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15 March 2005

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Journal of Craniofacial Genetics
and Developmental Biology 7:311–317 (1987)

Natal Teeth and Steatocystoma Multiplex: A Newly Recognized Syndrome

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A Chinese family is reported in which five generations have exhibited natal teeth and generalized multiple steatocystomas. This autosomal dominant condition is not similar to the two reported types of pachyonychia congenita, because nail lesions, palmoplantar keratosis and hyperhidrosis, follicular keratosis, and oral leukokeratosis were not observed. Therefore, it is suggested that this family exhibits a newly recognized syndrome.

Key words: mandibular incisors at birth, cystic skin lesions

INTRODUCTION

Natal and neonatal teeth occur with a frequency of approximately 1 in 3,000 births [Bodenhoff and Gorlin, 1963]. The condition probably is due to an abnormally superficial position of the tooth germs resulting in premature eruption [Boyd and Miles, 1951; Hals, 1957]; a strong familial trend has been observed in some cases [Massler and Savara, 1950], with an autosomal dominant inheritance pattern [Bodenhoff and Gorlin, 1963]. Natal teeth are associated with other anomalies in chondroectodermal dysplasia (Ellis-van Creveld syndrome), oculomandibulodyscephaly (Hallermann-Streiff syndrome), and pachyonychia congenita [Bodenhoff and Gorlin, 1963]. In 1976, McDonald and Reed reported a family with a syndrome of multiple steatocystomas and natal teeth; although the condition resembled pachyonychia congenita, there were no nail abnormalities. A Chinese family with a syndrome closely resembling that reported by McDonald and Reed is presented.

CASE REPORTS

Case 1

A 3-day-old Chinese female (Fig. 1, V-5) was referred for treatment of two mandibular central incisors that were erupted at birth. No other anomalies were detected. Both teeth appeared to be covered by immature enamel, and they were extremely mobile. Because of the poor prognosis, they were extracted under local anesthesia on the third day of life. The extraction sites subsequently healed. However,

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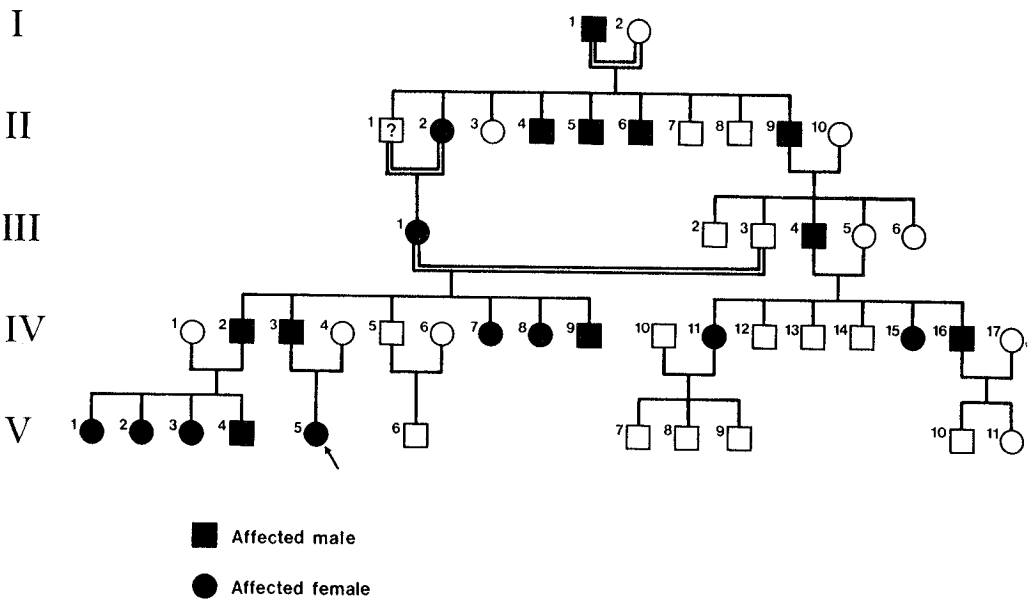

Fig. 1. Pedigree of family with natal teeth and steatocystoma multiplex.

