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Case Report
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Elephantiasis Gingivae: Report of a Rare Case and Literature Review

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Abstract

Elephantiasis Gingivae or Idiopathic gingival fibromatosis is a rare genetic disorder characterized by slow progressive enlargement of the gingiva caused by increase in the mass of submucosal connective tissue. Presented here is a case of a 32 years old female who presented with a massive and uniform enlargement of maxillary and mandibular gingiva. The diagnosis was made based on the clinical, drug and family history and after ruling out possibility of any associated syndrome. The excision of the enlargement was done surgically under general anesthesia.

Keywords: Hereditary gingival fibromatosis; Elephantiasis gingivae; Idiopathic gingival fibromatosis; Fibromatosis fingivae

Introduction

Elephantiasis Gingivae or Idiopathic Gingival Fibromatosis (IGF) is a rare painless slowly growing benign fibrous enlargement of the marginal gingiva, attached gingiva and interdentally papilla [1]. The hyper plastic gingival tissue is usually normal in color, non-hemorrhagic, has firm and leathery consistency and presents a characteristic "pebbled" surface [1]. Stippling may be exaggerated. The gingival tissues enlarge and cover the dental crowns partially or totally forming pseudo-pockets and poor oral hygiene which leads to generalized aggressive periodontitis. It can also cause diastema between teeth and delay or impede eruption of teeth [1,2]. In cases of massive gingival enlargement mastication of food may be painful as the tissues are traumatized during mastication. The speech is affected and the lips appear incompetent.

It may be an isolated finding or may be associated with a syndrome. Synonyms of IGF are hereditary gingival fibromatosis, congenital hypertrophy of gingiva, congenital macrogingivae, fibromatosis gingivae and hypertrophic gingiva. Males and females show equal predilection. Presented here is an unusual case of idiopathic gingival fibromatosis in a 32 years old female.

Case Presentation

A 32 years old female patient walked into the Department of Oral Medicine and Radiology with a C/O swollen and enlarged gums throughout the upper and lower jaws (Figure 1). The history suggested that there was gradual and progressive enlargement of the gums since the age of 13 years, making mastication and speech difficult. The family, postnatal, menstrual and drug history were noncontributory. Patient is unmarried and did not elicit history of any systemic disease. On extra oral examination, a massive enlargement of gingiva was seen giving the patient a convex profile and incompetent lips (Figure 2). Hypertricosis of the facial hair was an evident finding. Intraoral examination revealed gingival enlargement in relation to maxillary and mandibular arches involving both buccal and palatal gingiva in all the four quadrants with reduction in the palatal and lingual space (Figures 3-5). The gingival growth was smooth, normal in color, firm and fibrotic in consistency, teeth appeared submerged in the enlarged gingiva up to the incisal and occlusal third, the maxillary central incisor was displaced buccally, bleeding on probing of the enlargement was absent, and pseudo pockets were present with generalized mobility of teeth. Based on the above findings a provisional diagnosis of idiopathic gingival enlargement was given.

Investigations performed included complete blood picture which showed Hb (11.9 gm %), Peripheral blood smear (normochromic normocytic RBC), Thyroid profile (T3-1.25 mg/ml, T4-10 mg/dl, TSH-3.33 IU/ml) and random blood sugar (103 mg/dl) were within normal limits. A panoramic radiograph was taken which showed severe generalized horizontal alveolar bone loss, vertical bone loss is seen in relation to 46 with missing 26, 27 and 38 (Figure 6). Systemic examination was done by the specialists at St. Jones Medical College Bangalore, to rule out any syndromic association

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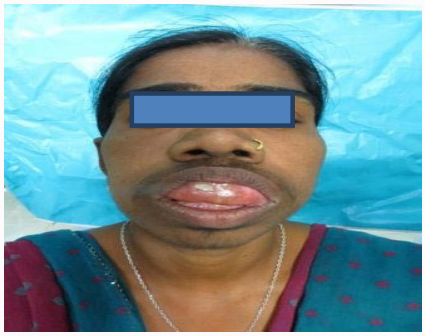

Figure 1: Extraoral photograph showing enlarged gingiva and hypertricosis.

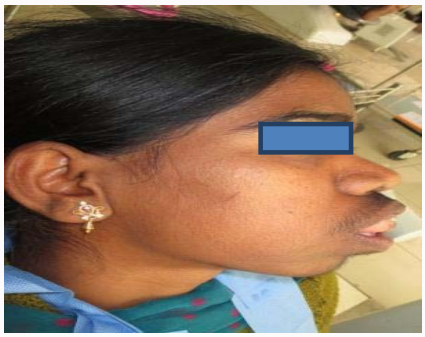

Figure 2: Lateral view showing convex profile and incompetent lips.

