Zimmermann-Laband Syndrome: A Case Report

Sawaki K * / Mishima K ** / Sato A *** / Goda Y **** / Osugi A ***** / Nakano M ******

Zimmermann-Laband syndrome is a very rare disorder characterized by gingival fibromatosis, abnormalities of soft cartilages of the nose and/or ears, hypoplastic or absent nails and terminal phalanges, joint hypermobility, hypatoslenomegaly, mild hirsutism and learning difficulties. Early presentation of Zimmermann-Laband syndrome in a newborn has rarely been described. This paper describes a newborn patient with Zimmermann-Laband syndrome.

Keywords: *gingival fibromatosis, gingival enlargement, Zimmermann-Laband syndrome, children.*
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INTRODUCTION

Zimmermann-Laband syndrome (OMIM 135500) is a very rare genetic disorder characterized by gingival fibromatosis, abnormalities of soft cartilages of the nose and/or ears, hypoplastic or absent nails and terminal phalanges, joint hypermobility, hepatosplenomegaly, mild hirsutism and learning difficulties.[1] Gingival fibromatosis is a characterized by slow and progressive enlargement of maxilla and mandibular gingiva. Gingival fibromatosis, which is sometimes associated with generalized hypertrichosis, mental retardation or epilepsy has been described as a distinct disorder, with autosomal dominant segregation.[2–4] Early presentation of Zimmermann-Laband syndrome in a newborn has rarely been described.[5,6] This paper describes a newborn patient with Zimmermann-Laband syndrome.

Case report

A 1-month-old female was referred to the Department of Dentistry and Oral Surgery, Hiroshima City Hospital, for assessment of gingival over growth. The main complaint was excessive gingival enlargement. She was the second daughter of healthy nonconsanguineous Japanese parents. The pregnancy was uneventful and the infant's birth weight was 3400g, birth length was 50cm, occipitofrontal circumference was 34.3cm, and perithoracic circumference was 34.5cm. Apgar scores at 1 min and 5 min were 1 and 5, respectively. The child had a very striking facial morphology. She had a large fleshy nose, and malformed external ear lobs (Fig. 1). There was massive gingival overgrowth in the

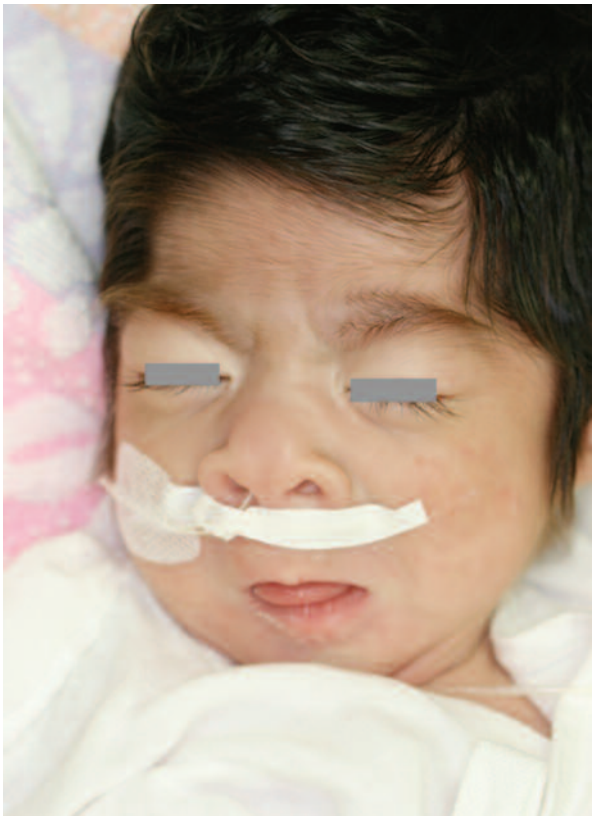

Figure 1. The infant had a very striking facial morphology, a large fleshy nose and malformed external ear lobs.

* Koichi Sawaki, DDS, PhD, sub-director, Department of Dentistry and Oral Surgery, Hiroshima City Hospital.
** Katsuaki Mishima, DDS, PhD, associate professor, Department of Oral and Maxillofacial Surgery, Graduate School of Medicine, Yamaguchi University.
*** Aki Sato, DDS, clinical fellow, Department of Dentistry and Oral Surgery, Hiroshima City Hospital.
**** Yu Goda, DDS, clinical fellow, Department of Dentistry and Oral Surgery, Hiroshima City Hospital.
***** Atsuo Osugi, DDS, clinical fellow, Department of Dentistry and Oral Surgery, Hiroshima City Hospital.
****** Makoto Nakano, DDS, PhD, sub-director, Department of Dentistry and Oral Surgery, Hiroshima City Hospital.

