ZIMMERMANN–LABAND SYNDROME – A CASE REPORT

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Abstract

The Zimmermann–Laband syndrome, also often termed as Laband–Zimmermann syndrome or Laband's syndrome, was first described by Zimmermann in the year 1928. It is one of the rare inherited autosomal dominant disorders `clinically characterized by pathognomonic triad of extensive gingival enlargement, abnormalities of the nose and ears along with dystrophic nails. The syndromic characteristics are highly variable and is quiet complicated in nature. The present study describes a case report of a 17 years old female, presenting features of Zimmermann–Laband syndrome.

Key Words Gingival fibromatosis, Zimmermann–Laband syndrome

INTRODUCTION

Gingival fibromatosis may manifest clinically as slow and progressive gingival enlargement of maxillary and mandibular arch. It can be observed either as an isolated feature or as a part of an associated syndrome. Various syndromes are associated with gingival fibromatosis such as Zimmermann-Laband syndrome, Murray–Puretic–Drescher syndrome, Cross syndrome, Rutherford syndrome, Jones syndrome, Cowden syndrome, Prune Belly syndrome and Ramon's syndrome. Zimmermann-Laband syndrome is usually associated with a classic triad of generalized enlargement of gingiva, abnormalities of nose and dystrophic nails[1]. Besides this classic triad many clinical signs like hyppertrichosis, joint hypermobility, protruberant lips, thick eyebrows, maxillary prognathism, arched foot may be present.

Zimmermann first described this syndrome in the year 1928.

CASE REPORT

A 17-year-old female patient reported to the Department of Oral & Maxillofacial Pathology, Dr. R. Ahmed Dental College & Hospital, Kolkata, with a complaint of gingival swelling causing esthetic problems. Proper questionnaire and history records revealed that the gingival growth is enlarging gradually and is present since the time of eruption of permanent dentition. Extraoral examination of the patient showed the presence of maxillary prominence, thick eyebrows, bulbous and large nose and thick protruberant lips. Besides, the patient had also presented with hypertrichosis extremities and recurrent nail infection.

Intraoral examination revealed diffuse and severe gingival enlargement in the maxillary and mandibular arch so much so that most of the teeth were covered with the gingiva itself. Patient also reported that she had delayed exfoliation and eruption of deciduous and permanent teeth respectively.

Thorough clinical examination revealed no hepatosplenomegaly and any other clinical significant finding.

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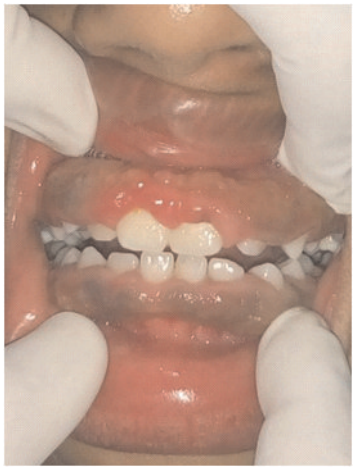

Fig. 1: Generalized gingival enlargement

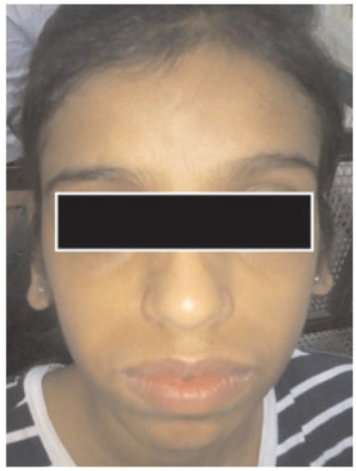

Fig. 2: Frontal profile of the patient showing prominent bulbous nose, thick protruberant lips as well as thick eyebrows.