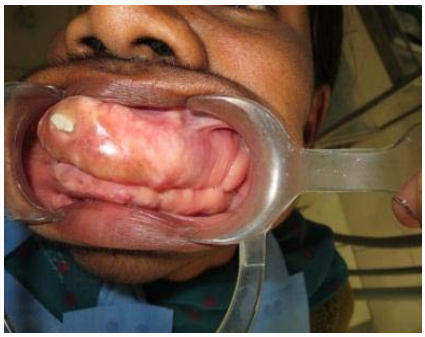

Figure 3: Enlarged maxillary and mandibular gingiva.

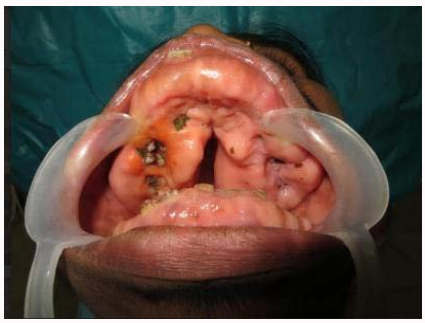

Figure 4: Maxillary gingival enlargement showing palatal space obliteration and submerged teeth.

for cardiovascular, respiratory and musculoskeletal systems. Dermatologic, Gynecologic and Gastrointestinal examination was performed which did not show any abnormal findings.

Histopathological examination of the Incisional biopsy specimen taken from the region of 13 and 14 revealed parakeratinized stratified

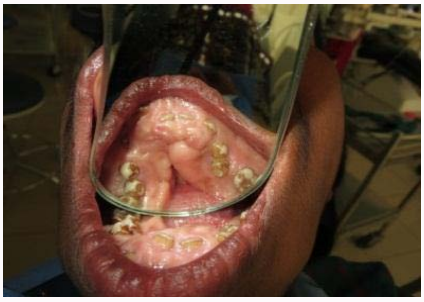

Figure 5: Mandiblar gingival enlargement showing obliteration of tongue space and submerged teeth.

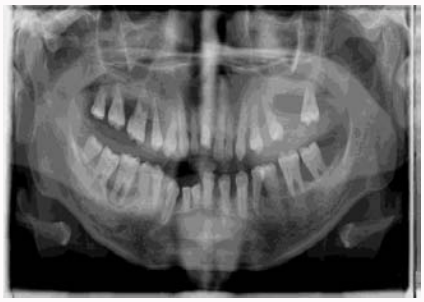

Figure 6: OPG showing generalized horizontal alveolar bone loss.

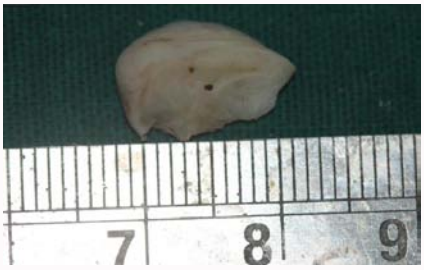

Figure 7: Biopsy specimen (1.3 cm × 0.7 cm × 0.6 cm).

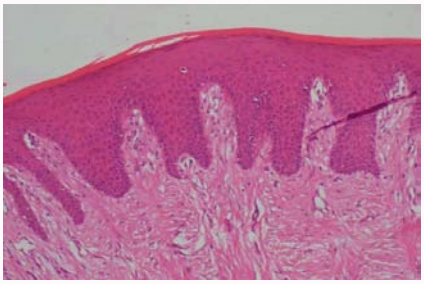

Figure 8: H and E staining showing parakeratinized stratified squamous epithelium, long rete ridges and hypocellular, hypovascular connective tissue with bundles of dense collagen fibers.

squamous epithelium of varying thickness with majority of areas showing long rete ridges (Figure 7). Underlying connective tissue was hypo cellular, hypo vascular with bundles of dense collagen fibers. A focal area of dense inflammatory infiltrate predominantly consisting of lymphocytes was seen (Figure 8). This lead to the final diagnosis of "Idiopathic gingival fibromatosis". Accordingly, gingivectomy for both maxillary and mandibular arches with extraction of 15, 16, 17, 25, 27, 36, 37, 46, 47 and 48 was performed under general anesthesia. Considering the young age of the patient the anterior teeth were retained. In order to restore function a removable partial denture

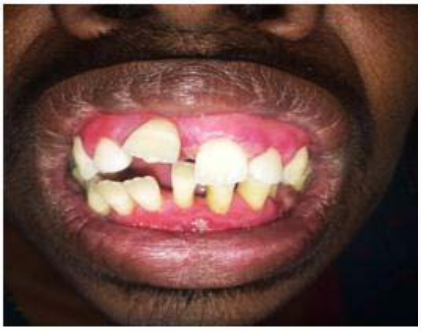

Figure 9: Follow up at 1 month.

was fabricated for the maxillary and mandibular posterior teeth. No recurrence was observed 1 month after the surgery (Figure 9).