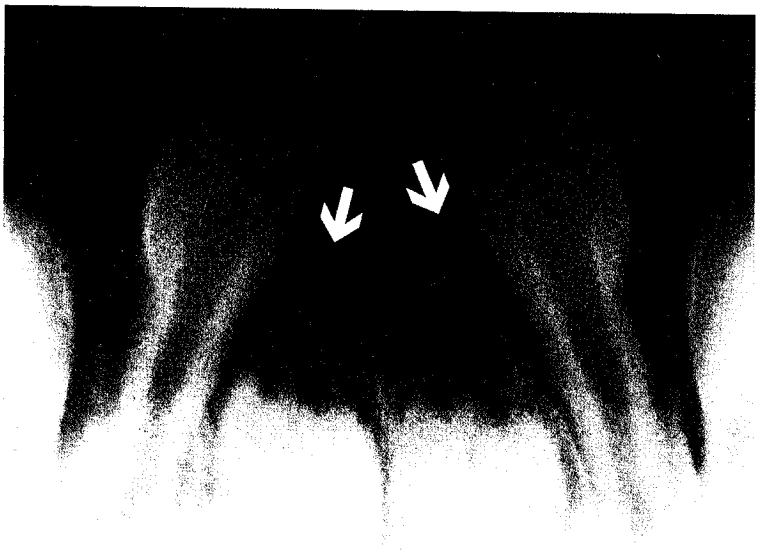

Fig. 2. Radiograph taken at 13 months showing two root like fragments (arrows) in the mandibular central incisor region of the propositus (V.5).

a small abscess developed a year later, which was surgically drained. Subsequent radiographs of the region revealed two rootlike structures at the sites of the previously extracted natal teeth (Fig. 2). These structures may have been part of the remaining portion of the tooth germs, which, after extraction of the erupted part of the natal teeth, had continued to develop and eventually became infected. The permanent successor teeth appeared from the radiograph to be developing normally.

At the age of 16 months, pinhead-sized milia-like whitish papules were found on the skin of the back and around the base of the nose (Fig. 3). The nails of the hands and feet developed normally (Fig. 4), and no other abnormalities were found.

Case 2

The father (Fig. 1, IV-3) of the propositus had two natal teeth in the lower jaw, which were extracted. He gradually developed numerous cystic skins lesions, with a generalized distribution. Although these lesions had been biopsied, no reports were available. No visible abnormalities were observed on the scalp or the oral mucosa, and all nails were normal (Fig. 5a, b).

Case 3

The aunt of the propositus (Fig. 1, IV-7) had natal teeth, which were extracted by the grandmother. Her arms and neck were found to have numerous cystic lesions (Fig. 6). Biopsy of these lesions had been performed 2 years prior to examination. The lesions were well encapsulated with a yellow, turbid fluid. Histological exami-

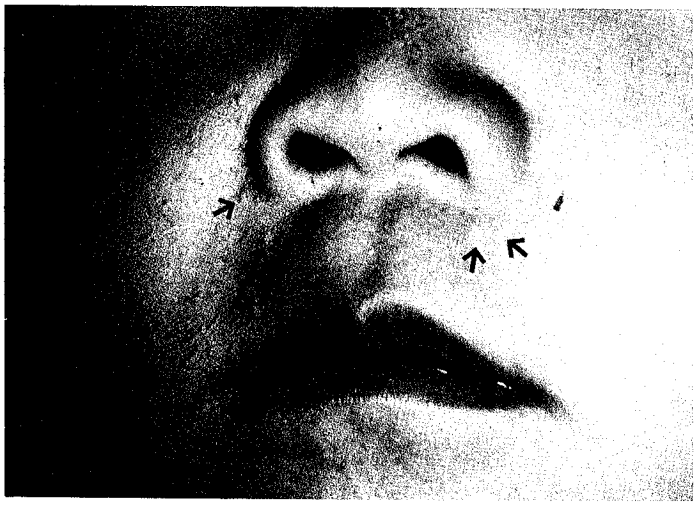

Fig. 3. Pinhead-sized, milia-like, whitish papular skin lesions on the face of propositus (arrows).

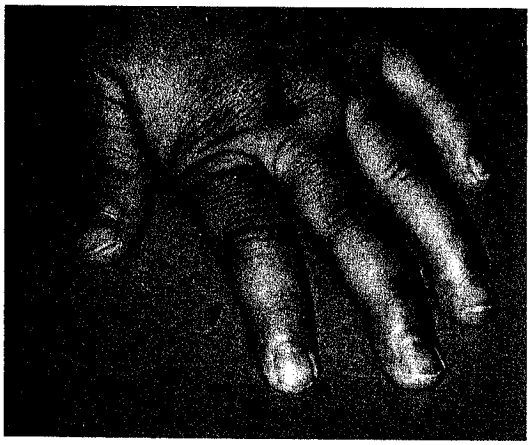

Fig. 4. The hand of the propositus (V.5), showing normal nail development.

