Familial gingival fibromatosis: A rare case report

Shweta Sharma, Dhawal Goyal, Gaurav Shah, Amit Ray[1]

Abstract

Hereditary gingival fibromatosis is a rare condition that can occur as an isolated disease or as part of a syndrome or chromosomal abnormality. In severe cases, the gingival enlargement may cover the crowns of teeth and cause severe functional and aesthetic concerns. Here, we present a case of an 8-year-old girl with severe enlargement of gums in maxilla and mandible. Both deciduous and permanent teeth were not erupted in the oral cavity at all. Mutation in the *Son-of-Sevenless (SOS-1)* gene has been associated with the disease. The diagnosis was made based on clinical examination and family history. Surgical removal of the hyperplastic tissue was performed under general anesthesia.

Keywords: Enlargement, familial, gingival fibromatosis

Introduction

Gingival hyperplasia is a bizarre condition causing aesthetic, functional, psychological and masticatory disturbance of the oral cavity. Familial gingival fibromatosis is a rare hereditary condition that has no definite cause.[1] Investigations are in evolution to establish the genetic linkage and heterogeneity associated with it.[2,3] This condition may manifest as an autosomal-dominant or, less commonly, an autosomal-recessive mode of inheritance, either as an isolated disorder or as part of a syndrome.[4,5] Autosomaldominant forms of gingival fibromatosis, which are usually non-syndromic, have been genetically linked to the chromosomes 2p21-p22 and 5q13-q22.

In modern times, a mutation in the *Son-of-Sevenless* (*SOS-1*) gene has been suggested as a possible cause of isolated (non-syndromic) gingival fibromatosis, but no definite linkage has been established.[6] Familial gingival fibromatosis is a gradually progressive benign enlargement that affects the marginal gingiva, attached gingival and interdental papilla.

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The fibromatosis may potentially cover the exposed tooth surfaces, thereby hampering the function of the stomatognathic system. The gingival tissues are usually pink, firm and fibrotic in consistency. Histopathologically, the bulbous increased connective tissue is relatively avascular and has densely arranged collagen-fiber bundles, numerous fibroblasts and mild chronic inflammatory cells. The overlying epithelium is thickened and acanthotic, and has elongated rete ridges.[7,8] The autosomal-dominant form is often associated with hypertrichosis, corneal dystrophy, nail defects, deafnessand craniofacial deformities whereas in the autosomal-recessive form, facial anomalies with hypertelorism have been observed but most forms are without defects, other than gingival enlargement. Consanguinity has been observed in the recessive form. Clinical abnormalities most commonly associated with gingival fibromatosis are hirsutism, epilepsy, oliogophrenia, mental retardation, nystagmus, strabismus, cataracts, soft tissue tumors and enlarged facial bones. We report a case of non-syndromic familial gingival fibromatosis along with its management.

Case Report

An 8-year-old female patient had reported to the Department of Pediatric Surgery at the Government Hospital with the complaint of enlargement of gums [Figure 1, 2]. The patient was unable to close her mouth because of severe enlargement of gums in both maxillary and mandibular arches [Figure 3]. There were no teeth visible in the oral cavity [Figure 4]. The patient was referred to the oral surgery outdoor for their opinion. After a detailed history, it was found that the patient's brother also had the same problem [Figure 5]. The patient's father was treated for the same condition in his childhood by alveolectomy with removal of primary and permanent teeth or tooth buds. This type of enlargement was also seen in the patient's uncle and grandfather. Provisional diagnosis made was familial gingival fibromatosis. She did not have any associated medical conditions, which ruled out the possibility of any syndromic involvement. An orthopentomogram was

