

Idiopathic gingival fibromatosis associated with progressive hearing loss: A nonfamilial variant of Jones syndrome

BAGAVAD GITA, SAJJA CHANDRASEKARAN, PRAKASH MANOHARAN¹, GARIMA DEMBLA

Abstract

Gingival fibromatosis is characterized by gingival tissue overgrowth of a firm and fibrotic nature. The growth is slow and progressive and is drug-induced, idiopathic, or hereditary in etiology. It occurs isolated or frequently as a component of various syndromes. Our patient presented with the complaint of gingival enlargement associated with progressive deafness, characteristic of Jones syndrome. This case report is important and unique since it is the first known one to have a Jones syndrome-like presentation without a family history. A male patient aged 14 years reported with the chief complaint of swelling of gums and progressive hearing loss in both ears for the past one year. There was no family history or history of drug intake. Enlargement was generalized, fibrotic and bulbous, involving the free and attached gingiva, extending up to the middle 1/3rd of the crown. Investigations such as pure tone audiogram, impedance audiometry, and Tone decay test concluded that there was severe right and moderate left sensorineural hearing loss. The case was diagnosed to be idiopathic, generalized gingival fibromatosis with progressive hearing loss. The gingival overgrowth was managed by gingivectomy and periodic review. The patient was advised to use high occlusion computer generated hearing aids for his deafness as it was not treatable by medicines or surgery. This unique case report once again emphasizes the heterogeneity of gingival fibromatosis, which can present in an atypical manner.

Keywords: Familial, gingival overgrowth, idiopathic, Jones syndrome, progressive hearing loss

Introduction

Gingival fibromatosis is an overgrowth of the gingiva characterized by an expansion and accumulation of the connective tissue with an occasional presence of an increased number of cells.^[1] The condition may be drug-induced, inherited, or idiopathic. The clinical and genetic heterogeneity of the disease have made the diagnosis and predictability of treatment success difficult. Gingival fibromatosis is often a part of many syndromes one of which is Jones syndrome. It was first reported by Jones *et al.*^[2] in an 11-year-old white male. Family history revealed five generations to be affected by the disease. Kasaboğlu *et al.*^[3] also reported gingival fibromatosis and sensorineural hearing loss in a 42-year-old man who gave an anamnestic history of four family members over three generations being affected.

Department of Periodontology, Sree Balaji Dental College and Hospital, and ¹Department of ENT, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India

Correspondence: Dr. Bagavad Gita,
Flat "H" Prajwal Vijay Nivas, 457, R.K. Shanmugam Salai,
K. K. Nagar, Chennai - 600 078, Tamil Nadu, India.
E-mail: gita70.geetha@gmail.com

Wynne *et al.*^[4] have reported on three generations of one family with an autosomal dominant expression of gingival fibromatosis with associated hearing deficiencies, hypertelorism, and supernumerary teeth. A variant of this, namely sensorineural hearing deficit and premature tooth loss has also been reported as a part of Coffin-Lowry syndrome (mental retardation, developmental delay and characteristic facial changes).^[5]

This case report is similar in its clinical presentation to Jones syndrome but lacks a definite familial inheritance. It is the third case in the literature so far to report gingival fibromatosis with hearing loss, and the first-ever reported one without a genetic predilection. This atypical presentation should make dentists, physicians and periodontists aware that isolated cases of gingival fibromatosis with hearing loss do occur without a familial history.

Case Report

A 14-year-old male patient reported to the Department of Periodontology with the chief complaint of swollen gums and deafness for the past 1 year. The patient also complained of poor esthetics due to a "gummy smile" and inability to close the mouth completely. The medical history was non-contributory for systemic diseases or drug intake. No history of ear trauma or exposure to loud noises was reported. Family history revealed that the patient was the only one to be affected over three generations.

On intraoral examination, there was a generalized gingival overgrowth. It was firm, nodular and fibrotic involving the marginal, attached gingiva and interdental papillae of all the teeth with no associated

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inflammation [Figures 1 and 2]. The gingiva was dark in color due to heavy melanin pigmentation. The lower anterior teeth were barely visible as the enlargement extended up to the incisal-third of the crowns. Examination of the teeth revealed that the patient was still in the mixed dentition stage though he was 14 years old. Orthopantomograph revealed all the missing permanent teeth to be present radiographically [Figure 3]. There was no evidence of alveolar

bone loss. Periodontal examination with a periodontal probe (UNC-15 periodontal probe, Hu-Friedy, Chicago, IL) revealed no associated clinical attachment loss or mobility.

