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HEREDITARY GINGIVAL FIBROMATOSIS;
RECURRENT, SEVERE ENOUGH TO CLOSE THE MOUTH.

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ABSTRACT: Hereditary gingival fibromatosis also referred to as idiopathic gingival hyperplasia, elephantiasis gingivae etc; is a rare hereditary localized or generalized condition, which represents as swelling of gingivae of varied degrees with no apparent cause. Swelling is usually firm, pink and usually no signs of inflammation if oral hygiene is up to fair. Condition may occur solitary or in a syndrome. Recent findings report a defect in the Son of seven less-1 gene on chromose 2p21-p22(HGF1) as a possible cause. This case report presents a severe HGF case, importance of earlier management (as it may unable the patient to close their mouth), possible gravitational role in enlargement of swellings and its proper remedy.

Key words: Hereditary gingival fibromatosis,idiopathic gingival hyperplasia.

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INTRODUCTION

Hereditary gingival fibromatosisis a rare (1 in 750,000) hereditary condition characterized by slow, progressive enlargement of the gingivae[4,2]; caused by an increased in submucosal connective tissue element[1]. It may be inherited or iatrogenic1. Condition when neglected gets severe enough to inhibit proper hygiene maintenance leading to further inflammation, which is not a usual feature in this highly stippled pink colored gingival swelling[3,5]. Due to massive gingival enlargement patient is unable to maintain normal occlusion, speech, mastication and oral flora[1,3]. The mode of inheritance is believed to be autosomal dominant, although reports of a recessive mode of inheritance have also been published[3],. Recent findings report a defect in the Son of sevenless-1 gene on chromose 2p21-p22(HGF1) as a possible cause[5,6]. The gingival enlargement may occur solely or in association with other abnormalities, as part of a syndrome, most commonly in association with hypertrichosis and epilepsy, with or without mental retardation. Other syndromes thathave occasionally been associated with hereditary gingival fibromatosis are Zimmerman– Laband syndrome (defects of bone, ear, nail and nose, accompanied by hepatosplenomegaly)[4,5].

Treatment approach ranges from conservative and surgical procedures to extraction of all teeth and reducing alveolar bone to prevent relapse[4]. Mostly repeated surgical excisions are required as it is a highly recurrent condition.

This report presents a case of a 25 years old female patient with severe non-syndromic generalized hanging gingival enlargements which hinders normal occlusion, mastication, speech and esthetics, which is managed with electro cautery and post surgical deep scaling and measures to prevent further recurrences.

