EDELWEISS PUBLICATIONS
OPEN ACCESS

https://doi.org/10.33805/2572-6978.116
Volume 3 Issue 1 | PDF 116 | Pages 3

Dental Research and Management

Case Report ISSN 2572-6978

Zimmerman-Laband Syndrome Oral Manifestations A Case Report

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Citation: Mathew AL, James M and Maria H. Zimmerman-laband syndrome oral manifestations a case report (2019) Dental Res Manag 3: 3-5

Received: Dec 12, 2018
Accepted: Jan 08, 2019
Published: Jan 15, 2019

Abstract

Zimmerman-Laband syndrome was reported by Zimmerman in the year 1928 which is a rare inherited autosomal dominant disease characterized by generalized enlargement of the attached and marginal gingiva, abnormalities of nose, ear, deformities of nails, joint hyperextensibility, hepatosplenomegaly, skeletal abnormalities and occasional mental retardation. Idiopathic gingival enlargement is usually evident after the eruption of the permanent teeth. Both sexes are equally affected. Genetic loci for autosomal dominant modes of gingival fibromatosis is localized to chromosome 2p21p22 (HGF-1) and chromosome 5q12-q22 (HGF-2). This syndrome is not a life threatening disorder. Hereditary gingival enlargement is associated with syndromes like Rutherford syndrome, Zimmerman-Laband syndrome, Murray-Puretic-Drescher syndrome, Cross syndrome and Ramon's syndrome. The most important feature of this syndrome is gingival enlargement appearing early in childhood. Idiopathic gingival enlargement is usually evident after the eruption of the permanent teeth. Surgical correction of gingival fibromatosis is recommended, although there is no information on the permanence of the results of this treatment. We present a case of a 14 year old female patient with Zimmerman-Laband syndrome. Gingivectomy was carried out in the upper and lower anterior region there by exposing the impacted teeth.

Keywords: Gingival enlargement, Autosomal dominant, Zimmerman-Laband syndrome

Introduction

Hereditary gingival enlargement is associated with syndromes like Rutherford syndrome, Zimmerman-Laband syndrome, Murray-Puretic-Drescher syndrome, Cross syndrome and Ramon's syndrome [1]. Zimmerman-Laband syndrome is a rare entity characterized by generalized enlargement of the attached and marginal gingiva, abnormalities of nose, ear and deformities of nails, joint hyperextensibility, hepatosplenomegaly, skeletal abnormalities and occasional mental retardation [2]. The prevalence of this syndrome is 1 in 10000000. Forty four patients have been reported till date. This syndrome has highly variable clinical expressions. Gingival fibromatosis is characterized by slow and progressive enlargement of maxillary and mandibular gingiva. It can be presented as an isolated feature or as a part of syndrome. This syndrome is not a life threatening disorder.

Etiology

The genetic basis is unknown. Mapping of breakpoints of two translocations, t (3; 8) and t (3; 7) found in two patients with typical clinical features of Zimmermann-Laband syndrome defined a common breakpoint region located in 3p14.3 but the lack of a specific coding-sequence lesion in the common region suggests that either some other type of genetic defect in this vicinity, or an alteration elsewhere in the genome, could be responsible for ZLS. Autosomal dominant inheritance has been suggested.

The most important feature of this syndrome is gingival enlargement appearing early in childhood. Idiopathic gingival enlargement is usually evident after the eruption of the permanent teeth [3]. The differential diagnosis includes other defined syndromes of hirsutism and coarsening of the face. Isolated gingival fibromatosis has been documented as a dominantly transmissible trait. Surgical correction of gingival fibromatosis is recommended, although there is no information on the permanence of the results of this treatment, surgical removal of the hyperplastic fibrous tissue and appropriate orthodontic treatment to improve esthetic appearance and eruption of the non-erupted teeth. Our case reports a 14 year old female patient with Zimmerman-Lanband syndrome the oral manifestation and treatment.

Case Report

A 14 year old female patient reported the Department of Oral Medicine and Radiology with the chief complaint of difficulty in chewing, enlarged gums and also about the un-erupted front teeth, inability to close the lips causing esthetic concern. Patient's parents reported that the gingival growth gradually increased in size to the present condition. History of recurrent cellulitis of leg was reported.

Family history revealed that her elder brother is also suffering with the same condition hepatic fibrosis with portal hypertension. Her parents were normal with no features of this syndrome. Patient had normal intelligence and is doing well in her studies. On general examination well was moderately built and nourished with normal vital signs. Systemic evaluation revealed no obvious internal organ abnormalities.

On extra oral examination showed the presence of thick eyebrows, bulbous nose and floppy ears, thick and incompetent lips as shown in **Figure 1.**

Figure 1: Extraoral photograph.

Patient also had plantar keratosis shown in **Figure 2**.

Figure 2: Plantar keratosis.

Intraoral examination revealed severe gingival enlargement of the maxillary and mandibular anterior region with high arched palate **Figures 3, 4**.

