CASE REPORT

Rare case report of idiopathic gingival fibromatosis in childhood and its management

Morankar Rahul,[✉] Krishan Gauba, Nitin Gorwade, Aman Kumar

Oral Health Sciences Center,
Post Graduate Institute
of Medical Education and
Research, Chandigarh, India

Correspondence to
Dr Morankar Rahul,
captainrahul88@gmail.com

Accepted 8 January 2019

SUMMARY

Idiopathic gingival fibromatosis (GF), also known as gingivomatosis, is a rare condition in childhood, with an unknown aetiology. The oral manifestations of the condition are varied and depend on the severity and age of involvement. This paper describes the case of a 5-year-old male child with extensive gingival enlargement covering almost all the maxillary and mandibular teeth resulting in difficulty with speech, mastication and poor aesthetics. Clinical and radiographic examination along with haematological investigations ruled out any systemic association. The case was managed with conventional scalpel blade surgery along with electrocautery under general anaesthesia yielding good results without any recurrence after a 12-month follow-up. The results revealed that the oral manifestations of GF depend on its severity and the age of onset. Timely intervention can help to prevent associated complications in a growing child.

BACKGROUND

Gingival enlargement can occur as an isolated pathology or can be associated with various syndromes. The aetiological factors considered responsible for this condition include heredity, hormonal fluctuations (pregnancy related, growth-hormone associated), drug-induced, inflammatory, systemic (associated with leukaemia, neurofibromatosis), idiopathic and those associated with the syndromes.^{1,2} Idiopathic enlargement of gingiva, which has no definite cause, is rare and affects only one in 750 000 individuals.¹ It is also known as gingivomatosis or fibromatosis.³ Clinically, the hyperplastic gingiva, is usually firm, pale pink in colour and leathery in consistency. It presents a characteristic pebbled surface.⁴ Histologically, it shows hyperplastic epithelium with hyperkeratosis and elongation of rete ridges. There is increased thickening of collagen bundles in the connective tissue which results in an increase in the tissue mass.⁵ Oral manifestations depend on the severity and age of involvement as mild cases causing minimal discomfort may go unnoticed whereas advanced cases with generalised involvement in children can lead to over-retained primary teeth, delayed eruption of permanent teeth, tooth displacement, inability to maintain oral hygiene, malocclusion, poor mastication and can also affect speech and aesthetics.⁶

This paper presents oral manifestations of a rare case of childhood idiopathic gingival fibromatosis

(GF) along with its surgical management and follow-up outcome.

CASE PRESENTATION

A 5-year-old male child reported to the Paediatric Dentistry Unit, Oral Health Sciences Centre, PGIMER, Chandigarh with swollen upper and lower gums. The gingival enlargement was extensive, involving almost all maxillary and mandibular teeth. History revealed, that this problem was present for the last 2 years resulting in difficulty with speech and mastication and causing poor aesthetics. It also resulted in inadequate lip seal and poor dental articulation. The patient had no history of seizures, drug intake, fever, weight loss or any other physical or mental disorder. There was no family history of a similar illness. On extraoral examination the patient had a convex profile and incompetent everted lips due to gingival mass protruded between upper and lower lips (*figure 1A*). Intraoral examination revealed gingival enlargement to be generalised, diffused, pale pink in colour, firm and fibrous in consistency involving the upper and lower arches. This gingival enlargement extended almost up to the incisal/occlusal third of teeth making them barely visible (*figure 1B–D*). An intraoral examination was also carried out for linear ulcerations, spontaneous gingival bleeding, mucosal pallor, cobblestone or corrugated appearance of buccal/labial mucosa and presence of petechiae but no such findings were noticed in the present case.

Figure 1 Preoperative photographs. (A) Extraoral view with incompetent lips. (B) Frontal view. (C) Maxillary occlusal view. (D) Mandibular occlusal view.

© BMJ Publishing Group Limited 2019. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: Rahul M, Gauba K, Gorwade N, et al. *BMJ Case Rep* 2019;**12**:e227942. doi:10.1136/bcr-2018-227942

No other mucosal anomalies were evident. Oral hygiene was fair with scarce plaque, no calculus or any other inflammatory component.

