Treatment of Recurrent Gingival Enlargement associated with Zimmermann-Laband Syndrome: A Case Report

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**ABSTRACT**

Background: Zimmermann-Laband syndrome is a rare disorder characterized by generalized gingival fibromatosis and defects of ear, nose, nail and bone.

Methods: This case report describes the clinical presentation, periodontal findings and treatment of recurrent gingival enlargement in a 22 year old male associated with a previously undiagnosed case of Zimmermann-Laband syndrome.

Results: Clinical and radiographic findings supported the diagnosis of Zimmermann-Laband syndrome. The most striking oral findings were gingival enlargement involving the maxillary and the mandibular arches and anterior open bite. Periodontal treatment comprised of Gingivectomy in second and third quadrants. Histopathological evaluation of the excised gingival mass supported the diagnosis of gingival fibromatosis. The patient has been closely followed for the earliest sign of recurrence of gingival enlargement.

Conclusion: Dental practitioners should be alert for the various developmental abnormalities that can occur in patients with gingival fibromatosis as it may indicate the presence of a rare disorder like Zimmermann-Laband syndrome. The successful therapy for gingival fibromatosis depends on correctly identifying the etiological factors and improving the impaired function and esthetic appearance through surgical intervention.

**Keywords:** Developmental abnormalities, fibromatosis, Zimmermann-Laband syndrome

**Introduction**

Gingival fibromatosis is a rare disease with unknown etiological factors, resulting in gingival enlargement.¹⁻³ This form of gingival enlargement usually appears in childhood and this is also known as idiopathic hyperplasia or hereditary gingival fibromatosis. It may appear as isolated entity, or it may be a part of one of the several syndromes.¹ The enlarged tissues are firm, pink and generally non-erythematous except for locally caused inflammation and have no tendency to bleed. The histopathologic analysis might show dense, avascular, bland connective tissue and elongation of rete pegs of the gingival epithelium.⁴

Gingival fibromatosis may be associated with Murray-Puretic-Drescher syndrome (multiple hyaline tumors), Rutherford syndrome (corneal dystrophy), Jones syndrome (sensorineural hearing loss), Ramon syndrome (hairy trunk, cherubism, mental and somatic retardation or it may be idiopathic.⁵ One such rare disorder is Zimmermann-Laband syndrome which is characterized by gingival fibromatosis, abnormalities of the nose and ears, hyperextensible joints and striking hypoplastic changes in the nails or in terminal phalanges of the fingers and toes. The condition has an autosomal dominance mode of inheritance⁶,⁷ and may involve a variable phenotype. Other characteristics of this syndrome include macroglossia, thick lips, speech defects, hirsutism, retinitis pigmentosa, seizures, spina bifida occulta, high foot arch, hepatosplenomegaly and mental retardation.⁸

Surgical intervention has to be carried out to reduce the excessive gingival tissue to enhance esthetics and function, in spite of gingival fibromatosis being a benign condition. This study reports an isolated case of Zimmermann-Laband syndrome in a 22 year old male patient and its dental management.

**Case Report**

A 22 year old male was referred for assessment of his gingival overgrowth. The main complaint of the patient was swelling of the gums since 3 years. It was associated with pain and bleeding from the gums while brushing and eating food. The pain was moderate and continuous in nature. Patient gave a history of similar enlargement of the gums ten years back for which he was operated and the enlargement had resolved completely at that time.

The examination of the patient showed that the patient had thick floppy years with low set and bulbous nose, prominent maxillae, anterior open bite (figure 1), mild hirsutism on his arms and legs, thick eyelashes and eyebrows, hypertelorism, deformed terminal phalanges of the toes and thumbs and hyper extensibility of the
Figure 1: Pre-operative view showing anterior open bite.

Figure 2: Pre-operative view with gingival overgrowth.

Figure 3: Histopathological picture of the excised gingival mass.