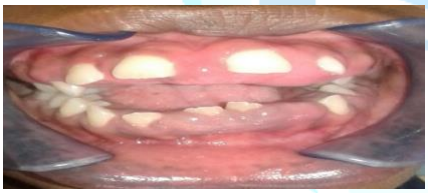

Figure 3: Intraoral photograph showing gingival enlargement.

Figure 4: Showing high arched palate.

Due to the gingival enlargement the upper and lower anterior teeth were embedded in the gingiva with partially erupted upper and lower central incisors. Gingiva was firm, leathery in consistency with lobulated and pebbled surface. There was melanosis of the buccal and labial mucosa. A panoramic radiograph was taken revealed the presence of all the permanent teeth arranged irregularly in the anterior region **Figure 5**.

Hematological and biochemical investigations were within the normal limits. Ultrasound revealed no signs of hepatosplenomegaly.

Patient was diagnosed with Zimmerman-Laban syndrome based on the clinical features and pediatric consultation done in a medical college where the patient was worked up initially.

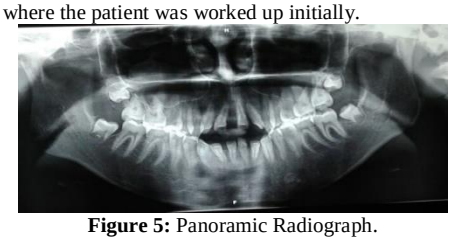

Figure 5: Panoramic Radiograph.

No gingival biopsy was done in our case. Treatment was planned to surgically expose the embedded teeth by gingivectomy under general anesthesia. Gingivectomy was performed with the objective of restoring the normal contour of the gingiva as well as exposing all the embedded teeth in order to improve the esthetics of the young girl **Figures 6, 7**.

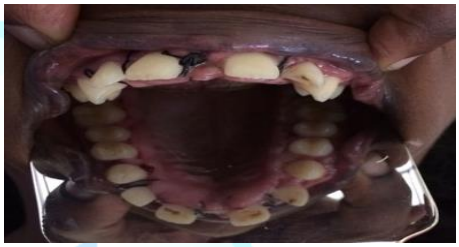

Figure 6: Post-operative photograph showing exposed upper teeth.

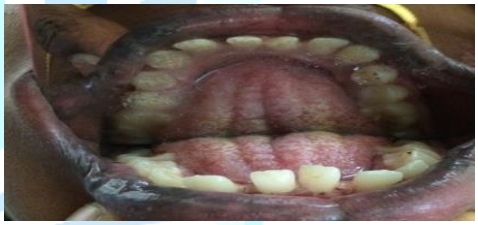

Figure 7: Post-operative photograph showing exposed lower anterior teeth.

Patient has been advised to go in for orthodontic correction of teeth after complete healing of the gingiva. Patient was asked to maintain a good oral hygiene by advising plaque control measures and regular follow up. Regular 0.12% chlorhexidine gluconate mouth rinse was prescribed. Patient is on regular follow up for assessing the recurrence.

Discussion

Zimmerman-Laband syndrome is a very rare syndrome with autosomal dominant mode of inheritance. Both sexes are equally affected. Genetic loci for autosomal dominant modes of gingival fibromatosis is localized to chromosome 2p21p22 (HGF-1) and chromosome 5q12-q22 (HGF-2) [4]. This syndrome is not a life threatening disorder. The most important feature of this syndrome is gingival enlargement appearing early in childhood. Swaki et al [5] stated that the major clinical findings of this syndrome will be gingival fibromatosis, hyperplasia or absence of terminal phalanx or nails of hand and feet, bulbous soft nose, thick lips, large ears and enlargement of soft tissues of face.

Hypoplasia of the nails and hyper flexibility of the joints are also reported to be associated with hereditary gingival fibromatosis [6]. Hepatomegaly was also reported to be associated with this syndrome [7]. Other clinical syndromic presentation with phenotypic overlap

includes Cowden syndrome, Cross syndrome, Gohlich-Ratmann syndrome, Avani syndrome and I-cell disease [8].

Our patient was concerned about the esthetic and functional needs indicated the need for surgical correction. The gingival enlargement cannot be cured but may be controlled with varying degrees of success for improving the esthetics and normal function. Future research could be directed towards the therapeutic field that could change dramatically by one of the recombinant DNA and monoclonal antibody technology for prevention of these syndromes. Early detection and timely recognition of this syndrome allows adequate dental care at periodic intervals to improve the overall quality of the life of such patients [9].

Conclusion

To conclude this syndrome is not a killer disease but it needs proper medical, and systemic evaluation is needed for the correct diagnosis, treatment and complications that can occur in such patients. Dental practioners should be aware of the developmental abnormalities that may occur in patients with gingival fibromatosis as this may indicate the presence of a rare disorder like Zimmerman-Laband syndrome. A comprehensive medical history and physical systemic evaluation are essential for correct diagnosis and treatment of these cases.

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