An Extremely Rare Presentation of Zimmermann–Laband Syndrome in a Twin

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Abstract

Zimmermann–Laband syndrome is a rare condition which manifests as hereditary gingival tissue hypertrophy along with other clinical features. Till date, only few cases have been reported in the literature. This case report focuses on an extremely rare presentation of Zimmermann–Laband syndrome in a twin. There has not been a single case report of Zimmermann–Laband syndrome in twins.

Keywords: Sisters, twins, Zimmermann–Laband syndrome

Introduction

Hereditary gingival fibromatosis is a rare condition which manifests as gingival tissue hypertrophy. Even erupting teeth become buried in varying degrees beneath the hypertrophic tissues. This gingival hypertrophy may encroach onto the hard palate. Other names used to describe this condition are elephantiasis gingivae, idiopathic hyperplasia of the gums, fibromatosis gingivae, and diffuse fibroma of the gums.[1] In 1964, Laband *et al*. reported a family in which the mother and five of her seven children exhibited gingival fibromatosis, splenomegaly, and skeletal and soft-tissue abnormalities. Two such cases had already been described by Zimmermann[2] in 1928 and one by Jacoby *et al*.[3] in 1940. The syndrome is apparently an autosomal dominant trait, as confirmed by Alavandar,[4] who observed five affected persons in three generations. The present case adds new information on the syndrome.

Case Report

The patient, 24-year-old female, reported to our clinic for gingival enlargement. She was born as one of twin females after full-term uncomplicated pregnancy. At conception, her mother was 32 and her father 34-year-old and both were healthy nonconsanguineous Indian parents. Her clinical examination revealed enlargement of the maxillary and mandibular gingiva including hard palate, partially or completely covering the crowns of maxillary posterior teeth [Figure 1], multiple carious teeth, prominent labial fullness on extraoral examination, mild mental retardation, hirsutism,[5] large cartilaginous parts of ears and nose, dysplastic fingernails, aplasia of nails of last two toes on both feet, and pes cavus deformity was present. Preoperative radiographs showed abnormal position of maxillary posterior teeth[Figure 2]. Her twin sister [Figure 3], mother, and father were examined and appeared normal. Radiographic examination of foot revealed hypoplastic distal phalanges of the fourth and fifth toes [Figure 4]. Cardiovascular, respiratory, gastrointestinal, ophthalmologic, and blood abnormalities were not found, the liver was normal sized and the spleen not palpable. Informed consent was obtained from all individual participants included in the case report.

The patient was treated with gingivectomy and gingivoplasty under local anesthesia to improve esthetics and facilitate oral health. Postoperative clinical photographs showed improvement of esthetics [Figure 5]. Histopathological examination of excised tissues revealed an acanthotic, nonkeratinized, stratified squamous epithelium, with elongated rete pegs. Beneath the epithelium, there were dense bundles

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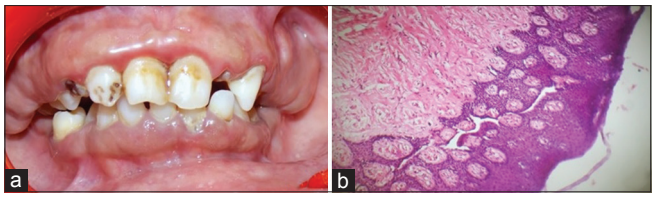

Figure 1: (a) Intraoral clinical photograph showing gingival enlargement. (b) Histopathological picture of excised gingiva

Figure 3: Clinical photograph of patient affected with Zimmermann–Laband syndrome (left) with her twin sisters (right)

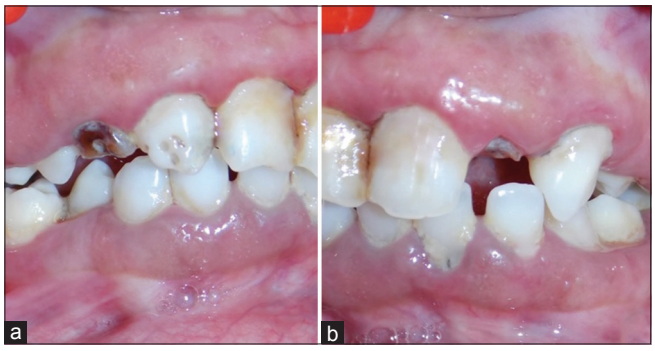

Figure 5: (a) Postoperative – right-side intraoral clinical photograph. (b) Postoperative – left-side intraoral clinical photograph

of collagenous fibrous tissue which confirmed the diagnosis of gingival fibromatosis.