The patient was referred to an ENT surgeon for management of hearing loss. Pure tone audiogram, impedance audiometry



Figure 1: Facial view of the enlargement



Figure 2: Lateral view of overgrowth



Figure 3: Orthopantomograph of patient showing all missing teeth to be present radiographically

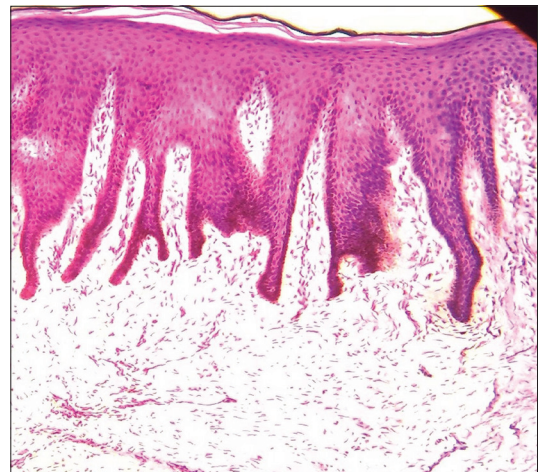


Figure 4: Parakeratinised stratified squamous epithelium with underlying dense fibrous connective

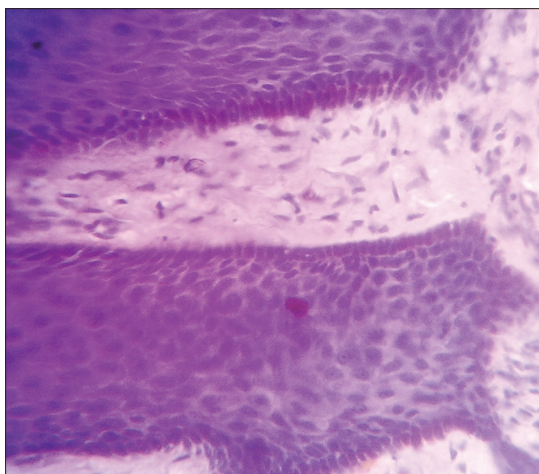


Figure 5: Hyperplastic epithelium showing prominent elongated rete ridges



Figure 6: Follow-up image after 24 months

and Tone decay test and brainstem evoked response audiometry tests were conducted. It was concluded there was severe to profound right and moderately severe to severe left sensorineural hearing loss. The case was provisionally diagnosed to be a case of idiopathic gingival fibromatosis with progressive hearing loss. The patient was recommended high occlusion computer generated hearing aids.

Management of the gingival overgrowth was done with phase I therapy and surgical excision. Under local anesthesia, quadrant by quadrant gingivectomy was carried out. A biopsy sample of the excised tissues was presented for histological evaluation. The histopathological picture showed parakeratinized stratified squamous epithelium with underlying dense fibrous connective tissue stroma [Figure 4]. The overlying epithelium was characteristically hyperplastic and showed prominent, elongated rete ridges [Figure 5]. Periodic review was carried out every 3 months. The patient was followed-up for a period of 24 months. There was a slight recurrence in the overgrowth, though lip competence was maintained [Figure 6]. The auditory tests, repeated after 1 year, revealed the hearing loss to be at the same level. The etiology was unknown, though an immunological influence could not be ruled out. Periodic review and audiometry once a year were suggested to monitor the progression of the hearing loss.

Discussion

Gingival fibromatoses can be either hereditary or idiopathic. Variations in the inheritance pattern and its frequent association with systemic defects or anomalies make it very complex to study.^[6] It is frequently a component feature of many syndromes. Jones syndrome was characterized by gingival overgrowth and progressive deafness in five generations of a family. All generations developed hearing loss in the 2nd decade of life. This case report fits into the diagnosis of Jones syndrome, but there is no familial history. However, the possibility of a new mutation has to be taken into consideration.^[7]

Clinical management of gingival fibromatosis is normally surgical excision of the enlargement and periodic review. Due to the continuous increase in mass of tissues, maintenance of oral hygiene become challenging during all phases of therapy.^[7] The tissue overgrowth further leads to spacing of the teeth, delayed eruption and displacement, which make oral hygiene practices further deteriorate. Recurrence rates cannot be predicted, but surgical excision of the enlargement improves not only the patient's esthetics and psyche, but also functionality and oral hygiene. The patients' maintenance of oral hygiene had to be emphasized as high recurrence rates have been associated with poor oral hygiene.^[8,9] On follow-up after a year, the patient showed a mild recurrence of the gingival overgrowth.

The influence of sex hormones on gingival tissues has to be taken into consideration as it is well established that sex steroids influence periodontal metabolism and gingival overgrowth worsens throughout adolescence.^[10,11] The patient should be reviewed for recurrence tendencies after the adolescent phase is over to overrule this influence. Future generations will have to be observed to come to a definitive conclusion if the condition is an isolated one or a new mutation.

Overlapping of the phenotypes of various syndromes of which hereditary gingival fibromatosis is a feature has been reported.^[12] This suggests that identification of the genetic pathways and mechanisms will be the most important factor in classifying these disorders, with the phenotype playing a minor role. The genes for sensorineural hearing loss^[13] and gingival fibromatosis^[14] have been mapped to chromosomes 2p21 and p22 respectively. However, there is as yet no evidence about the molecular/genetic basis of Jones syndrome. Early diagnosis and recognition of hearing loss can help in correction and improvement of the quality-of-life of these patients.

Acknowledgment

Dr. Rathnavel, Post Graduate, Sree Balaji Dental College and Hospital, Chennai.

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How to cite this article: Gita B, Chandrasekaran S, Manoharan P, Dembla G. Idiopathic gingival fibromatosis associated with progressive hearing loss: A nonfamilial variant of Jones syndrome. *Contemp Clin Dent* 2014;5:260-3.

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

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