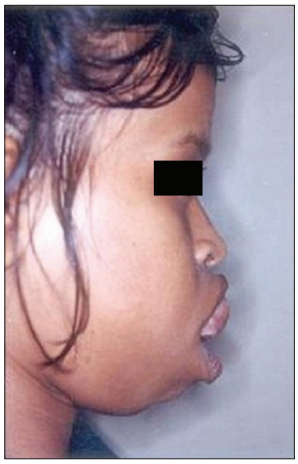

Figure 1: Right lateral extraoral view

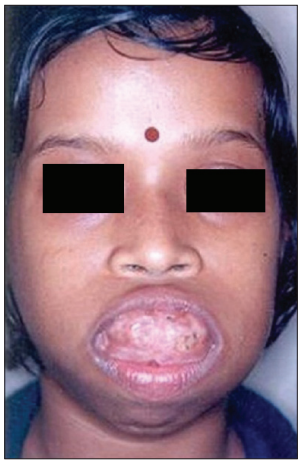

Figure 2: Pre-operative frontal view extraoral

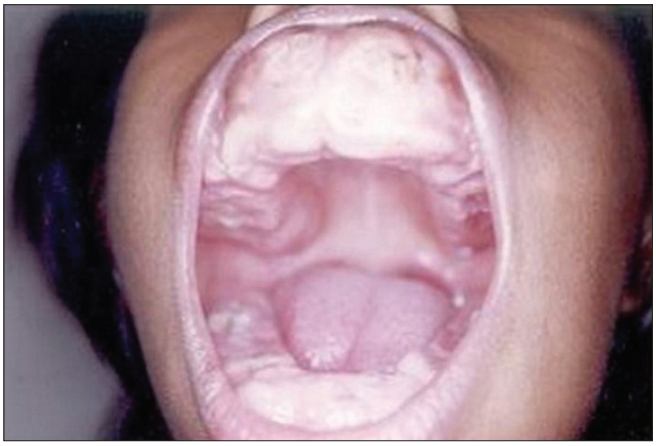

Figure 3: Intraoral open mouth view

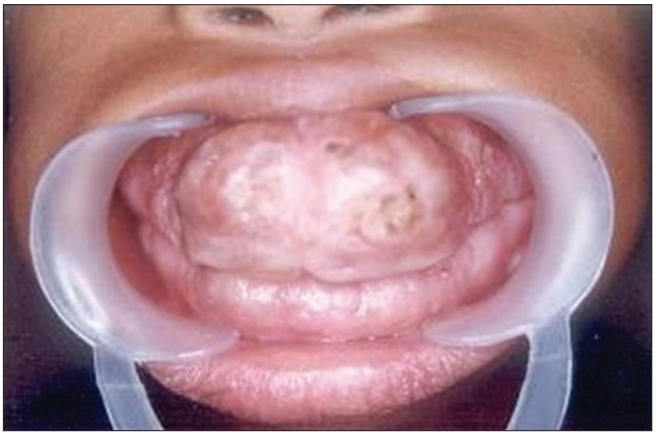

Figure 4: Intraoral close mouth view

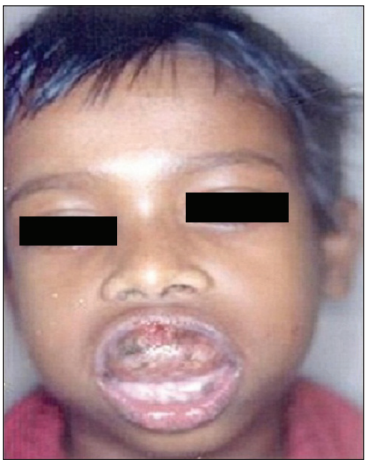

Figure 5: Patient's brother

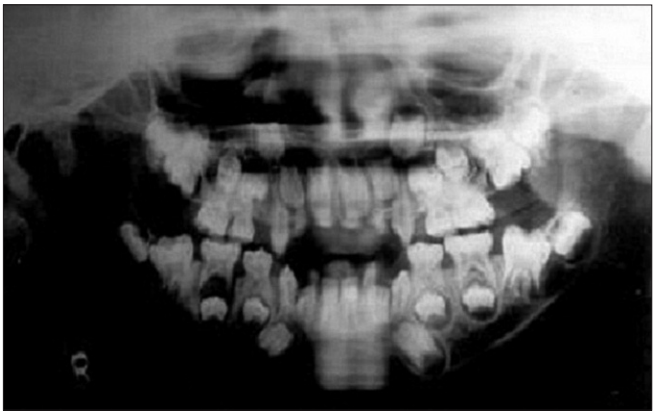

Figure 6: Orthopentomogram view

advised, which showed all primary teeth to be erupted in alveolar bone and developing permanent teeth [Figure 6] The treatment plan was to remove the fibrosed gingiva along with removal of mobile deciduous teeth. The patient was operated under general anesthesia. Surgery was planned in two stages after considering the age of the patient and duration of surgery. In the first stage, the mandibular arch was operated [Figure 7] and the maxillary arch was operated 2 weeks later. After the second surgery, fine periodontal surgery was performed in both arches and the patient was recalled till 2 years [Figure 8 and 9]. On histopathologic examination, thickened acanthotic epithelium with elongated rete ridges was seen with densely arranged collagen fibers, numerous fibroblasts and few chronic inflammatory cells, suggestive of gingival fibromatosis [Figure 10]. Initially, the patient had difficulty in closure of mouth as new occlusion had to