Send all correspondence to: Katsuaki Mishima, Department of Oral and Maxillofacial Surgery, Graduate School of Medicine, Yamaguchi University. Minamikogushi 1-1-1, Ube City, Yamaguchi, 755-8505, Japan.

Phone +81-836-22-2299
Fax +81-836-22-2298

E-mail kmishima@yamaguchi-u.ac.jp

maxillary region (Fig. 2). The enlargement also involved the anterior palate with soft swelling, and the midline of the palate displayed the furrow. In addition, there was a painless polypoid mass of 5mm in diameter with papillary surface in the median of the upper gingiva.

She had no neonatal teeth, no oral synechia, and no cleft palate. The tongue was normal. She had no nails at the toes and hypoplastic nails at digits, and displayed hirsutism in her arms and legs (Fig. 3). There was no hepatosplenomegaly. Complete blood count and blood chemical tests, including thyroid function tests, and amino acids and mucopolysaccharides were within normal ranges. Chromo-

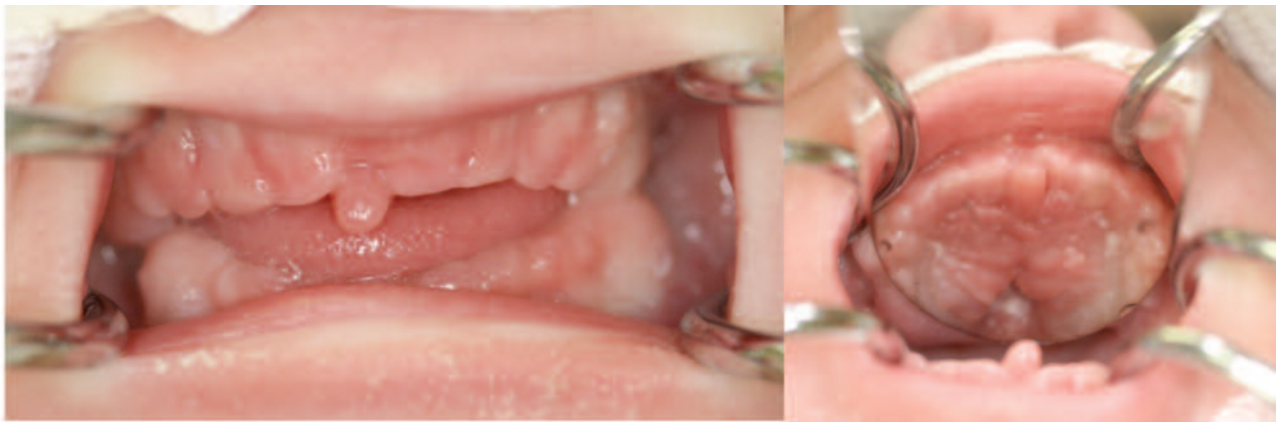

Figure 2. Massive gingival overgrowth in the maxillary region with a polypoid mass of 5mm in diameter and the presence of a papillary surface in the median of the upper gingiva. The enlargement also involved the anterior palate with soft swelling, and the midline of the palate displayed the furrow.