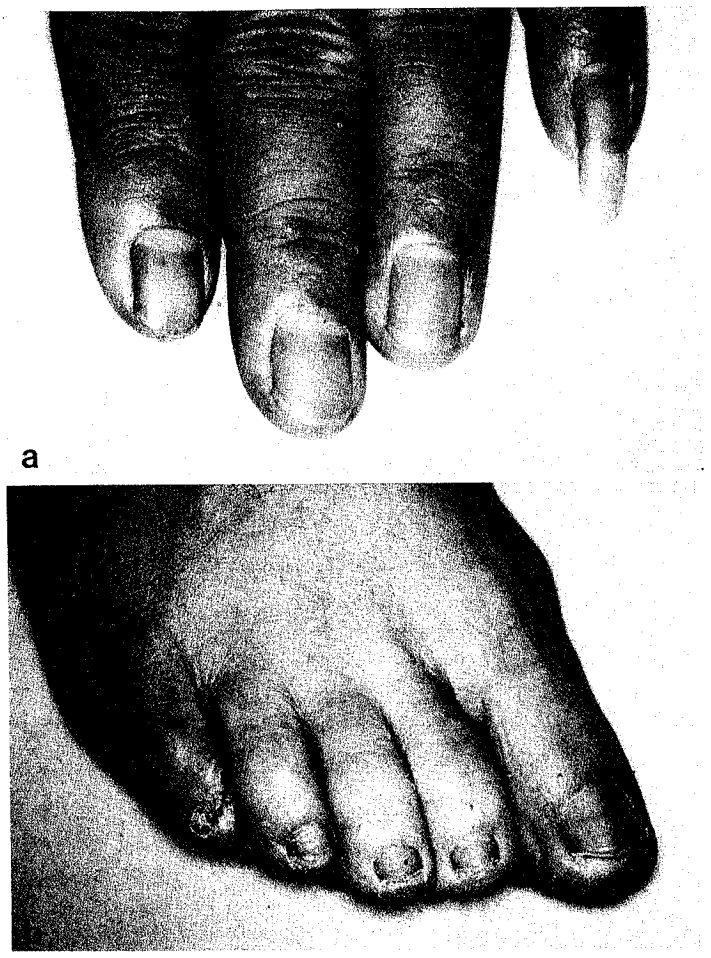

Fig. 5. Normal development of the nails of the left hand **(a)** and the right foot **(b)** of the father (IV.3).

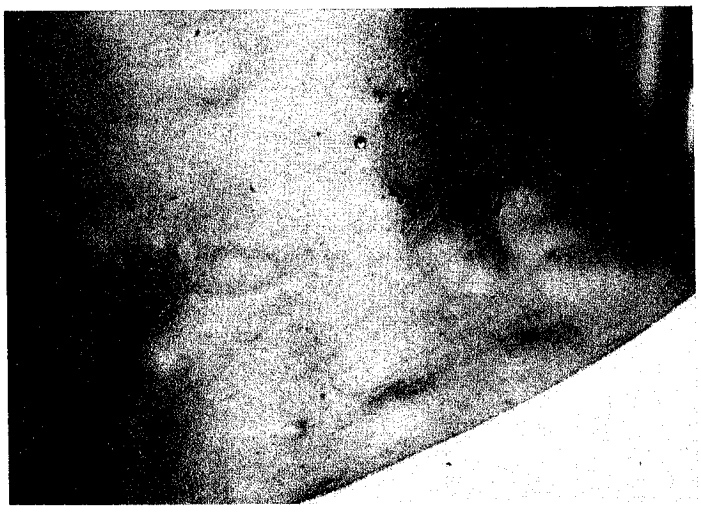

Fig. 6. The propositus' aunt (IV.7), exhibiting multiple steatocystomas on the neck.

nation showed lobules of sebaceous glands, which drained into ductlike structures lined by stratified squamous epithelium (Fig. 7). These findings were consistent with steatocystoma multiplex. She refused to have further biopsies. Her nails were normal (Fig. 8).

DISCUSSION

Pachyonychia congenita is probably genetically heterogenous, two syndromes being subsumed under the same term. The first, type I (Jadassohn-Lewandowski syndrome), involves congenital hypertrophy of the nails, palmoplantar keratosis and hyperhidrosis, follicular keratosis, and oral leukokeratosis [Akesson, 1967; Gorlin et al, 1976]. Type II (Jackson-Lawler syndrome) has the same features as type I, with the exception of oral leukokeratosis. In addition, large cutaneous epidermoid cysts and natal teeth have been observed [Murray, 1921; Vineyard and Scott, 1961; Moynahan and Shrank, 1966; Soderquist and Reed, 1968; Boxley and Wilkinson, 1971]. The disorder is believed to be inherited as an autosomal dominant trait [Gorlin et al, 1976]. Although the two syndromes exhibit many similarities, both have been seen in the same family only once. This was a Swedish family who were reported to have natal teeth associated with type I pachyonychia congenita [Anneroth et al, 1975].