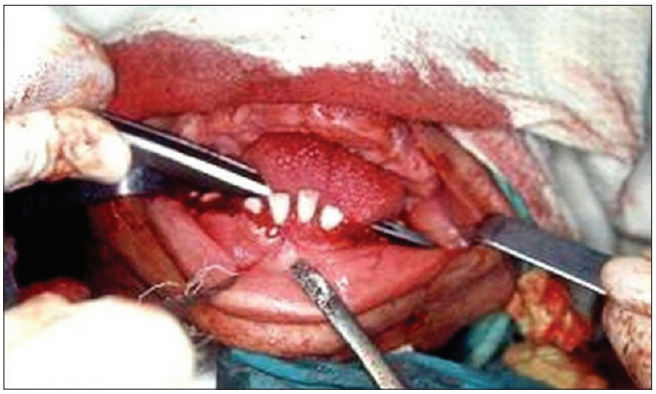

Figure 7: Intraoperative view

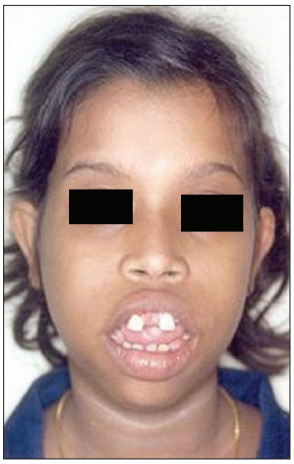

Figure 9: Post-operative frontal extraoral view

be achieved because the patient did not have any occlusion before the surgery.

Discussion

Gingival overgrowth varies from mild enlargement of isolated interdental papillae to segmental or uniform and marked enlargement affecting one or both of the jaws.[9] Here, we reported a case of non-syndromic familial generalized gingival fibromatosis with multidisciplinary approach. There are multiple causes of generalized gingival fibromatosis, like mouth-breathing gingivitis, drug-induced gingival overgrowth, scurvy, hereditary gingival fibromatosis, Wegener granulomatosis, acanthosis nigricans and idiopathic variety.

The precise mechanism of familial gingival fibromatosis is unknown, but it appears to be confined to the fibroblasts that harbor in the gingiva. The hyperplastic response does not involve the periodontal ligament, and occurs peripheral to the alveolar bone within the attached gingiva.[10] The growth is linked with eruption of teeth as seen in the present case, and the presence of teeth may be necessary for the commencement of the process. Fibromatosis gingivae may hinder tooth eruption, mastication and oral hygiene. In

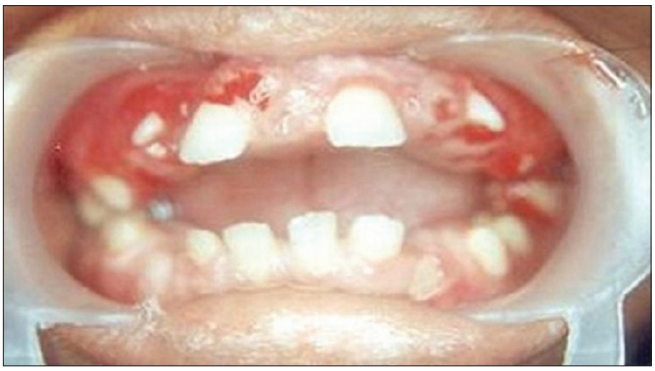

Figure 8: Post-operative intraoral view

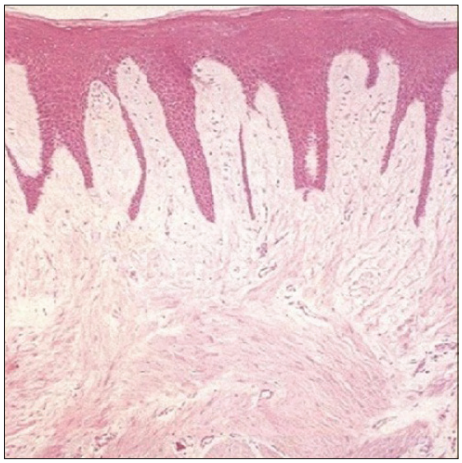

Figure 10: Histopathology shows thickened, acanthotic epithelium, elongated rete ridges with densely arranged collagen-fibers, numerous fibroblasts and few chronic inflammatory cells

severe cases, non-eruption of the primary or permanent teeth may be the chief complaint of the patient.[11] The finest and suggested treatment modality for familial gingival fibromatosis is gingivectomy.[12] Literature reports a high-recurrence rate after surgery, and needs a close follow-up. The present case has been followed for 2 years with no recurrence. There is debate regarding the time of surgery. Eruption of the complete set of permanent teeth is the recommended time for surgery.[13]

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