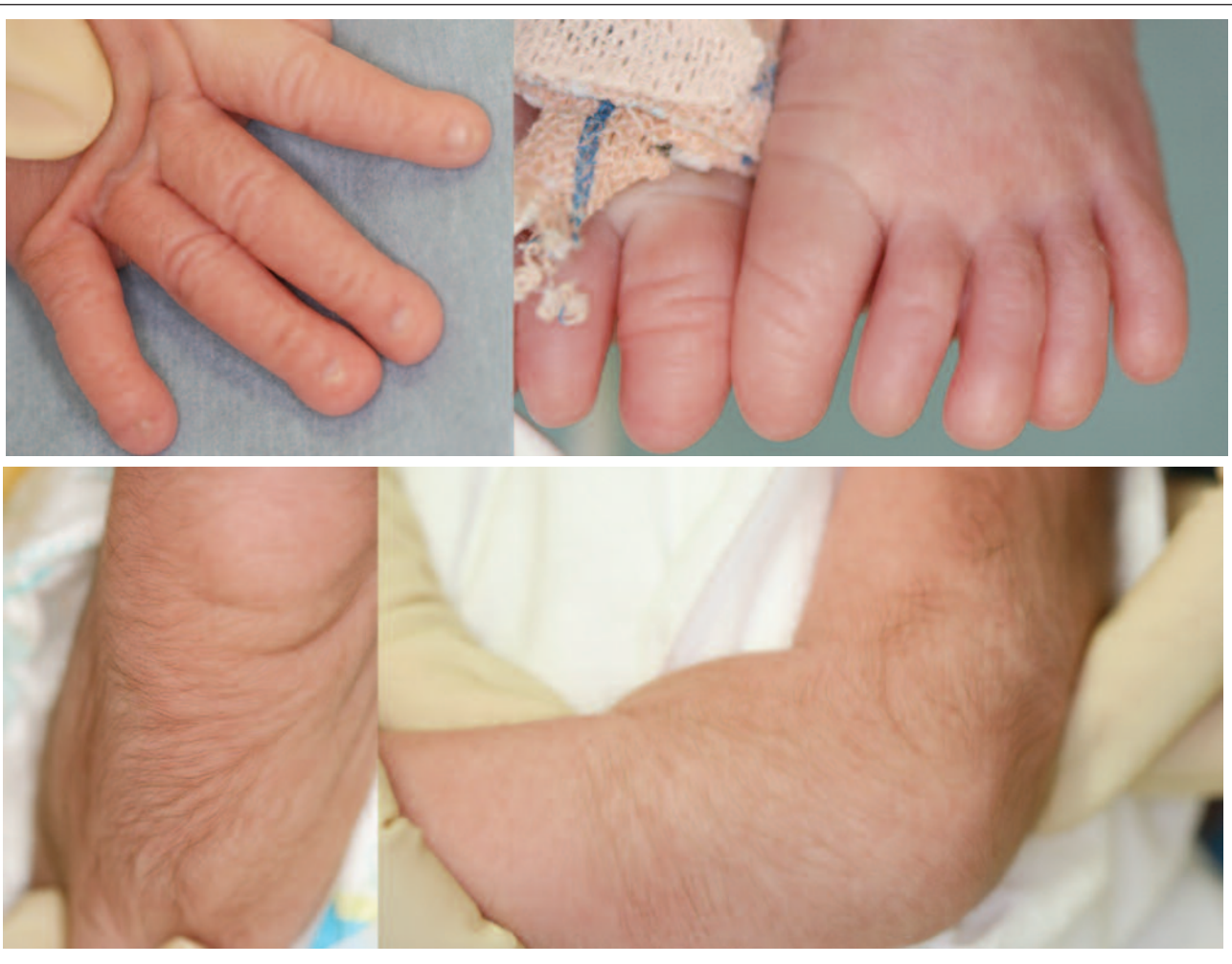

Figure 3. The infant displayed an absence of nails of the toes and hypoplasic nails of the digits, and hirsutism in her arms and legs.

somal examination using G-banding revealed a normal karyotype (46, XX). The patient had tremors from her first days of life. The episodes were initially treated with phenobarbital. The parents of the proposita were normal in appearance. An elder brother was clinically normal. At birth, the mother was 38 years old and the father was 38 years old. The family history was unremarkable. The patient was diagnosed with Zimmermann-Laband syndrome based on her clinical manifestations.

DISCUSSION

Zimmermann-Laband syndrome is a rare genetic disorder characterized by gingival fibromatosis, supernumerary teeth, a bulbous nose and/or ears, thick lips, hypoplastic or absent nails and terminal phalanges, splenomegaly, joint hypermobility, hepatosplenomegaly, hirsutism and mental retardation.[1,7,8]

A young patient with Zimmermann-Laband syndrome has been described in the present report. However, early presentation of Zimmermann-Laband syndrome in a newborn has rarely been described.[5,6] Usually, gingival hyperplasia does not become evident until the eruption of the permanent teeth.[5] Gingival hyperplasia is a key component of the syndrome; in this case it was presented in the newborn period.

