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Original Research Paper

Periodontology

ZIMMERMANN-LABAND SYNDROME: A RARE CASE REPORT

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ABSTRACT Zimmermann-Laband syndrome (ZLS) is a very rare condition characterized by gingival fibromatosis, coarse facial appearance and malformed nails or distal phalanges of hands/ feet and sometimes hepatosplenomegaly1. The term Zimmermann-laband syndrome was for the first time given by Carl Jacob Witkop. It is attributed to be because of autosomal disease characterized by a classical triad of extensive gingival enlargement (gingival fibromatosis), abnormalities of the nose and ears, abnormally developed nails or terminal phalanges of the hands and feet. The present case report is of a 12year old boy who was diagnosed as ZLS based on the classical sign of gingival overgrowth.

KEYWORDS : labands syndrome, gingival overgrowth, nail dysplasia, gingivectomy.

Case report

A twelve-year male child reported to this clinic with the chief complaint of increase in size of the gums in the lower anterior region causing difficulty in chewing and giving an unpleasant smile since 2 months. On eliciting the history revealed that the increase in size was gradual and had reached the present size. The patient did not reveal any familial history. The child was otherwise systemically healthy, not on any medication for any known reason.

On general examination of the patient was moderately built with a normal gait. On extraoral examination revealed the presence of thick eyebrows, bulbous nose and increase in the size of the lips. On examination of his extremities, showed the absence of nails in the fingers. The nails of the toes were partially missing [Fig 1,2]. The examination of extremities showed irregularity in the size of both the feet [Fig 3].

Intraoral examination revealed mixed dentition stage, absence of any decayed teeth. Soft tissue examination showed severe gingival overgrowth in the lower anterior region with macroglossia. The gingival tissue on examination was firm and fibrous in consistency. More in the lower anterior region. Mild increase in size in the posterior, all four quadrants were involved. Deep pseudo-pockets were seen in the anterior mandibular teeth. Minimal bleeding on probing was also present.

An ortho-pantograph was taken. It showed a mixed dentition stage arranged in a irregular manner. [Fig 4]. The routine blood investigation and urine examination revealed findings which were within the normal range. Ultrasonography showed no signs of any abnormality in the size of liver as well as the spleen. The child was diagnosed with Zimmermann–Laband syndrome which was confirmed based on the clinical features and physical examination.

Treatment was planned based on the chief complaint, initial phase one therapy included deep scaling and oral hygiene maintenance, followed by gingivectomy to improve aesthetics and function. The surgery was performed with a classical reverse bevel incision. [Fig5,6] Gingivectomy was performed under 2 % lidocaine (local anesthesia) the excised tissue sample was sent for histological analysis. [Fig 7] Periodontal pack (Coe pack) was placed. Patient was prescribed with mild analgesic (paracetamol) and an antiseptic mouth wash (0.2% chlorhexidine gluconate) for 10 days.

Histological examination of gingival tissue excised showed hypertrophy of stratified squamous epithelium with elongated rete pegs. The blood vessels were reduced in number and compressed due to increased collagen. No evidence of dysplasia was noted. The patient was periodically recalled for maintenance therapy. [Fig 8]

Discussion

A rare syndrome known as Zimmermann–Laband syndrome or Laband's syndrome was first reported by Zimmermann in 1928. It is known to be an autosomal dominant disease characterized by a triad of extensive gingival overgrowth [Fig 9], abnormalities of the nose and ears, absence or hyperplasia of the nails or terminal phalanges of the hands and feet. Other findings include hepato-splenomegaly and hyper extensible metacarpophalangeal joints. Intellectual deficit is occasional and usually mild to moderate. The genetic changes are unknown. Studies have shown two translocations t(3;8) and t(3;17) found in a few patients with ZLS.[3]

The most common and consistent feature of ZLS is gingival overgrowth appearing in early childhood. This condition is not often seen at birth.[4] In the diagnosis, systemic evaluation is necessary to differentiate gingival fibromatosis from various other types of acquired generalized gingival overgrowths that can occur from inflammation, pregnancy, leukemia, and drug induced gingival overgrowth caused by phenytoin, diltiazem, cyclosporine, verapamil, and nifedipine. In such cases, the gingiva is usually not as enlarged or as fibrotic as in hereditary gingival fibromatosis.[5]

Sawaki *et al*.,[6] confirmed the clinical features of ZLS as gingival overgrowth, hyperplasia or absence of the nails of hand and feet, bulbous soft nose, thickened lips, elongated ear. Other Syndromes such as Murray–Puretic– Drescher presents with hyaline fibromas, osteolysis of terminal phalanges, recurrent infection, stunted growth, and early death. It is partially similar to Jones Syndrome, which otherwise is a genetically inherited disease first described in 1977.[7] Jones syndrome is associated with slow deafness. In cases where gingival fibromatosis is associated with nasal as well as nail defects it may be characterized as ZLS. Our patient did not show embedded supernumerary teeth, unlike Wynne *et al* [8]. in 1995 reported with gingival fibromatosis associated with hearing deficit and supernumerary teeth.

Treatment in cases with ZLS is gingivectomy and orthodontic intervention where ever required to improve aesthetic appearance and eruption of the non-erupted teeth. Gingivectomy can be carried out using the classical reverse bevel gingivectomy, as done in this case or by the help of lasers.[9]

Recurrence is very common; hence prognosis is fair. Periodic physical /systemic evaluation is very essential.

Conclusion

The Zimmermann–Laband syndrome is never life-threatening. The present case was diagnosed due to gingival overgrowth and a thorough clinical examination. Dentists too can sometimes play a

very important role in identification of certain systemic disorders and help in early intervention if required.

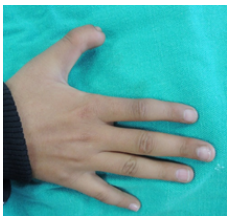

Fig 1. Nails of the toes were partially missing

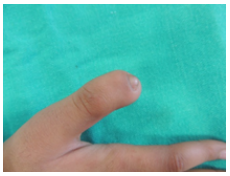

Fig 2. Nails of the toes were partially missing

Fig 3. Extremities showed irregularity in the size of both the feet

Fig 4. Ortho pantograph showing mixed dentition

Fig 5. Classical reverse bevel incision given for gingivectomy

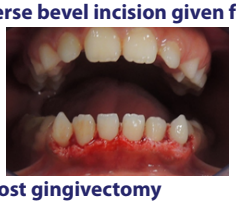

Fig 6. Immediate post gingivectomy

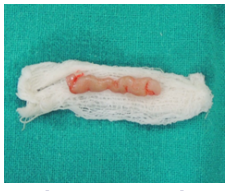

Fig 7. Excised Gingival tissue sample sent for histological examination

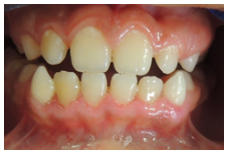

Fig 8. follow up after 6 months

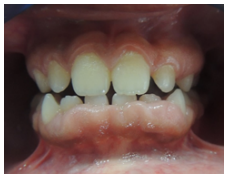

Fig 9. Excessive Gingival overgrowth covering the entire dentition.

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