GINGIVAL FIBROMATOSIS WITH HEARING LOSS: A JONES SYNDROME CASE REPORT

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Abstract:

Introduction: Hereditary Gingival Enlargement (HGF), a rare entity, is also known as familial elephantiasis, elephantiasis gingivae, diffuse fibromatosis. It is a benign, non-haemorrhagic fibrous enlargement of gingival tissue. It is frequently a component feature of many syndromes. Jones syndrome is one such syndrome, characterized by gingival overgrowth and progressive deafness.

Case report: A 27-year-old male patient reported to the Department of Oral Medicine and Radiology with the chief complaint of swollen gums for the past 7 years. The patient also complained of bleeding gums on brushing. The medical history stated a concurrent gradual hearing loss 7 years back. An incisional biopsy was done to confirm the fibrous nature of the diagnosis.

Conclusion: This reporting is of unique case that remained undiagnosed for almost a decade. It also emphasises the need of a multidisciplinary approach during diagnosis and treatment.

Introduction:

Gingival fibromatosis (GF) is a rare disorder manifesting as a benign, progressive, non-haemorrhagic, fibrous enlargement of maxillary and mandibular gingiva.¹ It can be generalized involving all quadrants or isolated to one quadrant. It is characterized by an expansion and accumulation of the connective tissue with an occasional presence of an increased number of cells.² The prevalence is one per 175 000 population, and men and women are equally affected.³

The pathogenic pathway is yet to be determined and multiple hypothesis abound. The most accepted one pertains to the overzealous increase

of fibroblasts and fibroblastic activity in gingiva followed by diminished apoptosis. This in turn results in increased collagen synthesis followed by decreased breakdown and thus manifesting as fibrotic gingival overgrowth.³

By way of aetiology, clinical manifestation and histopathological findings, the gingival fibromatosis is categorized as hereditary gingival fibromatosis (HGF), lysosomal storage disorders, vascular disorders, and disorders associated with dental abnormalities.⁴

Case Report

A 27-year-old male patient reported to the Department of Oral Medicine and Radiology with the chief complaint of swollen gums for the past 7

years. The patient also complained of bleeding gums on brushing. The medical history stated a concurrent gradual hearing loss 7 years back with no history of ear trauma or high noise exposure. On examination of his medical records, the hearing loss was stated to be sensorineural in nature. The patient had been treated for the condition and wearing a hearing aid for the same. The systemic diseases and drug history were non-contributory. The personal history revealed oral

home care practices of the patient comprised brushing once a day in horizontal motion using tooth brush and tooth paste. The family history revealed that the patient's mother and sister had similar gingival enlargement. The mother suffered hearing loss as well, however the sister was unaffected. There was no history of consanguineous marriage in the family. On intraoral examination, there was a generalized gingival overgrowth (Figure 1).

Figure 1: Frontal and profile view of patient

It was firm, nodular and fibrotic involving the marginal, attached gingiva and interdental papillae of all the teeth with mild inflammation of marginal gingiva. The gingiva was pale pink in colour with mild irregular melanin pigmentation. Only the incisal third of upper anterior teeth were visible owing to the enlargement extension. Generalised pseudo pockets were observed along with bleeding on probing.

Figure 2: Generalised fibrotic gingival enlargement

Examination of the teeth revealed that the patient had complete permanent dentition with an unerupted upper third molar. Orthopantomograph (Figure 3) confirmed the absence of additional impacted teeth. The alveolar bone loss, attachment loss and tooth mobility was observed to be absent on examination and confirmed by the radiograph.

Figure 3: OPG demonstrating impacted right maxillary molar and absence of alveolar bone loss.

The provisional diagnosis was declared as a case of idiopathic gingival fibromatosis with progressive hearing loss (Jones syndrome). An incisional biopsy was done to confirm the fibrous nature of the diagnosis. The histopathological picture showed stratified squamous epithelium with underlying dense fibrous connective tissue stroma. The sub epithelium demonstrated dense collagenisation and perivascular inflammatory cell inflammation was also observed. (Figure 4).

Figure 4: Incisional biopsy specimen and histopictograph demonstrated stratified squamous epithelium with underlying dense fibrous connective tissue stroma. The sub epithelium demonstrated dense collagenisation and perivascular inflammatory cell inflammation was also observed.

The final diagnosis was thus determined as Hereditary gingival fibromatosis with hearing loss or Jones syndrome.

Discussion

Hereditary Gingival Enlargement (HGF), a rare entity, is also known as familial elephantiasis, elephantiasis gingivae, diffuse fibromatosis.⁸ It is a benign, non-haemorrhagic fibrous enlargement of gingival tissue. It has varying intensity and expressivity even in individuals within the same family. The disease may present as an autosomal dominant or autosomal recessive mode of inheritance. The mode of genetic transmission in this patient points to an autosomal dominant gene because family members of both sexes were affected, and the condition was present in successive generations.⁵

HGF is frequently a component feature of many syndromes. Jones syndrome is one such syndrome, characterized by gingival overgrowth and progressive deafness. The progressive hearing loss develops in the 2nd decade of life. This reported case conforms to the same criteria and demonstrated familial inheritance as well.³ On the genetic front, chromosomal abnormalities

reported for syndromes with gingival fibromatosis include duplications, deletions, and/or other anomalies of chromosomes 2p13-16, 4q, 8, 14q, 19p, 19q, and Xq.⁶

The hyperplastic gingiva manifests a normal coral pink colour and has a rigid consistency. The enlargement of the gingiva concurs with the eruption of the permanent dentition.⁵ Furthermore, the presence of teeth seem to be necessary for HGF to occur because the condition disappears or recedes with the loss of the teeth.⁷ It may be localized (nodular) or generalized (symmetric), thus potentially impeding speech, lip competency, and mastication ensuing both aesthetic and functional complications.⁵ The associated effects include diastemas, teeth mispositioning, retention of primary dentition, delayed eruption, cross and open bites, prominent lips, and open lip posture. The gingival enlargement does not affect the alveolar bone. Nevertheless, owing to the false pockets and the subsequent bacterial plaque accumulation, induces gingivitis, periodontitis, bone resorption and halitosis.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. Recurrence rates cannot be predicted, but surgical excision of the enlargement improves not only the patient's appearance, but mastication, speech and oral hygiene. The patients' maintenance of oral hygiene had to be emphasized as high recurrence rates have been associated with poor oral hygiene.³ Also, the progressive hearing loss must be evaluated and managed on a yearly basis by the ENT specialist.²

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