METHODOLOGY

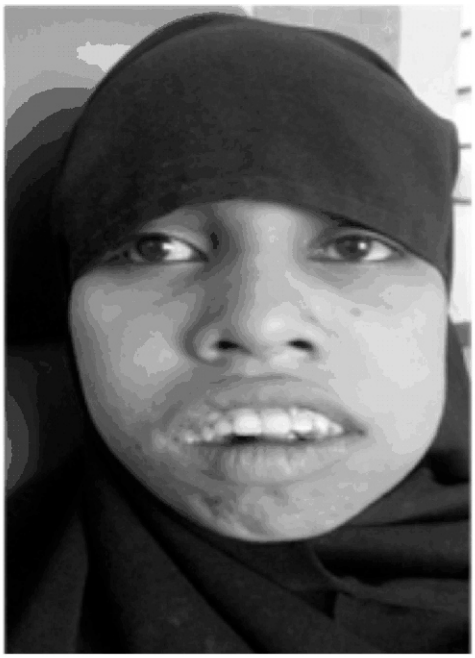
Case Report

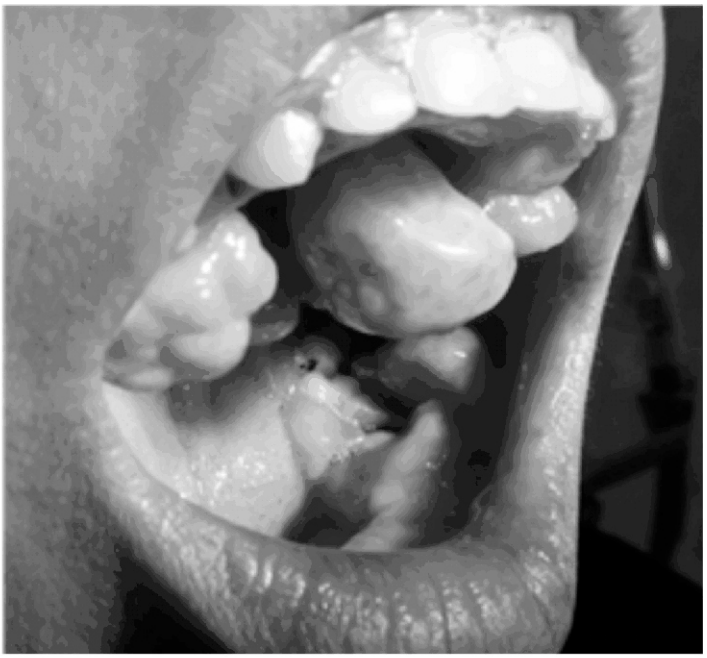
A 25 years old female patient presented in the out patient department of oral surgery in LUMHS (Liaquat University Of Medical and Health Sciences) with recurrence of severe gingival enlargement. Her mother and sister used to have

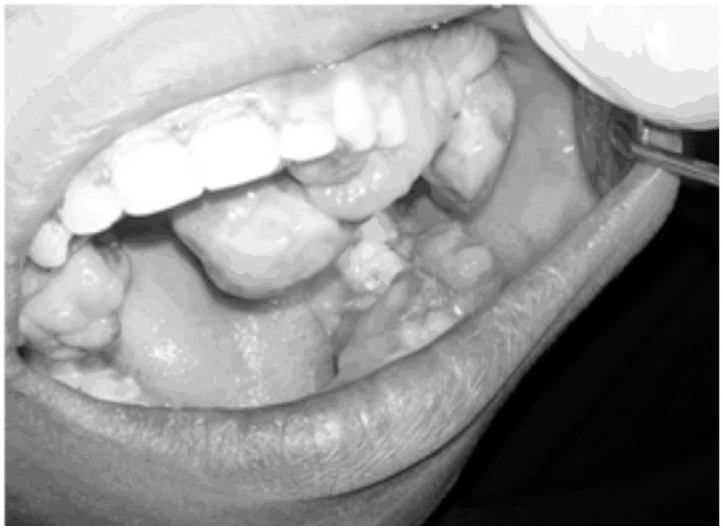
same kind of condition. Swelling gradually reached to so sever condition (that she is unable to close her mouth and maintain proper oral hygiene) because of negligence on the part of patient. She neither had any history of fever, weight loss, seizures, hearing loss nor she had any physical or mental deterioration that may suggest a syndrome and also no drug history that may suggest drug induced gingival enlargement. Chairman of our department treated her previously for same lesion in 2008.

