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Case Report

Dental findings and management in a child with hypomelanosis of Ito

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ABSTRACT

Hypomelanosis of Ito (HI) is a rare neuroectodermal disorder characterized by hypopigmented whorls of skin along the Blaschko lines. Mental retardation and intractable epilepsy are also commonly observed in HI. However, there are only a few published reports describing dental observations in HI patients. Here, we report the oral management and novel dental findings of a male HI patient aged 3 years and 10 months. The chief complaint was spontaneous gingival bleeding due to gingival hyperplasia induced by anticonvulsants, which was improved with plaque control and gingival massage. Numerous additional dental abnormalities were also evident, including enamel hypoplasia and large pulpal chambers.

1. Introduction

Minoru Ito, a Japanese dermatologist, reported an unusual case of incontinentia pigmenti (IP) in 1952 ultimately recognized as the first described case of incontinentia pigmenti achromians (IPA) [1]. The features and inheritable traits of IPA differ from those of IP; thus, the disorder was classified as hypomelanosis of Ito (HI) in 1973 [2]. The incidence of HI is reportedly very rare; it was estimated to affect only 1 in 1000 new patients in a pediatric neurology service and 1 in 8000–10,000 patients in a children's hospital [3].

HI is characterized by unilateral and bilateral macular hypopigmented holes, streaks, and patches, which is the opposite pattern evident in hyperpigmented areas of IP [1,2,4]. Importantly, HI is significantly associated with afflictions of the central nervous system including mental retardation and intractable epilepsy [2,5]. Further, HI frequently involves ophthalmic abnormalities including strabismus, nystagmus, and congenital cataracts [2].

Abbreviations: CRP, C-reactive protein; HI, hypomelanosis of Ito; IP, incontinentia pigmenti; IPA, incontinentia pigmenti achromians.

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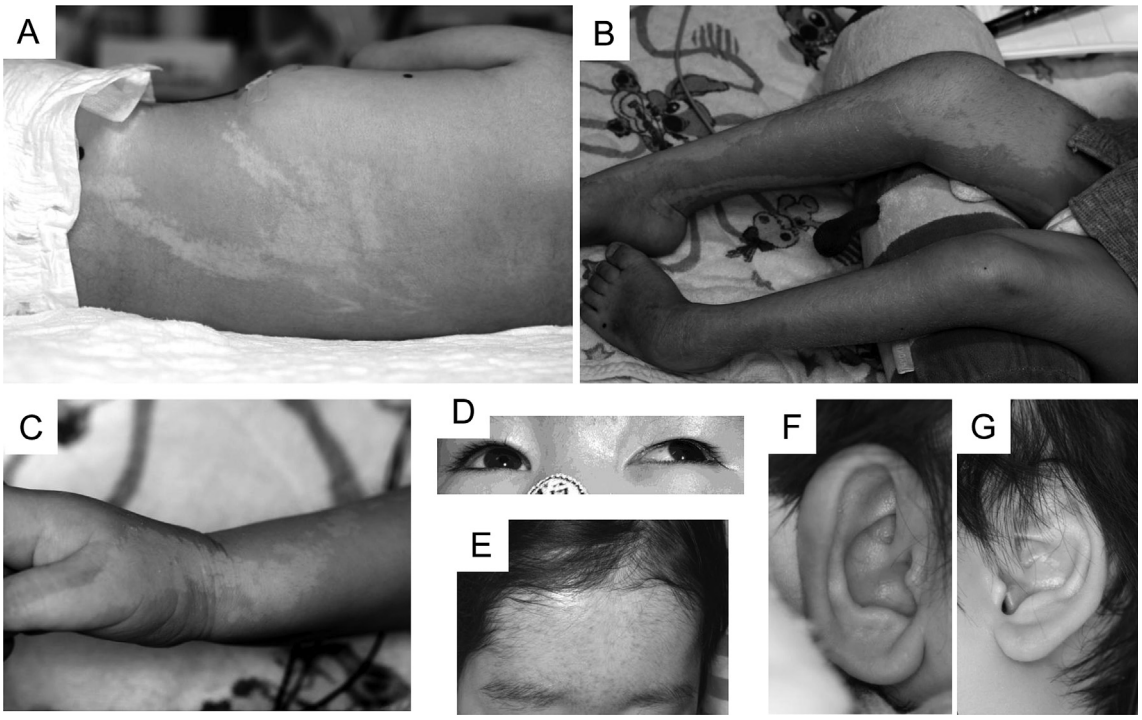

Fig. 1 – Linear and whorled hypopigmented patches along the lines of Blaschko were observed on the trunk (A), the right leg (B), and the right arm (C). Heterochromia iridis was observed on the left eye (D). Hirsutism was apparent on the forehead (D). Auricle asymmetry was evident on the right (F) and the left (G).

A number of dental abnormalities including hypodontia, conical tooth, delayed eruption, and malformed teeth were reported in a study investigating IP [6]. However, there are only a few reports of dental findings in HI. The purpose of this report was to document the dental observations and oral management of a case of the rare disorder, HI.

2. Case report

A Japanese male boy aged 3 years and 10 months with HI was referred to our clinic for examination of gingival swelling and bleeding. His birth had involved vaginal breech at 35 weeks and 2 days gestation, and occurred in the local maternity hospital. He was the second boy of a 32-year-old woman. At birth, his weight was 2276 g (−0.4 standard deviation (SD)) and head circumference was 35 cm (+1.8 SD). He had suffered from severe neonatal asphyxia, and Apgar scores after 1 and 5 min were 2 and 6, respectively. He was immediately admitted to the neonatal intensive care unit of our hospital. At that time, he had a healthy 5-year-old brother, and his family history was otherwise unremarkable.

At birth, systematized linear and patchy depigmentations, distributed in a pattern following the lines of Blaschko, were observed on the trunk and the right extremities (Fig. 1A–C). From the age of 12 days, he began to have intractable epileptic seizures. Magnetic resonance imaging demonstrated lissencephaly. Further, he was diagnosed with heterochromia iridis at the age of 3 months (Fig. 1D), and optic nerve hypoplasia at 7 months. Hirsutism (Fig. 1E) and auricle asymmetry (Fig. 1F and G) were also observed. Based on these findings, HI was diagnosed.

Artificial respiration management via tracheostomy had been initiated in this patient at the age of 10 months, due to

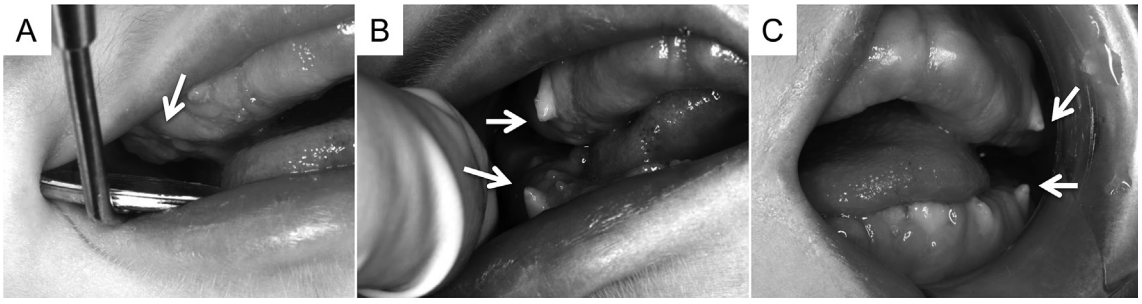

Fig. 2 – The gingiva of the upper right second primary molar bleed easily upon brushing (A). Enamel hypoplasia was observed at the tip of the cusps of the primary canines on the right (B) and left (C).