

ATRICHIA WITH CYSTS AND PAPULES

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A robust, intelligent male aged 32 years had atrichia with cystic lesions. Scanty hairs were present at birth on the scalp and were shed in the course of a few weeks. Multiple papular and cystic swellings started appearing 10 years back and were seen over the shoulders, axillae and the elbows. Nails, although normal, were racquet shaped while the teeth were normal. The mode of inheritance of this rare condition has not yet been established.

Key words : Atrichia, Cysts, Papules.

Damste and Prakken¹ reported in 1954, three women patients with atrichia and papular lesions. In 1961 Lowenthal and Prakken² described another girl with similar lesions. One of Damste and Prakken's cases had a family history and the case of Lowenthal and Prakken was born out of consanguinity. Ten such cases have so far been reported. Although an autosomal recessive inheritance was postulated, the exact mode of inheritance is not yet settled. We report a case of atrichia with papular and cystic lesions in an adult male with no family history.

Case Report

A 32-year-old male patient had absence of hair since birth and multiple painless swellings over the body for the last ten years. The patient did not remember to have had hairs on the scalp but was told that at birth there were scanty hair which were shed in the course of a few weeks. Born of non-consanguinous parents, his only sister had abundant hair growth; none of his parents or close relatives had similar problem.

The patient noticed the appearance of multiple painless swellings over the right knee to start with 10 years back, and later over the back of the elbows, nape of the neck, axillae

and the back. One or two swellings used to become painful periodically and heal either by spontaneous rupture or after incision and drainage by the local practitioners.

There was total absence of hairs all over the body including the secondary sexual hairs, except a few fine eyelash hairs present over the upper eyelids (Fig. 1). Multiple skin-coloured cystic swellings varying in size from 1 to 3 cm in diameter, and papules were seen on the back of the elbows, axillae, shoulder and the back punctuated by pitted atrophic scars (Fig. 2). Sweating was normal; so also the teeth and the nails except that the latter were racquet-shaped. Physical examination otherwise was unremarkable. Routine laboratory inves-



Fig. 1. Complete loss of hairs on the scalp, eyebrows and eyelashes.

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Fig. 2. Numerous papules and cysts on the shoulder and the back.

tigations did not reveal any abnormality. Biopsy of one of the swellings showed keratin-filled cyst in the mid-dermis.

Comments

In one of the three unrelated women patients described by Damste and Prakken¹ atrichia associated with cystic lesions was familial while the girl reported by Lowenthal and Prakken² had consanguinous parents. Three others reported from Germany³ were brothers while two patients from Mexico⁴ were sisters. The boy and his father reported by Kanzler and Rasmussen³ had normal hair until the age of 2 years. The patient reported by us was the only one in the family to have atrichia. It is evident that the mode of inheritance of atrichia with papules is, thus far, not certain.

The associated cystic lesions have been unique in that these were consistently present in all the patients so far reported although other features like mental retardation, polyposis of the gastro-intestinal tract, immunodeficiency were, in all probability, only coincidental. The

age of onset of these cystic papular lesions in our patient was around 22 years; it has varied from the age of 2 to 26 years in the other cases. Histopathology of the lesions showed keratin-filled cysts in the dermis independent of the epidermis and the hair follicle. The patient showed no other evidence of ectodermal or craniofrontal dysplasia.

The presence of hair over the scalp at birth rules out the possibility of total agenesis of hair follicles as reported in some patients with ectodermal dysplasia. The fact that he had hair on the scalp at birth speaks for the functional viability of the hair follicles to form hairs in early life. As postulated by Damste and Prakken¹ these follicles might have been damaged later on to a point when continued hair growth became almost impossible. What causes the damage is, of course, a moot point. Similarly, the suggestions of a hereditary factor acting during the intrauterine life or an infection or intoxication of the mother are only speculative.

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