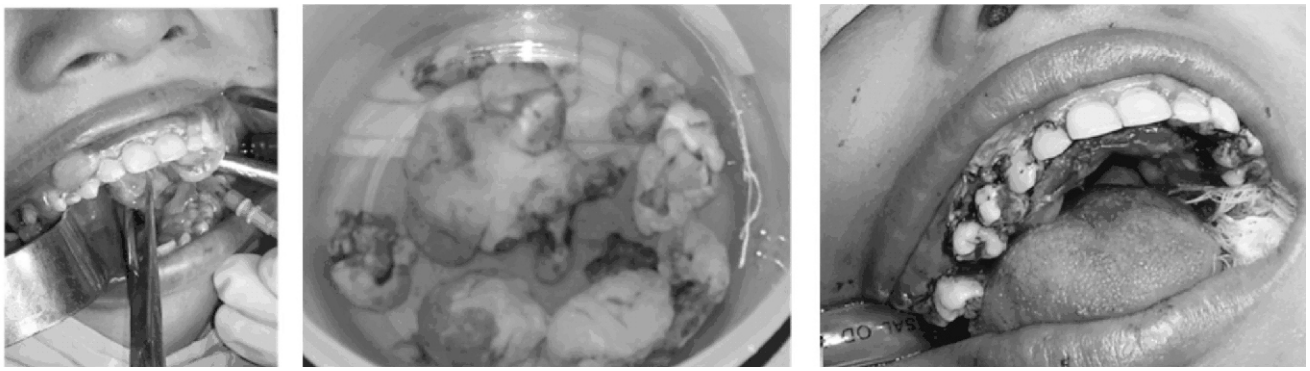
Examination

Extra orally appeared to have a full mouth appearance. Intra orally patient had multiple hanging enlargements originating from maxillary palatal gingiva with slight marginal gingivitis in anterior maxilla, right palatal maxillary swelling had characteristic nodular appearance which was not present in other swellings. Gingival enlargements also covered mandibular teeth but not as much swelling as in maxilla, left posterior mandibular region had some purulent discharge.









Diagnosis

On the basis of patient and family history, clinical presentation and biopsy report it was diagnosed to be a recurrence of HEREDITARY GINIGIVAL FIBROMATOSIS.

Treatment

Surgical resection in first instance under general anesthesia to reduce the miserable picture of the patient and let her at least have her meal. Local anesthesia was injected to the site to reduce the chances of bleeding, further electro cautery was preferred to reduce bleeding (unlikely to occur in this condition) followed by deep sub-gingival scaling, root planning and curettage was performed after the surgical site showed some healing. Meanwhile patient was strictly prescribed to maintain proper oral hygiene to prevent recurrence. Margins of the swellings were also cauterized to damage the potential of the site to grow. There was no complication during surgery except the greater palatine vessel was severed which was easily managed by electro coagulation. Maxillary left 7th tooth become so mobile after this surgical resection that it had to be extracted. Excised tissue was completely immersed in formalin solution in a big jar and was sent for biopsy report.

DISCUSSION

Gingival swelling, either localized or generalized might be due to a number of causes, ranging from inflammation, leukemia, and association with use of medicines like phenytoin, cyclosporine, and nifedipine etc[6].

Here, we report a case of hereditary gingival fibromatosis. It is usually an autosomal dominant condition but also reported to be autosomal recessive. We justify our diagnosis of the gingival fibromatosis as hereditary and as an autosomal dominant, solely relying on the occurrence of the enlargement in both her mother and sister and biopsy report. In this case, HGF occurred as a solitary condition. But a thorough look into the literature reveal that it can be a part and parcel of multi-system syndromes, such as Zimmermann-Laband syndrome (Ear, nose, bone and nail defects with hepatosplenomegaly), Murray-Peretic-Drescher syndrome (juvenile hyaline fibromas), Rutherfurd syndrome (corneal dystrophy, mental retardation, impairment of dental eruption by radicular resorption) and Cross syndrome (microphthalmia, mental retardation, athetosis and hypopigmentation)[4,5]. In this case, a thorough history and examination of the patient, revealed no associated clinical features which may suggest any of the above syndromes.

Numerous remedy options have been suggested for the excision of the enlarged gingival masses, including conventional surgery, electrosurgery, an apically positioned flap and lasers[6]. Surgical method using conventional means like scalpel may sometimes be technically impractical for example in children or mentally handicapped, or in patients suffering from impaired haemostasis[6]. Hence use of electrosurgery in these situations would be beneficial.

Surgical resection was performed at the first instance to let the patient out of that miserable

condition. General anesthesia was justified because of the need of proper access, patient compliance and the severity of the lesion. Electro cautery was decided as there were chances of vessel approximation, in a mouth full of scattered swellings.

It was noted that masses were much bigger in size and were hanging in maxillary region while in mandible swelling was covering teeth but was not having any huge masses as like in maxilla. Gravitational role was suggested to be in it (not confirmed yet).

CONCLUSIONS

Hereditary gingival fibromatosis is a highly recurrent, rare hereditary lesion, which should be managed by surgical resection as soon as possible. In this report we have highlighted the importance of earlier management to prevent severe swelling, which may unable the patient to close the mouth, gravitation role in supporting the swellings to grow and inflammation leading to necrosis if proper hygiene is not maintained.

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