruption of the primary dentition. The most common side-effects related to the gingival lesions are diastemas, malpositioning of the teeth, and prolonged retention of primary teeth, as well as delayed eruption of permanent teeth.

Gingival fibromatosis can be controlled with varying degrees of success. When the enlargement is minimal, oral prophylaxis and home care may be sufficient to maintain good oral health. As the excess tissue increases, appearance and functional impairment dictate the need for surgical intervention. Many techniques have been used for the excision of the enlarged gingival tissues including an external or internal bevel gingivectomy; an apically positioned flap; electrocautery and CO₂ laser. Because of the severity of the involvement in this case and the patients inability to cooperate, gingivectomy using the electrocautery technique under general anesthesia was the treatment of choice.

Early diagnosis of gingival fibromatosis is imperative for maintaining optimum gingival health, monitoring the eruption of permanent teeth, improving oral function, aesthetics, and reducing psychological effects. Long-term follow up is necessary in patients with gingival fibromatosis because of the risk of recurrence, which is not predictable. Treatment of such cases involves close collaboration between the practitioners of different dental specialties.

Conclusion

Gingival overgrowth as a clinical characteristic of idiopathic GF is compatible with life, but the related dental complications worsen patient’s adaptation in daily emotional, social, and functional requirements. Management of such cases is based on a multidisciplinary approach and psychological counseling plays a very important role in patient management. Although the appropriate time for treatment is typically found to be after the eruption of permanent teeth, when recurrence is found to be minimal, surgical intervention should not be delayed. However, further research is needed to establish a syndromic association between gingival fibromatosis, MR, and epilepsy which is done based on genetic evaluation and linkage studies.

References

1. Pappachan B, Narayan J, Nayak A. Idiopathic gingival fibromatosis: A neglected case. Indian J Radiol Imaging 2002;12:335-8.
2. Gorlin RJ, Pinborg JJ, Cohen MM. Syndromes of the Head and Neck. 2nd ed. New York: McGraw-Hill Co; 1976.
3. Oikarinen K, Salo T, Kääriä ML, Lahtela P, Altonen M. Hereditary gingival fibromatosis associated with growth hormone deficiency. Br J Oral Maxillofac Surg 1990;28:335-9.
4. Breen GH, Addante R, Black CC. Early onset of hereditary gingival fibromatosis in a 28-month old. Pediatr Dent 2009;31:286-8.
5. Hart TC, Pallos D, Bozzo L, Almeida OP, Marazita ML, O’Connell JR, et al. Evidence of genetic heterogeneity for hereditary gingival fibromatosis. J Dent Res 2000;79:1758-64.
6. Xiao S, Bu L, Zhu L, Zheng G, Yang M, Qian M, et al. A new locus
for hereditary gingival fibromatosis (GINGF2) maps to 5q13-q22. Genomics 2001;74:180-5.
7. Shashi V, Pallos D, Pettenati MJ, Cortelli JR, Fryns JP, von Kap- Herr C, et al. Genetic heterogeneity of gingival fibromatosis on chromosome 2p. J Med Genet 1999;36:683-6.
8. Bittencourt LP, Campos V, Moliterno LF, Ribeiro DP, Sampaio RK. Hereditary gingival fibromatosis: Review of the literature and a case report. Quintessence Int 2000;31:415-8.
9. Ramer M, Marrone J, Stahl B, Burakoff R. Hereditary gingival fibromatosis: Identification, treatment and control. J Am Dent Assoc 1996;127:493-5.

How to cite this article: Duddu MK, Muppa R, Prasad Reddy GS, Reddy PV. Non syndromic gingival fibromatosis in a mild MR child. Contemp Clin Dent 2012;3:S206-9.

Source of Support: Nil. Conflict of Interest: None declared.