INVESTIGATIONS

Haematological investigations including complete blood count, differential leucocyte count, serum iron and folate levels, erythrocyte sedimentation rate were carried out to rule out any systemic involvement and results were within normal limits. Mantoux and sputum tests were negative for tuberculosis and so was the chest X-ray. Cone beam CT (CBCT) revealed no abnormal bony contour or alveolar bone loss. Consultation was also sought from the department of endocrinology to rule out diabetes mellitus and hypothyroidism but there were no abnormal endocrine findings leading to the present condition.

DIFFERENTIAL DIAGNOSIS

Chronic inflammatory gingival enlargement,⁷ drug-induced gingival overgrowth,^{7,8} familial GF,^{9,10} scurvy,¹¹ plasma cell gingivitis,¹² Crohn's disease,^{13,14} Wegener's granulomatosis,¹⁵ juvenile hyaline fibromatosis,¹⁶ leukaemic gingival enlargement,¹⁷ sickle cell disease,¹⁸ primary peripheral T-cell lymphoma¹⁹ were considered in the differential diagnosis. The clinical features of these conditions pertaining to the gingival enlargement are described in [table 1](#).

TREATMENT

Gingivectomy was performed under general anaesthesia. External bevel incision was made to achieve better gingival contour and aesthetics. Incisions on facial and lingual gingival surfaces were made using Kirkland gingivectomy knives whereas for interdental areas Orban periodontal knives were used ([figure 2A–C](#)). Electrocautery was used to achieve haemostasis as well as to reshape the gingival tissue. The wound was covered with a Coe-Pak surgical dressing to avoid postoperative pain and discomfort. Proper instructions were given to the patient and parents regarding brushing and maintenance of oral hygiene.

The excised gingival tissue, which was sent for histopathological examination, revealed hyperplastic gingival epithelium covered with stratified squamous epithelium with keratinisation. There was elongation and interlacing of rete ridges. The subepithelium showed extensive fibrosis and focal lymphoplasmacytic inflammation more prominent in the perivascular region ([figure 3A,B](#)). There was no evidence of dysplasia or malignancy present. Findings were consistent with idiopathic GF.

OUTCOME AND FOLLOW-UP

A postoperative review after a month revealed complete wound epithelisation. The patient was followed-up at every 3 months. An intraoral examination after 12 months revealed competent lips with good dental occlusion and there was no tendency toward further enlargement ([figure 4A–D](#)).

DISCUSSION

Gingival hyperplasia in children is less common and is mostly associated with poor oral hygiene and the use of drugs. The granulomatous diseases (Wegener's granulomatosis, Crohn's disease, sarcoidosis, tuberculosis), malignancies (acute myeloid leukaemia) and hereditary gingival hyperplasia also should be taken into consideration in the differential diagnosis of gingival overgrowth in children.^{20,21} According to Gagliano *et al* there are different mechanisms of gingival overgrowth for different aetiologies which include increased proliferation of resident tissue

Table 1 Differential diagnoses of gingival enlargement and their characteristic features