Discussion

Idiopathic gingival fibromatosis is a rare genetic disorder characterized by slow progressive enlargement of the gingiva. Usually this condition is not seen at birth but has been reported at the time of eruption of the permanent dentition [3]. The enlargement appears to be most extensive during the loss of deciduous teeth or in early stages of permanent teeth eruption. The growth is rapid during the phase of active eruption and ceases at the end of eruption. Some rare cases affecting the deciduous dentition have also been reported [4].

The condition may present as multiple nodules involving the interdental papillae or as symmetric uniform enlargement of gingiva or a combination of both forms unilaterally or bilaterally and can involve both the arches [5]. IGF is a slowly progressive, benign enlargement which involves the marginal gingiva, interdental papillae and the attached gingiva unlike drug-induced gingival enlargement, which limits itself to marginal gingival and interdental papillae [6].

Gingival Fibromatosis has been classified as [7]:

1. Isolated HGF.
2. Isolated IGF.
3. GF with hypertrichosis.
4. GF with hypertrichosis and mental retardation and/or epilepsy.
5. GF with mental retardation and/or epilepsy.
6. GF associated with other diseases as part of a syndrome.

Syndromes associated with IGF are Zimmermann-Laband syndrome (defects of bone, nail, ear and nose accompanied by splenomegaly), Murray-Puretic-Drescher syndrome (multiple dental hyaline tumors), Rutherford syndrome (corneal dystrophy), Jones syndrome (sensorineural deafness), Cowden syndrome (multiple hamartomas), and Cross syndrome (hypopigmentation with athetosis Cornelia de Lange syndrome (primordial growth deficiency, severe mental retardation, anomalies of the extremities and a characteristic face) [8,9]. Klippel-Trenaunay syndrome (GF, hemi hypertrophy, Nevus flammeus, hemangioma, hypertlorism, and macrocephaly) and Ramon syndrome (Hypertrichosis, Mental retardation, Delayed development, Epilepsy and Cherubism) [10,11]. Takagi et al. [12] has also reported a syndrome associated with IGF and hearing deficiencies, supernumerary teeth and hypertelorism Present case was diagnosed as idiopathic gingival fibromatosis without syndromic association was evident.

A single gene mutation may result in isolated GF while syndromic forms may result from alterations of multiple genes or a gene dosage effect [13]. Autosomal dominant non-syndromic forms have been associated with chromosome 2p21-p22 and 5q13-q22 [13]. A single nucleotide-insertion mutation in codon 1083 of the SOS-1 gene localized to chromosome 2p21-p22 is the cause of Hereditary GF in humans [1,12]. Recently, IGF has been attributed to a mutation in Son of Sevenless-1 (SOS-1) gene. SOS-1 gene codes for a protein that activates RAS pathway, which signals cell growth. When this gene is mutated, it results in gingival fibromatosis; otherwise it is associated with growth of normal gingiva [1].

Histologically, there is accumulation of relatively a vascular dense fibrous connective tissue with densely arranged collagen-fiber bundles, numerous fibroblasts and chronic inflammatory cells. The overlying epithelium is thickened and acanthotic with elongated rete ridges [1-3,5-7,9]. An imbalance between synthesis and degradation of extracellular matrix composed mainly of collagen, increased proliferative activity and decreased apoptosis of fibroblasts could contribute to fibrotic overgrowth of gingiva [14,15]. It is also attributed to alteration in fibroblast proliferation. The precise mechanism process has not been identified [3]. Our case presented with similar histological findings. Presence of small calcified particles, amyloid deposits, islands of odontogenic epithelium, and osseous metaplasia in the connective tissue are unusual findings [16].

Treatment modalities comprise of surgery, use of a carbon dioxide laser and electrocautery. In the present case, gingivectomy for maxilla and mandible was performed under general anesthesia. Regular follow up of the patient is being done to assess her periodontal status and any recurrence of gingival overgrowth. The patient will undergo orthodontic treatment and prosthetic replacement of missing teeth so that the esthetic and masticatory function is restored.

Post surgery the recurrence rate is very high and therefore repeated surgeries may be required [17]. More the fibroblasts present, greater is the chance for recurrence [18]. A permanent remolding of gingival tissue is seen after teeth extraction which suggests the role of teeth and the gingival crevicular environment of in the pathogenesis of GF [19]. According to Emerson the best time to excise the enlarged gingival tissue is post eruption of the permanent teeth [20].

Conclusion

Gingival Fibromatosis (GF) is a disorder of heterogeneous origin; it may be inherited or idiopathic. Our case presented with a rare condition of non-syndromic idiopathic gingival enlargement, which had to undergo a series of clinical, blood, hormonal, radiographic and histopathological examinations to arrive at the current diagnosis. In this condition easthetic, masticatory and psychological status of the patient is affected; therefore a multidisciplinary approach for the management is required. Complete excision and prosthetic rehabilitation will appreciably improve the aesthetic and masticatory competence of the patient. As it is associated with a high rate of recurrence, a periodic observation is mandatory.

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