CASE REPORT

GINGIVAL FIBROMATOSIS ASSOCIATED WITH ZIMMERMANN LABAND SYNDROME - A RARE ENTITY

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ABSTRACT

Zimmermann Laband syndrome or Laband syndrome or Laband disease (ZLS) is a rare inherited autosomal dominant disease characterised by gingival fibromatosis, hypertrichosis, aplasia or hypoplasia of nails or terminal phalanges of hands and feet. However, the syndromic features of ZLS are highly complicated and variable. This paper, reports a case of ZLS with few manifestations and its dental management.

KEY WORDS

Zimmermann Laband Syndrome (ZLS); Gingival fibromatosis

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INTRODUCTION

ZLS is a disorder characterized by gingival fibromatosis, abnormalities of the nose and/or ears, and absence and/or hypoplasia of the nails or terminal phalanges of the hands and feet (Fig.1). Other variable features may include hyper-extensibility of joints, mild hirsutism, hepatosplenomegaly, and mental retardation. The first case was described by Zimmermann[1] in 1928 and Laband et a1.[2] described the first familial occurrences in 1964.

Individuals with gingival fibromatosis presents with a generalized benign, slowly progressive, nonhemorrhagic, painless, fibrous enlargement of the buccal and lingual aspects of the attached and the marginal gingiva. This is most noticeable at the time of eruption of the dentition and produce a cosmetic defect which may completely overgrow the occlusal surfaces of the teeth. Thus its impairmentmay be considered as a definite reason for surgical intervention in order to reduce the excessive gingival tissue. Most of the cases have no apparent associated systemic disorders.

The histopathologic analysis of gingival fibromatosis might show dense, avascular, bland collagenic connective tissue and elongation of rete processes of the gingival epithelium.[3] Hereditary gingival fibromatosis may be associated with Murray-Puretic-Drescher syndrome, Rutherford syndrome, Jones syndrome, Ramon syndrome, or Zimmermann Laband syndrome, or it may be idiopathic[4].

This study, reports the dental management of one such patient with idiopathic gingival fibromatosis associated with few features of ZLS.

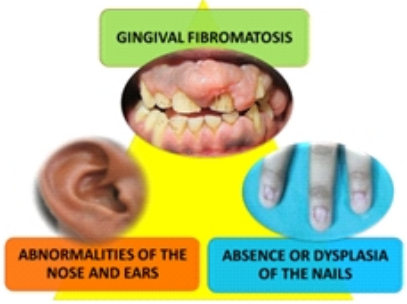


Fig. 1 - Pathognomic triad

CASE REPORT

A 21-year old male patient reported to the Department of Periodontics, with the complain of gingival overgrowth. He had pronounced generalized asymmetric gingival fibromatosis affecting buccal and lingual/palatal aspects both in the maxilla and mandible. The gingival enlargement was noted by his mother at the age of 7 years.

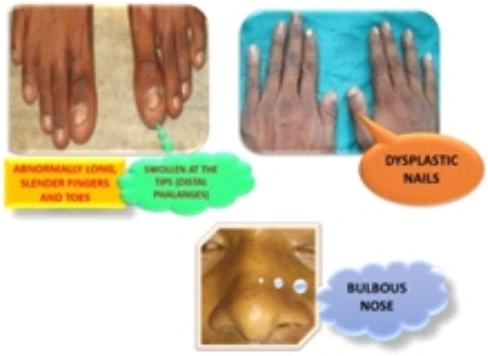


Fig. 2 -Showing extraoral features

He presented with the following clinical features like thick floppy ears with bulbous soft nose, prominent maxillae, thick lips, thick eyebrows, dysplastic toenails, deformed terminal phalanges of the toes and thumbs, and hyper extensibility of the hands and foot (Fig. 2) which was confirmed by the radiograph of both hands and legs. No hepatomegaly, splenomegaly or hypertrichosis were noted. The patient had mild mental retardation. There was no history of any medication known to cause gingival fibromatosis. Haematological findings were within normal range. The diagnosis of Zimmermann-Laband syndrome was made based on his medical history and the presence of the characteristic manifestations.

The intraoral examination revealed an extensive pale-pink firm enlargement of the gingiva in both maxilla and mandible (Fig.3). The enlargement also involved the palate which affected his speech. Presence of high-arched and narrow palate and macroglossia was noted.

OPG revealed spacing in between the tooth and carious upper right second premolar and first molar and root stump in lower left molar and an impacted upper right third molar(Fig. 4).

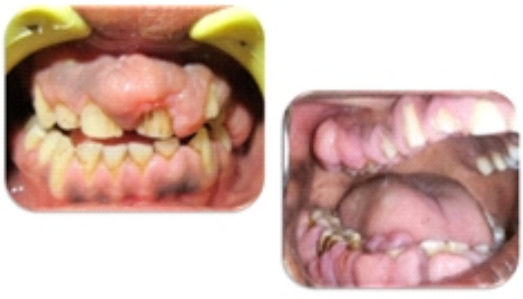


Fig. 3 - Intraoral view showing gingival enlargement in maxilla and mandible

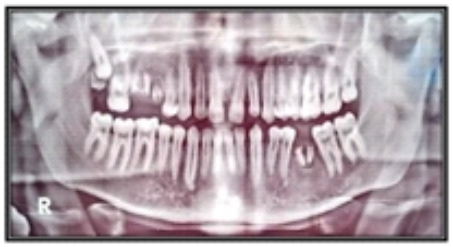
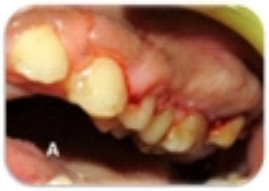


Fig. 4 - Pre -operative orthopantomogram

SURGICAL PROCEDURE

In an attempt to facilitate the normal functioning and improve the aesthetics, Gingivectomy surgeries were performed under local anaesthesia quadrant-by-quadrant in an interval of approximately 4 weeks between surgeries.