Differential diagnosis	Clinical presentation
Chronic inflammatory gingival enlargement ⁷	<ul style="list-style-type: none"> ▶ Occur as an inflammatory response to local irritants like plaque, calculus, ill-fitting prosthesis, orthodontic brackets and so on. ▶ Characterised by bluish or deep red gingiva, frequently friable, soft and bleeds easily. ▶ Occasionally may present as a firm, resilient, pink and fibrotic enlargement usually resolves with effective oral hygiene measures.
Drug-induced gingival overgrowth ^{7,8}	<ul style="list-style-type: none"> ▶ Signs and symptoms generally appear within 2–4 months after initiation of drug intake. ▶ Characterised by mulberry shape, firm, pink and resilient gingiva with minute lobulations and no bleeding on probing. ▶ Involvement is more prominent in maxillary and mandibular anterior region. ▶ After secondary infection, it may show features of inflammatory enlargement.
Familial gingival fibromatosis ^{9,10}	<ul style="list-style-type: none"> ▶ Diagnosis is made by a positive family history of gingival enlargement. ▶ Begin with the eruption of primary or permanent dentition. ▶ Usually develop during childhood, although some cases may not become evident until adulthood. ▶ Condition present as a slow growing generalised or occasionally localised non-tender, firm, pale pink enlargement of gingiva.
Scurvy ¹¹	<ul style="list-style-type: none"> ▶ Gingival enlargement is usually bluish red, soft and friable with a smooth, shiny surface. ▶ Bleeding may occur spontaneously or on slight irritation. ▶ Surface necrosis with pseudomembrane formation is also frequently seen.
Plasma cell gingivitis ¹²	<ul style="list-style-type: none"> ▶ It is considered to be a hypersensitivity reaction to allergens. ▶ Gingiva is reddish in colour; almost involve complete attached gingiva, slightly granular surface appearance is typical. ▶ Patient usually experience burning sensation on eating hot and spicy food.
Crohn's disease ^{13,14}	<ul style="list-style-type: none"> ▶ Gingiva becomes pink, firm and almost leathery in consistency, with a characteristic minutely pebbled surface. ▶ Usually associated with swelling of lips, bowel disorders, fever and ulcers.
Wegener's Granulomatosis ¹⁵	<ul style="list-style-type: none"> ▶ Strawberry gingivitis with reddish-purple exophytic gingival swelling and patchial haemorrhage. ▶ Oral lesions could persist for a long time before multiorgan involvement.
Juvenile hyaline fibromatosis ¹⁶	<ul style="list-style-type: none"> ▶ Gingival hypertrophy with nodular lesions. ▶ Multiorgan involvement with mucocutaneous nodules in the neck, elbow, knees, shins and ankles may be seen.

Continued

Table 1 Continued

Differential diagnosis	Clinical presentation
Leukaemia ¹⁷	<ul style="list-style-type: none"> ▶ Clinically it may mimic inflammatory enlargement. ▶ Other associated features could be oral ulceration, spontaneous gingival bleeding, petechiae, mucosal pallor, herpetic infections and candidiasis. ▶ Rarely, uncommon features such numbness in chin and/or tooth pain is present.
Sickle cell disease ¹⁸	<ul style="list-style-type: none"> ▶ Oral manifestations include mucosal pallor, delayed tooth eruption, disorders of enamel and dentine mineralization, changes in superficial cells of the tongue, malocclusion, hypercementosis and a degree of periodontitis which is unusual in children. ▶ Other features include midfacial overgrowth, facial swelling and osteonecrosis.
Primary peripheral T-cell lymphoma ¹⁹	<ul style="list-style-type: none"> ▶ Primary sites of involvement include palate, gingiva, tongue, buccal mucosa, floor of mouth and lips. ▶ Gingival swelling is usually painless and progressive in nature and may be associated with symptoms such as fever, fatigue and difficulty in breathing on severe exhaustion. ▶ Marginal and interdental gingiva become erythematous, bulbous and tender associated with bleeding on probing.

fibroblasts, reduction in matrix metalloproteinase synthesis (MMP-1 and MMP-2) with inhibitory effect on extracellular matrix degradation, increased production of type-I collagen, formation of heat-shock protein 47 (hsp47) and components of extracellular matrix.²²

In the present case report, the child had no signs of any genetic disorder, the enlargement was not associated with pregnancy or puberty, there was no family history of gingival hyperplasia, no evidence of medication associated gingival enlargement and no known systemic condition associated with gingival hyperplasia. Clinical appearance of bilateral non-inflammatory uniform gingival enlargement without any contributory factors led to a diagnosis of a generalised isolated idiopathic gingival fibromatosis. The generalised form is more common in males compared with females in whom the localised form is more prevalent. It has been postulated that an idiopathic GF may manifest due to a

Figure 3 Histopathology of excised gingival tissue. (A) Stratified squamous epithelium with hyperplasia and interlacing rete ridges. Subepithelial tissue shows marked fibrosis (×50). (B) Mild perivascular lymphomononuclear cell infiltrate (×400).

congenital/hereditary cause but an exact aetiology of the condition is still unknown.

