Case Report

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Hereditary Gingival Fibromatosis: Genetic Lineage

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ABSTRACT

Background: Hereditary Gingival Fibromatosis (HGF) is a rare, benign, genetically inherited gingival condition that presents as localized or generalized fibrotic enlargement of gingiva. It can occur both as an isolated entity as well as a part of a multisystem disorder. It is now reported to be due to defect in the Son of Sevenless-1 gene on chromosome 2p21-p22. Clinically it is characterized by firm, nodular, pale pink, stippled gingiva covering almost the whole teeth. The treatment plan is not definite including surgical intervention with a good follow up and maintenance. This paper presents a case report of a patient suffering from Hereditary Gingival Fibromatosis and highlights the various diagnostic criteria along with its treatment.

Keywords: Gingival fibromatosis, Periodontology, Dentistry.

INTRODUCTION

This case report describes Hereditary Gingival Fibromatosis which is a rare type of gingival hyperplasia having clinical as well as genetic variations but is usually autosomal dominant in inheritance. The etiology of this kind of enlargement was not clear until recently and so it is not frequently documented. Of late it has been found that this condition is genetically inherited and is present in generations of families. This case report thus highlights the importance of knowledge needed regarding such rare hereditary conditions along with its treatment protocol.

CASE REPORT

A 17 year old girl reported to the Department of Periodontics, Karnavati School of Dentistry, Uvarsad with the chief complaint of a swelling behind the back teeth on both sides of the upper jaw (Figure 1). Her history revealed that the swelling was present from past 6-7 years and the patient also had difficulty in chewing and swallowing food. Her family history revealed that her mother and brother were also having similar kind of over growths. The patient's medical history was non- contributory to the development of the gingival enlargement and there was no history of any drug usage.

The intraoral examination revealed bilateral gingival enlargement distal to the 2nd maxillary molar. These gingival enlargements were pale pink in color, firm in consistency, nodular, painless, well circumscribed and had a smooth surface devoid of stippling.

Investigation

Radiographic examination revealed resorption of root in relation to permanent maxillary right first molar. Orthopantomograph was taken to rule out any odontogenic or osseous lesion. Routine blood investigations were also carried out which revealed normal haemogram.

Differential Diagnosis

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HGF is a kind of gingival enlargement which has been found to occur either as an isolated entity or as a component of multisystem syndromes.[1,2] Apart from HGF, other causes of isolated gingival enlargements can be plasma cell gingivitis, plaque induced gingival enlargement, scurvy or trauma. Plasma cell gingivitis shows enlargement which is erythematous and its histopathological examination shows a dense infiltration of plasma cells. It is found to be a hypersensitivity reaction to certain agents. Plaque induced gingival enlargement presents with abundant amount of local factors, reddish and odematous gums. In scurvy severe bleeding of gums is associated with gingival enlargement and altered vitamin C levels.[3]

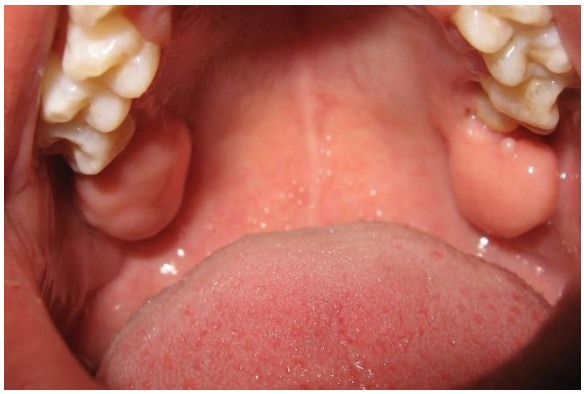

Fig 1: Pre-operative photograph showing bilateral gingival hyperplasia.

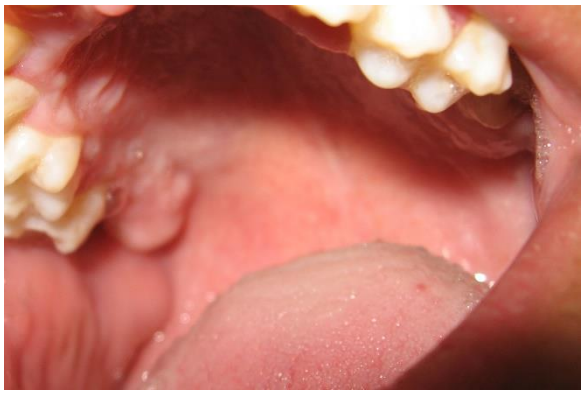

Fig 2: One year post-operative photograph.