Figure 4: 10 months post-operative view (Facial)

Figure 5: 10 months post-operative view (Buccal)
the metacarpophalangeal joints. The patient had low intelligence and no history of medication which could have caused this gingival overgrowth. No signs of hepatosplenomegaly were evident. The intraoral examination revealed extensive pale pink enlargement of the gingiva in both maxilla and mandible (figure 2). The presence of an anterior open bite, high arched and narrow palate and a large tongue was also noted. The diffuse gingival enlargement, involving buccal and palatal gingiva was present between 22 and 23. The overlying mucosa appeared normal but stretched. The gingival overgrowth was lobulated in the upper left quadrant. The enlargement was fibrotic in consistency and bleeding on probing was present. Hard tissue examination revealed grossly decayed 36, root stumps with 46 and occlusal caries with 16. One characteristic finding which was noted in this case was that the patient was diagnosed with primary hypertension with blood pressure being 170/106 mmHg at the first visit and 160/96 mm/Hg, 160/90 mm/Hg and 150/90 mm/Hg at subsequent follow up visits. Patient was then put on Amlodipine 5mg per day.

The diagnosis of Zimmermann-Laband syndrome was performed by a pediatric geneticist based on his medical history and the presence of characteristic manifestations. Treatment planning was done which included non surgical periodontal therapy following which gingivectomy was planned in the second and third quadrant. Scaling and root planing was done at the first visit. It was followed by quadrant wise internal bevel Gingivectomy. The patient was prescribed 0.12% chlorhexidine rinse twice a day post surgically.

The diagnosis of gingival fibromatosis was given after histopathological examination of the excised gingival mass showed hyperplastic orthokeratinised stratified squamous epithelium with elongated rete ridges. The underlying connective tissue showed densely packed collagen bundles, few blood vessels and few lymphocytes. (figure 3)

The patient has been followed periodically for plaque control and no recurrence of gingival enlargement was observed 10 months after periodontal surgery. Considerable improvement of esthetics was successfully achieved.(figure 4, 5)

Discussion and Conclusion

The pathogenesis of gingival fibromatosis is still unknown but an increase in proliferation of the gingival fibroblasts as well as increased collagen synthesis may be involved. Hereditary gingival fibromatosis may exhibit autosomal dominant mode of inheritance. Genetic loci for autosomal dominant modes of gingival fibromatosis have been localized to chromosome 2p21-p22 (HGF-1) and chromosome 5q13-q22 (HGF-2). It has been discovered that a mutation in the son of sevenless-1 (SOS-1) gene is responsible for gingival fibromatosis.

Gingival fibromatosis is a frequent feature of Zimmermann-Laband syndrome. It is reported to occur at birth or within first few months of life and more severely affects the maxilla as compared to the mandible. Other clinical manifestations of this syndrome include enlargement of the soft tissue of the ears and nose, shortening of the terminal phalanges, absence or hypoplasia of the nails and hyperflexibility of the joints. Anterior open bite, hepatosplenomegaly, hirsutism, mental deficiency, high foot arch, spina bifida occulta of the fifth lumbar vertebra and retinitis pigmentosa have also been documented with variable severity.

The existence of gingival enlargement may also be a result of inflammation, pregnancy, leukaemia and response to certain drugs such as phenytoin, diltiazem, cyclosporine A, verapamil and nifedipine. However in these disorders, the gingiva is usually not enlarged or as fibrotic as in hereditary gingival fibromatosis. Therefore a detailed medical history and physical examination should be carried out before a diagnosis of generalized gingival enlargement is given. The various types of acquired or hereditary generalized gingival enlargement should be considered in the differential diagnosis.

A surgical treatment is considered when the gingival enlargement is responsible for impairment of esthetics and interferes with function. Conventional Gingivectomy is the most efficacious treatment in such cases. In this case also, surgical treatment improved the esthetics and enhanced the function.

The patient should maintain good oral hygiene for stabilization of the effective treatment as the presence of inflammation and infection can be associated with the risk of recurrence of gingival enlargement as it happened in this case. However, recurrence of gingival fibromatosis in such patients can also be attributed to genetic predisposition. Therefore, it is not possible to predict the long term results of gingival fibromatosis treatment even when associated with good oral hygiene.

Thus to conclude, etiological factors should be identified in cases of gingival fibromatosis and surgical intervention results in improving enhanced function and esthetic appearance.
References


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