GF is a progressive enlargement of the gingiva due to an increase in the size of submucosal collagenous connective tissue.^{23 24} Despite what the name suggests GF is not related to soft tissue fibromatosis that occur elsewhere in the body. Gingiva is typically normal in colour with a firm consistency. It can progress to cover the crowns of teeth and cause retention of primary dentition and tooth impaction. GF often becomes clinically evident with the eruption of primary or permanent teeth, despite the normal appearance of gingival tissue at birth.²⁵ Most of the patients present with difficulty in mastication and/or phonation. Some may have difficulty in achieving adequate oral hygiene. The mean age of presentation is 26 years (range, 10–65 years). The gingival enlargement usually involves both maxillary and mandibular arches. It is gradual in onset and develops slowly over time.²⁶

The child in the present case report is a 5-year-old boy and had a history of gingival enlargement for the last 2 years. As all the primary teeth erupt around the age of 30 months, gingival enlargement had not affected the eruption of any of the primary teeth in the present case. The permanent first molars and mandibular central incisors start erupting at the age of 6 years. Therefore, a surgery before the eruption of permanent teeth may help to prevent complications associated with gingival overgrowth. The recurrence rate of this condition may show variation among different families or among different members of the same family. The reason for this variation is still unknown. The present case was managed with external bevel gingivectomy resulting in significant improvement in mastication, speech and facial appearance. No recurrence was seen in the present case at the 12-month follow-up. Kavvadia *et al*²⁷ reported a case of diffuse idiopathic GF in an 11-year-old boy. It was managed with surgery and no recurrence of the overgrowth was seen after 30

Figure 2 Intraoperative photographs. (A) External bevel gingivectomy. (B) Immediate postoperative view. (C) Excised gingival tissue.

Figure 4 Twelve-month follow-up photographs. (A) Extraoral view with competent lips. (B) Frontal view. (C) Maxillary occlusal view. (D) Mandibular occlusal view.

months.²⁷ Kelekis-Cholakis *et al*²⁸ presented a case of hereditary GF in a 13-year-old girl. Treatment composed of four-quadrant gingivectomy followed by an orthodontic treatment. Complete recurrence was noted 3 years after the initial surgery. This was followed by another full-mouth gingivectomy that did not reveal any signs of recurrence after a year of follow-up.²⁸

The management of gingival overgrowth varies according to its aetiology but overall the maintenance of good oral hygiene and periodic surgical excision yield good results. It is recommended that one should wait for repeat surgery until the eruption of all the permanent teeth.²⁹ Considering the high recurrence rate in cases with GF and the need for repeated surgery, the patient should be kept on regular follow-up.

Patient's perspective

Being a father, it was disturbing for me to see my son like this as no other family member was affected with the similar condition. We approached PGIMER Chandigarh where surgery was done and there is a significant improvement in aesthetics, speech and mastication compared with what it was before.

Learning points

- Idiopathic gingival fibromatosis is a very rare condition in childhood.
- Oral manifestations of the condition depend on its severity and the age of onset.
- The recurrence rate of this condition is highly variable.
- Timely intervention can help to prevent associated complications of the disease in a growing child.