HGF has also been found to be associated with other systemic manifestations like oro-facial granulomatosis, Crohn's disease, Sarcoidosis, Wegeners' granulomatosis, Zimmer-man Laband syndrome, Rutherford syndrome, Jones syndrome, Cross syndrome, Murray-Puretic Drescher's syndrome, Ramon syndrome etc.[3,4]

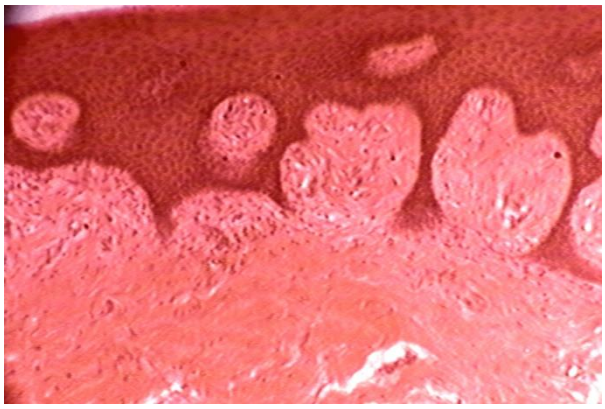

Fig 3: Photomicrograph of the excised tissue.

Oro-facial granulomatosis in addition to gingival enlargement is also associated with oral ulceration, angular cheilitis, tongue fissuring, cobblestoning, lip swelling, Bell's palsy and cervical lymphadenopathy. Crohn's disease shows similar kind of oral features and is also associated with additional GIT manifestations. Sarcoidosis shows oral ulceration, bony lesions with oversized jaws alongside granular gingival enlargement. Wegeners' granulomatosis is associated with salivary glands enlargement, lip swelling, oroantral fistula, osteonecrosis and shows specific strawberry like gingivitis. Zimmer-man Laband syndrome is associated with enlarged nose and pinnae of the ears, nail dystrophy, hepatosplenomegaly, hypoplastic distal phalanges, mental retardation and deafness. Rutherford syndrome has been found to be linked with corneal dystrophy, aggressive behavior and mental retardation. Jones syndrome shows progressive deafness, multiple maxillary odontogenic cysts and undescended testis. Cross syndrome shows microophthalmia, hypopigmentation, athetosis and mental retardation. Murray-Puretic Drescher's syndrome has involvement of various bones, joints, cartilage, skin and muscles. Ramon syndrome constitutes cherubism, hypertrichosis, stunted growth, juvenile rheumatoid arthritis, seizures and mental retardation. Based on all the above facts and absence of syndromic associations, positive family history along with clinical features a diagnosis of Hereditary Gingival Fibromatosis was made.

Treatment

The patient was given oral hygiene instructions and adequate plaque control was done. The surgical intervention was carried out under

local anaesthesia, bleeding points were marked using a pocket marker and external bevel gingivectomy was performed on both sides over a gap of one week.(Figure 2) Periodontal pack was applied over the surgical site and patient was prescribed Amoxicillin 500mg thrice daily for 5 days and Aceclofenac 100mg twice a day for 5 days. The excised tissue was of the dimension 1x1.5 cms on right side and 1x2 cms on the left side. The tissue was then subjected to histopathological examination. The patient was followed up every one month for first 3 months and then after every 3 months for next one year. No re-occurrence has been observed so far.

DISCUSSION

HGF is a rare, benign, slow growing, non-hemorrhagic fibrous enlargement of gingiva[5] not extending beyond the mucogingival junction.[6] It was first reported by Goddard and Gross in 1856[7] and was previously known as elephantiasis gingiva, hereditary gingival hyperplasia and hypertrophic gingiva.[8] Males and females have been found to be equally affected at a phenotype frequency of 1:750,000 and genotype frequency of 1:350,000 with varying intensity and expressivity even in individuals within the same family.[5] The condition may have autosomal dominant or recessive mode of inheritance. The onset of this condition usually occurs with the eruption of permanent teeth but may also be seen with the eruption of primary dentition. This condition can lead to mal-positioning of teeth, interference in speech, closure of the lips and mastication resulting in aesthetic and functional problems.[1]

Till now the etiology of this condition was not known and hence it was also called idiopathic gingival enlargement. But recent studies have shown it to be a condition of genetic origin commonly following a Mendelian autosomal dominant pattern of inheritance. Three genetically separate loci have been found on chromosome 2p21-p22 (GINGF), 2p22.3-23 (GINGF3) and 5q13-q22 (GINGF2).[9] A single nucleotide mutation in codon 1083 of GINGF of SON of sevenless-1 (SOS1) gene has also been isolated.[10] These mutations follow autosomal dominant mode but inheritance by recessive mode has also been reported.[8] Since the patient revealed a positive family history and absence of other systemic involvement, this case was suggested to be an example of HGF.

Various treatment modalities have been implemented for such cases ranging from conservative approaches to extraction of teeth.[11] In this case as the enlargement was localized and present beyond the distal surface of second molars, external bevel gingivectomies were planned with restoration of physiological contours of gingiva. The excised tissue was sent for histopathological examination and adequate post-operative care was ensured to minimize the recurrence.

Histopathological examination revealed highly fibrous connective tissue with dense collagen bundles, increased fibroblasts, inflammatory infiltrate consisting of few lymphocytes, plasma cells, neutrophils and mast cells.(Figure 3) Connective tissue was found to be relatively avascular with compressed capillaries. The overlying epithelium was hyperkeratotic and acanthotic, stratified squamous epithelium with elongated rete ridges thus complementing our diagnosis.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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