To diagnosis generalized gingival enlargement, a detailed medical history and physical systemic evaluation are necessary in order to differentiate gingival fibromatosis from other acquired abnormalities or from similar effects caused by a more extensive syndrome. Namely, the gingival enlargement that occurs as a result of inflammation, pregnancy and leukemia, and as a response to certain drugs such as phenytoin, diltiazem, cyclosporine A, verapamil and nifedipine, should be ruled out. In addition, other syndromes associated with hereditary gingival fibromatosis, that is, Murray syndrome with multiple hyaline dermal tumors, Rutherford syndrome with corneal dystrophy, Cowden syndrome with hypertrichosis, oligophrenia and giant fibroadenomatosis of the breasts and Cross syndrome with hypopigmentation, oligophrenia and asthetosis, should be differentiated.[6,9,10]

Shah *et al* have listed both the major and the occasional findings of this syndrome.[11] Out of the occasional findings, tremor and generalized and facial hypertrichosis were present in our case (Table 1). According to their review, a definite diagnosis of Zimmermann-Laband syndrome can be based on the clinical characteristics of gingival enlargement or fibromatosis, a characteristic facial appearance, and hypoplastic or absent nails.

It is anticipated that the gingival hyperplasia will worsen as the girl grows, and that gingivectomy and gingivoplasty will be necessary. It will be very important to follow this patient closely.

Table 1. Variable clinical findings in patients with Zimmermann-Laband syndrome[11]

Major findings	Gingival fibromatosis or gingival hypertrophy*	
	Dysplasia, hyperplasia or absence of the terminal phalanx and/or nails of hand and feet*	
	Enlargement of soft tissues of the face	Bulbous soft nose*
		Thick lips
		Large ears*
Occasional findings	Hyperextensibility of joints	
	Kyphosis	
	Spina bifida	
	Hallux valgus	
	Clubbed fingers	
	Large tongue	
	High arched palate	
	Partial anodontia	
	Learning disability	
	Tremor*	
	Hypertrichosis of face*	
	Retinitis pigmentosa	
	Hepatosplenomegaly	
	Scoliosis	
	Pes cavus	
	Genu valgum, cubitus valgus	
	Flexion contractures	
	Furrowed tongue	
	Macrocephaly	
	Prognathia	
	Epilepsy	
	Generalized hypertrichosis*	
	Deep palmar and plantar creases	
	Cataract	

*Represents finding in the present case

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REFERENCES

1. Zimmermann. Uber anomalien des ektoderms. Vjschr Zahnheilkd, 44: 419–434, 1928.
2. Anderson J, Cunliffe WJ, Roberts DF, Close H. Hereditary gingival fibromatosis. Br Med J, 3: 218–219, 1969.
3. Horning GM, Fisher JG, Barker BF, Killoy WJ, Lowe JW. Gingival fibromatosis with hypertrichosis. A case report. J Periodontol, 56: 344–347, 1985.
4. Cuestas-Carnero R, Bornancini CA. Hereditary generalized gingival fibromatosis associated with hypertrichosis: report of five cases in one family. J Oral Maxillofac Surg, 46: 415–420, 1988.
5. Dumić M, Crawford C, Ivković I, Cvitanović M, Batinica S. Zimmerman-Laband syndrome: An unusually early presentation in a newborn girl. Croat Med J, 40: 102–103, 1999.
6. Atabek ME, Pirgon O, Sert A, Toy H. Zimmermann-Laband syndrome in an infant with an atypical histologic finding. Pediatr Dev Pathol, 8: 654–657, 2005.
7. Chadwick B, Hunter B, Hunter L, Aldred M, Wilkie A. Laband syndrome. Report of two cases, review of the literature, and identification of additional manifestations. Oral Surg Oral Med Oral Pathol, 78: 57–63, 1994.
8. Lin Z, Wang T, Sun G, Huang X. Report of a case Zimmermann-Laband syndrome with new manifestations. Int J Oral Maxillofac Surg, 39: 937–941, 2010.

9. Witkop CJ Jr. Heterogeneity in gingival fibromatosis. Birth Defects Orig Artic Ser, 7: 210–221, 1971.
10. Gorlin RJ, Cohen MM, Hennekam RCM, editors. Syndromes of the Head and Neck, 4th ed. Oxford: Oxford University Press, 1093–1106, 2001.
11. Shah N, Gupta YK, Ghose S. Zimmermann-Laband syndrome with bilateral developmental cataract – a new association? Int J Paediatr Dent, 14: 78–85, 2004.