Furthermore, Resective osseous surgery was planned for left upper teeth region after all the gingivectomy procedures were completed where first sulcular incision was given along with the vertical incision (Fig. 5). Bulbous bony contour was noticed after reflection of full thickness flap Vertical grooving, radicular blending, flattening of interdental bone and gradualisation of marginal bone were done subsequently (Fig. 6). Flap was then closed with suture and periodontal pack given. A 0.12% chlorhexidine gluconate rinse was prescribed for administration twice a day post-surgically after each surgery.



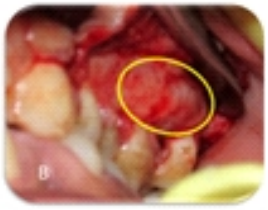
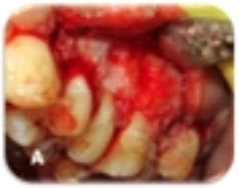
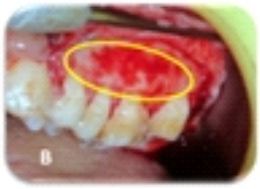


Fig. 5 - Intraoral view after sulcular incision and vertical releasing incision. Intraoral view showing bulbous bony contour following reflection.





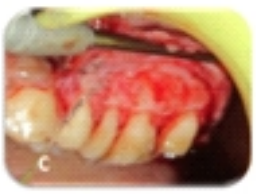


Fig. 6 - Intraoral view showing vertical grooving. Intraoral view showing radicular blending. Intraoral view showing Flattening of interproximal bone and granularization of marginal bone.

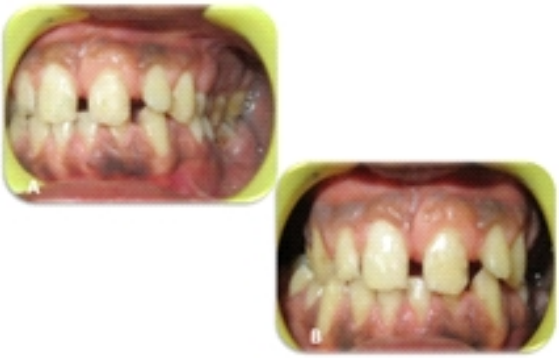


Fig. 7 - Post-operative view after 18 months granularization of marginal bone.

The histopathological examination of the excised tissues, revealed a non-keratinizing stratified squamous epithelium, with elongated rete pegs. Beneath the epithelium there were dense bundles of collagenous fibrous tissue. Due to increased collagen fibre bundle, compression of blood vessels were noted. A pathological diagnosis of gingival fibromatosis was given.

The patient has been followed periodically for plaque control and no recurrence of gingival hyperplasia was observed 18 months after periodontal surgery (Fig. 7). Considerable improvement of aesthetic and function was successfully achieved.

DISCUSSION

Zimmermann–Laband syndrome is a very rare genetic disorder which appears to have autosomal dominant inheritance and variable expressibility.[2] Both the sexes may be affected in equal ratio. Eleven cases from two families in the literature were well documented with evidence of autosomaldominant inheritance. A total of forty cases have been reported till 2010.[5] This syndrome is coded with phenotype MIM number 135500. The gene responsible for ZLS is located in 3p14.3 and implicates four likely candidate genes in this region. CACNA2D3 gene encoding avoltage dependent calcium channel, LRTM1 gene of unknown function, WTN5A gene encoding a secreted signalling protein, and ERC2 gene, which encodes fora synapse protein.[6]

Gingival fibromatosis is one of the most important and consistent features of ZLS. It may exist as an isolated finding or as part of a genetic syndrome.

The pathogenesis of gingival fibromatosis is still unknown, but an increase in collagen synthesis and increase in the proliferation of gingival fibroblasts may be involved. It is known that gingival fibromatosis may exhibit autosomal dominant inheritance[7].

In diagnosis of gingival fibromatosis, a detailed medical history and physical systemic evaluation is necessary rule out various other types of acquired or hereditary generalized gingival enlargement that can occur as a result of inflammation, pregnancy, leukaemia, and as a response to certain drugs such as phenytoin, diltiazem, cyclosporine, verapamil, and nifedipine. But in such cases, the gingiva is usually not as enlarged or as fibrotic as in hereditary or idiopathic gingival fibromatosis[4].

In this case reported here, the most efficacious method was the conventional gingivectomy procedure which allowed aesthetic improvement and speaking, and hence improved the psychological status of the patient. Presence of inflammation and infection can be associated with a risk of recurrence of the gingival enlargement. Thus, maintaining good oral hygiene is important in obtaining the stabilization of the treatment.

CONCLUSION

In conclusion, ZLS is not a life-threatening disorder but needs a comprehensive medical and family history and systemic examination that is necessary for correct diagnosis, treatment and prevention of any complications.

The first step in successful therapy related to the oral hygiene is the surgical correction of gingival fibromatosis, to improve the impaired function and aesthetic appearance which may further establish the necessary conditions for an efficacious orthodontic treatment.

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