Contributors MR: involved in writing of the case report, patient care and performing the surgery. KG: involved planning the treatment. NG: involved in intervention/performing the surgery and patient care. AK: involved in patient care.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Parental/guardian consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- Neville D, Allen B. Periodontal disease. In: *Oral and maxillofacial pathology*: Elsevier Saunders, 2004:148–50.
- Regezi JA, Sciuba JJ. Connective tissue lesions. In: *Oral pathology: clinical pathologic correlations*: W.B. Saunders, 1999:179–83.
- Ball EL. Case of gingivoma, or elephantiasis of the gingiva. *J Periodontol* 1941;12:96–100.
- Bozzo L, de Almedia OP, Scully C, *et al*. Hereditary gingival fibromatosis. Report of an extensive four-generation pedigree. *Oral Surg Oral Med Oral Pathol* 1994;78:452–4.
- Rateitschak-Plüss EM, Hefti A, Lörtscher R, *et al*. Initial observation that cyclosporin-A induces gingival enlargement in man. *J Clin Periodontol* 1983;10:237–46.
- Doufexi A, Mina M, Ioannidou E. Gingival overgrowth in children: epidemiology, pathogenesis, and complications. A literature review. *J Periodontol* 2005;76:3–10.
- Agrawal AA. Gingival enlargements: Differential diagnosis and review of literature. *World J Clin Cases* 2015;3:779–88.
- Hallmon WW, Rossmann JA. The role of drugs in the pathogenesis of gingival overgrowth. A collective review of current concepts. *Periodontol* 2000 1999;21:176–96.
- Kasaboğlu O, Tümer C, Balci S. Hereditary gingival fibromatosis and sensorineural hearing loss in a 42-year-old man with Jones syndrome. *Genet Couns* 2004;15:213–8.
- Witkop CJ. Heterogeneity in gingival fibromatosis. *Birth Defects Orig Artic Ser* 1971;7:210–21.
- Omori K, Hanayama Y, Naruishi K, *et al*. Gingival overgrowth caused by vitamin C deficiency associated with metabolic syndrome and severe periodontal infection: a case report. *Clin Case Rep* 2014;2:286–95.
- Konidena A, Puri G, Singh S, *et al*. Plasma cell gingivitis: a case report. *Journal of Indian Academy of Oral Medicine and Radiology* 2014;26:219–21.
- Ejil AL, Thomas A, Mercier S, *et al*. Unusual gingival swelling in a 4-year-old child. *Oral Surg Oral Med Oral Pathol Oral Radiol* 2014;118:627–31.
- Rowland M, Fleming P, Bourke B. Looking in the mouth for Crohn's disease. *Inflamm Bowel Dis* 2010;16:332–7.
- Stewart C, Cohen D, Bhattacharyya I, *et al*. Oral manifestations of Wegener's granulomatosis: a report of three cases and a literature review. *J Am Dent Assoc* 2007;138:338–48.
- Rajendran P, Karmegaraj B, Vij M, *et al*. Unusual cause for gum hypertrophy and skin nodules in a child. *BMJ Case Rep* 2015;2015:bcr2015211506.
- Patil S, Kalla N, Ramesh D, *et al*. Leukemic gingival enlargement: a report of two cases. *Arch Orolfac Sci* 2010;5:69–72.
- Tonguç MÖ, Ünal S, Arpacı RB. Gingival enlargement in children with sickle cell disease. *J Oral Sci* 2018;60:105–14.
- Ghattamaneni S, Guttikonda VR, Yeluri S, *et al*. Early diagnosis of an isolated primary peripheral T-cell lymphoma masquerading as massive gingival enlargement in a pediatric patient. *J Oral Maxillofac Pathol* 2017;21:421–4.
- Krishna KB, Raju PK, Chitturi RR, *et al*. Prevalence of gingival enlargement in Karnataka school going children. *J Int Oral Health* 2014;6:106–10.
- Olczak-Kowalczyk D, Krasuska-Slawinska E, Rokicki D, *et al*. Case report: infantile systemic hyalinosis: a dental perspective. *Eur Arch Paediatr Dent* 2011;12:224–6.
- Gagliano N, Moscheni C, Dellavia C, *et al*. Morphological and molecular analysis of idiopathic gingival fibromatosis: a case report. *J Clin Periodontol* 2005;32:1116–21.
- Aneundi RT, Sudha P, Nayak UA, *et al*. Idiopathic gingival fibromatosis: a case report. *Hong Kong Dent J* 2006;3:53–7.
- He L, Ping FY. Gingival fibromatosis with multiple unusual findings: report of a rare case. *Int J Oral Sci* 2012;4:221–5.
- Breen GH, Addante R, Black CC. Early onset of hereditary gingival fibromatosis in a 28-month-old. *Pediatr Dent* 2009;31:286–8.
- Ko YC, Farr JB, Yoon A, *et al*. Idiopathic gingival fibromatosis: case report and review of the literature. *Am J Dermatopathol* 2016;38:e68–71.
- Kavvadia K, Pepelassi E, Alexandridis C, *et al*. Gingival fibromatosis and significant tooth eruption delay in an 11-year-old male: a 30-month follow-up. *Int J Paediatr Dent* 2005;15:294–302.
- Kelekis-Cholakis A, Wiltshire WA, Birek C. Treatment and long-term follow-up of a patient with hereditary gingival fibromatosis: a case report. *J Can Dent Assoc* 2002;68:290–4.
- Gawron K, Łazarz-Bartyzel K, Fertala A, *et al*. Gingival fibromatosis with significant de novo formation of fibrotic tissue and a high rate of recurrence. *Am J Case Rep* 2016;17:671–5.

Copyright 2018 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <https://www.bmj.com/company/products-services/rights-and-licensing/permissions/>
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow