Zimmermann-Laband Syndrome: Report of a Case with Early Cardiac Complications

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Laband syndrome (LS) is a rare autosomal dominant inherited disorder characterized by coarse facial appearance, gingival fibromatosis, and hypoplasia of the terminal phalanges and nails of hands and feet. Additionally, more variable features include hyperextensibility of joints, hepatosplenomegaly, mild hirsutism, and mental retardation. This paper reports a case study of 22-year-old male exhibiting symptoms of Laband syndrome, including cardiac involvement described only in one other case. Detection and timely recognition of such syndrome associated with gingival fibromatosis allows adequate dental care and cardiac evaluation at periodic intervals is merited to prevent complications and improve the overall quality of life for these patients. Dental practitioners should be alert for developmental abnormalities that may occur in patients with gingival fibromatosis as this may indicate the presence of a rare disorder like Zimmermann-Laband syndrome. A comprehensive medical history and physical systemic evaluation are essential for correct diagnosis, treatment and prevention of cardiovascular complications in future in these patients.

Keywords: Laband syndrome, Gingival fibromatosis, Dysplastic nails, Aortic root dilatation.

On examination patient had generalized gingival enlargement affecting buccal and lingual/palatal aspects both in maxilla and mandible (Fig. 1). Patient also had an enlarged nose but normal ears, anterior open bite (Figs 2A and B), onychodystrophy of most of the nails and hypoplastic terminal phalanges (Figs 3A and B). Patient was mentally retarded. Hypertrichosis in the form of thick curly hair and thick eyebrows was noted. Examination of cardiovascular system revealed primary hypertension with left ventricular hypertrophy. Echocardiography was otherwise normal with mild increase in aortic root diameter. However, per abdominal, ophthalmologic and blood examinations were within normal limits.

CASE REPORT

A 22-year-old Indian male attended Department of Oral Medicine with the complaint of gingival enlargement. His medical history revealed enlargement of maxillary and mandibular gingiva since birth and was treated (gingivectomy) for the same ten years back. On examination patient had generalized gingival enlargement affecting buccal and lingual/palatal aspects both in maxilla and mandible (Fig. 1). Patient also had an enlarged nose but normal ears, anterior open bite (Figs 2A and B), onychodystrophy of most of the nails and hypoplastic terminal phalanges (Figs 3A and B). Patient was mentally retarded. Hypertrichosis in the form of thick curly hair and thick eyebrows was noted. Examination of cardiovascular system revealed primary hypertension with left ventricular hypertrophy. Echocardiography was otherwise normal with mild increase in aortic root diameter. However, per abdominal, ophthalmologic and blood examinations were within normal limits.

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Figs 2A and B: Facial appearance of patient shows coarse facial features with anterior open bite, broad bulbous nose, thick curly hair but normal ears

Fig. 5: Lateral head radiograph showing mild microcephaly with flattening of occiput, increased gonial angle with appearance of relative mandibular prognathism

Figs 3A and B: Fingers and toes of patient with onychodystrophy of nails

Fig. 6: Radiograph of hands showing hypoplasia of several terminal phalanges

Fig. 4: Panoramic radiograph showing diastemas and prominent antegonial angle

Panoramic radiography showed a complete, permanent dentition with few grossly decayed teeth. The anterior open bite was obvious (Fig. 4). Lateral cephalometric radiography showed labial protrusion of anterior teeth in both arches, mild microcephaly, flat occiput and increased gonial angle (Fig. 5). Radiographs of hands and feet showed hypoplasia of several terminal phalanges (Fig. 6).

DISCUSSION

Laband syndrome is a very rare genetic disorder. The main facial features are marked gingival fibromatosis, bulbous nose and thick, floppy ears. Other clinical findings are absence or dysplasia of nails, hyperextensibility of joints especially the metacarpophalangeal joints, hepatosplenomegaly, hypertrichosis and mental retardation.1

Our case manifests most of the characteristics of the disorder and it is sporadic, representing a new mutation. Mapping of the translocation breakpoints of t(3;8) and t(3;17) found in patients with the typical clinical features of Zimmerman-Laband Syndrome defined a common breakpoint region of approximately 280 kB located in 3p14.3, which includes the genes CACNA2D3 and WNT5A.2 In addition to above features patient also manifests...
anterior open bite, left ventricular hypertrophy and mild increase in aortic root diameter in echocardiogram (aortic orifice value is 39, normal range: 20-37). In one of the case report cardiomyopathy and dilatation of the aortic root and arch, was also described in this syndrome.\textsuperscript{4}

The patient did not manifest hepatosplenomegaly. However, this finding was variably expressed in the various reported cases.\textsuperscript{5} Hirsuitism, in our case was manifested as thick curly hair and thick eyebrows. Mental retardation which is not a typical feature of this syndrome was seen in our case. In one of the cases of Zimmermann-Laband syndrome, bilateral developmental cataract was reported.\textsuperscript{6} However, this feature was absent in our case.

Laband syndrome is not a life threatening disorder. The successful therapy for gingival fibromatosis depends on correctly identifying the etiological factors and improving the impaired function and esthetic appearance through surgical intervention and adjunctive orthodontics, although there is no information on the permanence of the results. Our patient already had surgical correction of gingiva ten years back but over the period of time gingival growth reoccurred. This patient was managed with antihypertensives for the control of hypertension and gingivectomy for function and esthetics.

Dental practitioners should be alert for developmental abnormalities that may occur in patients with gingival fibromatosis as this may indicate the presence of a rare disorder like Zimmermann-Laband syndrome. A comprehensive medical history and physical systemic evaluation are essential for correct diagnosis, treatment and prevention of cardiovascular complications that can occur in such patients.

REFERENCES