Discussion

Zimmermann–Laband syndrome is a very rare genetic disorder. The main facial features are marked gingival

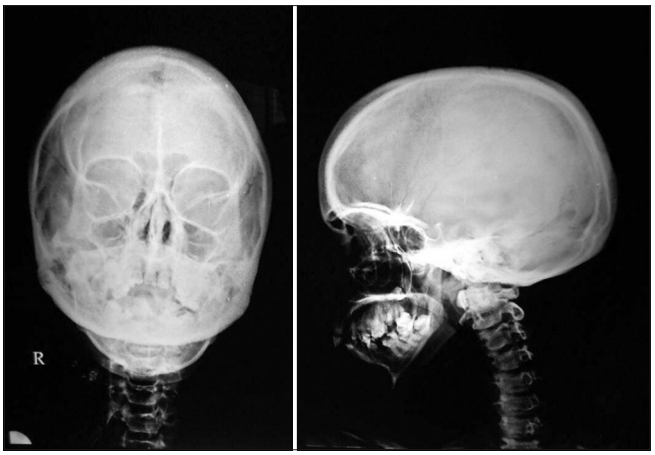

Figure 2: Preoperative radiograph

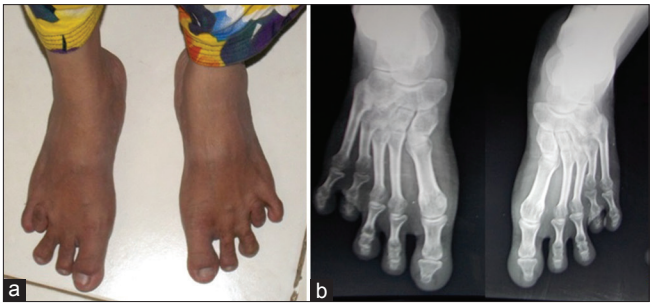

Figure 4: (a) Clinical photograph of feet showing hypoplastic distal phalanges of the fourth and fifth toes, dysplastic fingernails, and aplasia of nails with the last two toes of both the feet. (b) Radiographic photograph showing hypoplastic distal phalanges of the fourth and fifth toes

fibromatosis, thick, floppy ears, and bulbous soft nose. Other clinical findings are hepatosplenomegaly, absence or dysplasia of nails and/or terminal phalanges, hyperextensibility of joints, especially the metacarpophalangeal, and rarely, hypertrichosis and mental retardation.[6] The syndrome appears to have autosomal dominant inheritance and variable expressivity.[1,4] Our case manifests most of the characteristics of the disorder.

A young patient with Zimmermann–Laband syndrome has been described in the present report. However, presentation of Zimmermann–Laband syndrome in twins with one of the twins being normal has rarely been described in the literature. The patient did not manifest hepatosplenomegaly. According to Gorlin *et al*.,[7] enlargement of the liver and spleen has been specifically denied in his cases by Zimmermann,[2] and it was variably expressed in the patients of the reported families.[1,4] Mental retardation is not a typical feature of the syndrome. It has been described in two cases.[2,8] In our case, there was mild mental retardation.

Conclusion

Zimmermann–Laband syndrome is a rare genetic disorder. Although quite a few cases have been reported until now, manifestation of this syndrome in one of two twins with the

other being clinically normal has not been documented till date which makes this particular case unique and extremely rare.

Zimmermann–Laband syndrome is not a life-threatening disorder. Surgical correction of gingival fibromatosis is recommended.[6] Gingivectomy and gingivoplasty in our case will establish the necessary conditions for efficacious orthodontic treatment, which may improve the facial esthetics. However, the possibility of retreatment cannot be overlooked in these types of cases. The ultimate goal of the treatment is to improve esthetics and function. This in turn will boost the confidence of the patient reducing social embarrassment of the patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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