

Labial talon cusp in a child with incontinentia pigmenti achromians: case report

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Introduction

Incontinentia pigmenti achromians (IPA), first reported by Ito (Ito 1952), is a rare disease, involving the skin, hair, eyes, central nervous system, and musculo-skeletal system. The cutaneous manifestations consist of macular hypopigmented whorls, streaks, and patches in a bilateral or unilateral distribution affecting almost any portion of the body surface. Central nervous system dysfunction, ocular and musculoskeletal anomalies also have occurred with significant frequency (Schwartz et al. 1977; Takematsu et al. 1983).

IPA superficially resembles incontinentia pigmenti (IP), but IPA has been regarded as distinct and separate from IP (Jelinek et al. 1973; Takematsu et al. 1983). IP is seen almost exclusively in females and has been considered an X-linked inherited disease (Gorlin et al. 1976; Jelinek et al. 1973; Schwartz et al. 1977; Takematsu et al. 1983). On the other hand, the mode of inheritance of IPA remains unclear (Schwartz et al. 1977; Takematsu et al. 1983).

IP is associated frequently with delayed tooth eruption, pegged or conical crowned teeth, missing teeth, and malformed teeth (Gorlin et al. 1964). IPA also may have associated dental anomalies, but only a few reports have been published. Browne and Byrne (1976) reported on an unusual form of dental dysplasia and the microscopic structure of the teeth in IPA. Happle and Vakilzadeh (1982) reported multiple dental cusps in both primary and permanent incisors. Bartholomew et al. (1987) reported congenital absence of a maxillary primary central incisor.

This paper describes a patient with IPA having an abnormally shaped maxillary permanent central incisor.

Case Report

The patient, a 6-year-old, well-developed, well-nourished Japanese female, was the middle of three sisters. The mother had no miscarriages and siblings were reported to be free of systemic illness or abnormalities. Family history was negative for IPA.

The patient's in utero course and delivery were uneventful. At birth, areas of pigmentation and depigmentation were present on the child's trunk and extremities without inflammation. These progressed gradually until 2 months of age. The patient was referred to the Department of Dermatology, Hokkaido University, School of Medicine at 4 years and 5 months of age, and was diagnosed with IPA.

At the age of 6 years and 6 months, the patient was referred to the Department of Pediatric Dentistry, Hokkaido University, School of Dentistry, with the chief complaint of an abnormally shaped maxillary permanent central incisor. Oral examination revealed that her dental age was Hellman's IIC (Hellman 1932). The mandibular permanent central incisors had erupted. The mandibular permanent lateral incisors, first molars, and the maxillary permanent central incisors were erupted partially. The mother reported a normal primary dentition.

The maxillary left permanent central incisor exhibited a well-defined labial cusp located perpendicular to the mesiodistal plane of the tooth that extended from the cervical to the incisal edge of the crown. A minor cusp was also present on the cingulum area (Fig 1). The cusp present on the labial surface resembled the talon cusp which has been reported to be present on the cingulum area of anterior teeth (Mader 1981). Developmental grooves were observed between the cusp and the tooth. The radiographic examination revealed that the cusp was composed of normal enamel and dentin, and contained a pulp horn (Fig 2, see next page). Comparison of the structure, shape, and size of the two maxillary permanent incisors showed no other clini-



Fig 1. Labial cusp of the maxillary left permanent central incisor. The cusp extended from the cervical portion to the incisal edge (front view).



Fig 2. Apical radiograph showing the labial cusp with normal enamel and dentin. Note the horn of the pulp extending into the cusp.

cally significant differences. Defects of the other teeth were not observed. The gingiva, tongue, and oral mucosa were normal in color, contour and texture. The patient had no irritation of the upper lip.

Discussion

Dental anomalies may occur in IPA, but only a few reports have been published (Browne and Byrne 1976;

Happle and Vakilzadeh 1982; Bartholomew et al. 1987). In the present case, the maxillary left permanent central incisor exhibited a well-defined labial cusp. No hereditary origin of the tooth defect was identified. The cusp resembled a talon cusp which is defined as a cusp-like structure projecting from the cingulum area of a maxillary or mandibular incisor (Mellor and Ripa 1970; Mader 1981). Whether the cusp anomaly in this case is associated with IPA could not be resolved. Since this anomaly is rare in the normal population (Nishijima et al. 1959; Schulze 1970), it is possible that it is a finding of IPA. If

so, this cusp anomaly may help establish a diagnosis when major clinical findings are minimal or atypical. More reports are needed to establish the oral features of IPA.

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