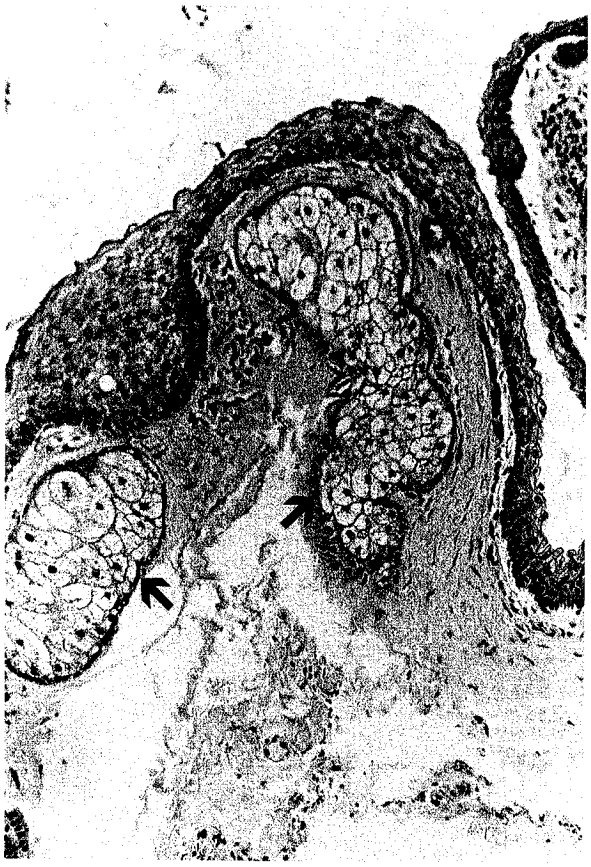

Fig. 7. Histologic appearance of the skin lesion of the propositus' aunt (IV. 7), showing multiple sebaceous glands (arrows) within the cyst wall ×16.

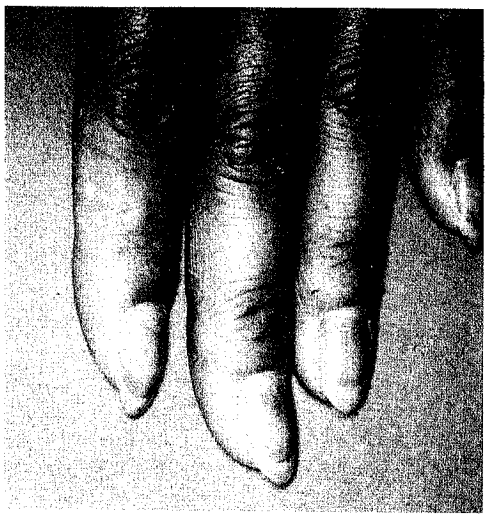

Fig. 8. The propositus' aunt (IV.7), showing normal nail development.

Therefore, doubt remains regarding whether these syndromes should be separated into types I and II.

Not all the affected members of the present family have been examined because of their location in Burma. However, the father (Fig. 1, IV-3) was able to indicate which relatives had been affected by natal teeth and skin lesions and to confirm the absence of nail involvement. Since the condition exhibited by the propositus has persisted within the family for five generations, the detailed knowledge of the father about the appearance of the condition, his extensive personal investigation, and the old family records lead the authors to trust his reliability.

From the data collected, more than 50% of the offspring of the reported family had natal teeth and skin lesions, with both males and females in every generation being equally affected. The present family and the one reported by McDonald and Reed [1976] exhibited natal teeth and steatocystomas as the only traits. Natal teeth and steatocystoma multiplex are reported to be hereditary traits singly [Noojin and Reynolds, 1948; Hals, 1957]; they have been reported together in only one individual once before [McDonald and Reed, 1976]. Nail abnormalities are a consistent component in the pachyonychia congenita syndrome. Since this was not present in this family nor in that reported by McDonald and Reed [1976], it is reasonable to conclude that these two families have a distinct syndrome that should be differentiated from pachyonychia congenita.

ACKNOWLEDGMENTS

The authors wish to thank Dr. E. Gwi and staff at Kwong Wah Hospital for their assistance with the histologic aspects of this report.

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