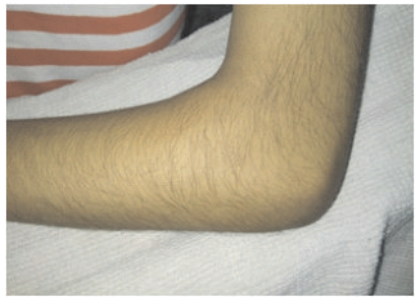

Fig.3: Hypertrichosis in hands

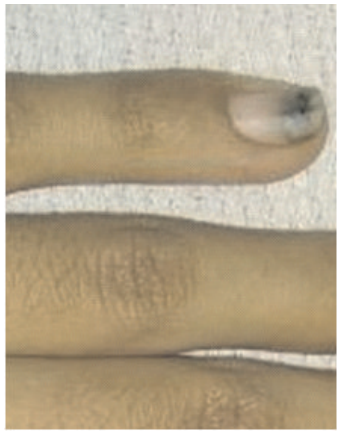

Fig 4. Dystrophic nails in little finger

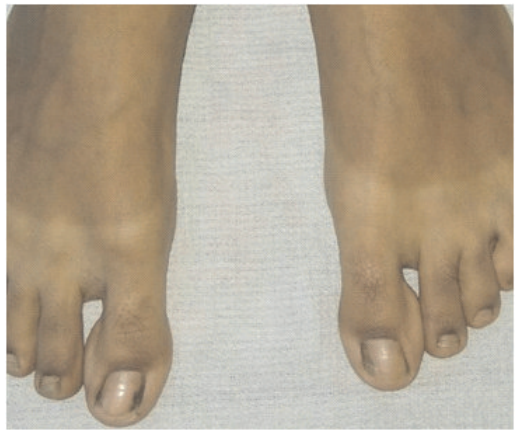

Fig. 5. Dystrophic nails of great toe

Differential diagnosis with other syndromes associated with Gingival fibromatosis was excluded in the following manner.

As patient had no subcutaneous tumor and muscle weakness, so Murray–Puretic–Drescher syndrome was excluded.

Absence of corneal opacity and failure of tooth eruption also do not favour the diagnosis of Rutherford Syndrome.

In Ramon Syndrome, besides Gingival fibromatosis, patients usually present with a history of seizures, fibrous dysplasia of jaw bones, stunted growth, but apart from gingival fibromatosis, all other clinical findings were absent in this patient.

Cowden syndrome was not considered because patient had no oral and dermal papules, acral keratosis, cutaneous neuromas and goitre.

Patients with Cross syndrome usually present with blond hair with yellow grey metallic sheen, small eyes with cloudy corneas, jerky nystagmus along with physical retardation which were absent in the present case.

Patient had no complaint of hearing loss, so diagnosis of Jones syndrome was also ruled out.

Considering all these clinical findings, the diagnosis of Zimmermann–Laband syndrome has been made.

Patient was referred to the Dept. of Periodontia for necessary treatment.

DISCUSSION

Zimmermann–Laband syndrome is one of the very rare autosomal dominant disorder, but not a life-threatening one. Both males and females have same predilection. Genes responsible for Zimmermann-Laband syndrome are located in 3p14.3. These candidate genes are CACNA2D3 gene encoding a voltage dependent calcium channel, LRTM1 gene of unknown function, WTN5A gene encoding a secreted signalling protein, and ERC2 gene, which encodes for a synapse protein[2].

The most clinically appreciable finding in this

syndrome is gingival fibromatosis which may appear at an early age. Various differential diagnosis of generalized gingival enlargement exists ranging from altered physiology like pregnancy to pathology like leukemia, drugs such as phenytoin, diltiazem, cyclosporine, verapamil, and nifedipine induced gingival enlargement. should be excluded. But, in these situations, the gingiva is not so much enlarged or fibrotic as in hereditary gingival fibromatosis.[3]

Confirmational diagnosis of this syndrome demands the mutation of the aforementioned candidate genes. Further genetic studies is advocated to understand the real pathophysiology of this disease linking the various clinical signs observed in this syndrome.

Surgical correction of gingival fibromatosis is routinely recommended as far as the aesthetics and oral hygiene is concerned. Although there may be a chance of recurrence. Some authors also suggest that ideal treatment to prevent recurrence of gingival growth is extraction of all the teeth followed by prosthesis. A team comprising of multiple specialities like dermatologist, general medicine, oral & maxillofacial surgeon and Periodontist is needed solely to render quality treatment to the patient.

CONCLUSION

Zimmermann–Laband syndrome is an autosomal dominant disorder which needs to be diagnosed early by thorough clinical examination. All differential diagnosis should be properly excluded prior to establishment of diagnosis of Zimmermann Laband syndrome. Though it is not a life-threatening syndrome, but it interferes with the patient's aesthetics. So, correct diagnosis followed by right periodontal treatment is very much needed.

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