A case of aquagenic syringeal acrokeratoderma with involvement of periungual area of the hand

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Aquagenic syringeal acrokeratoderma (ASA) is a rare form of palmoplantar keratoderma occurring after short-term contact with water. Although ASA usually involves the palmar region, there are also several cases with the involvement of dorsum of hand and sole of the feet. We described 15-year old girl who had white keratodermic plaques observed on the flexor side of distal phalanxes especially the periungual area after a 10 min contact with water. Our patient represents a rare case of ASA with the involvement of periungual region of the fingers.

Key words: aquagenic syringeal acrokeratoderma, adolescent, water.

Aquagenic syringeal acrokeratoderma (ASA) is a rare form of palmoplantar keratoderma occurring after short-term contact with water.¹ It was first defined in 1996 by English and McCollough^{1,2} in the name of 'transient reactive papulotranslucent acrokeratoderma' and then called 'aquagenic palmoplantar keratoderma' and 'aquagenic syringeal acrokeratoderma' by other authors.

Case Report

A 15-year old female patient was admitted to clinic with the complaint of blistering for about 1 year after water exposure which began on the palm of the hand and spread around the fingers. The patient reported that her complaints increase with water contact for 10 min and alleviate with a 20 min resting period after drying the hands. The patient also stated that she had excessive sweating on her hands. After a 10 min contact with water, white keratodermic plaques were observed on the flexor side of the distal phalanxes and especially periungual area (Fig. 1-2). Lesions completely disappeared after about 20 min following drying the hands. She had palmar hyperhidrosis. Neither the patient nor her family had history of cystic fibrosis. Punch

biopsy was performed from the periungual area of the left third finger of the patient.

Histopathological examination of the lesions showed orthokeratotic hyperkeratosis on the surface with the dilatation of acrosyringium and a tortuous appearance. Slightly evident irregular acanthosis was found in the epidermis with slight perivascular lymphocytic infiltration in the superficial dermis (Fig. 3 A-D).

In light of the above-mentioned clinical and histopathological findings, a diagnosis of ASA was made and topical 20% aluminum chloride hexahydrate was prescribed. Partial response was observed after 1-month of treatment. Written informed consent was obtained from the parents of the patient.

Discussion

ASA is characterized by localized, transparent and white-colored papules and plaques commonly in the palmar region of hand.¹⁻³ Although it is commonly asymptomatic, patients may complain from burning, pain, itching, and tingling. Hyperhidrosis may accompany the active disease.⁴

Although ASA usually involves the palmar

Fig. 1. Aquagenic syringeal acrokeratoderma. White keratodermic plaques on periungual area

region, there are also several cases with the involvement of dorsum of the hand and sole of the feet. A comprehensive literature search revealed only 5 ASA cases with the involvement of dorsal aspect of hand.⁵⁻⁹ Thus, our patient represents a rare case of ASA with the involvement of periungual region of the fingers.

Although the pathogenesis is not clear, there are several theories suggesting that the stratum corneum and eccrine ductus and glands play a role in this disease.³ It has been suggested that abnormalities of the sweat glands, hyperkeratosis and impairment in the barrier function of the stratum corneum increase the water absorption and lead to the swelling of stratum corneum.

Fig. 2. Aquagenic syringeal acrokeratoderma. White keratodermic plaques on flexor side of distal phalanges.

This condition has a genetic basis with both autosomal dominant and recessive inheritance.¹⁰ Familial cases have also been reported in the literature.¹¹ ASA is seen in 40-80% of the

Fig. 3. (A-D) Microscopic photos of the lesion illustrating the intraepidermal eccrine ductus (acrosyringium) dilatation and tortuosity (arrows) (Hematoxylin-eosin stain; original magnification $\times 40$, $\times 100$, $\times 200$, and $\times 400$, respectively).

patients with cystic fibrosis, while the rate is approximately 25% in the case of being a carrier.¹² In cystic fibrosis patients, defective chlorine channels create an osmotic gradient and hypertonic sweat secretion results in the dilatation of eccrine channels.¹³ Further investigation was not performed because there was no finding compatible with cystic fibrosis in our patient's evaluation.

In the literature, there are a number of cases secondary to the use of selective cyclooxygenase-2 (COX-2) inhibitors and aspirin.¹⁴⁻¹⁶ COX-2 inhibitors increase the sodium retention in epidermal keratinocytes resulting in an increase in the water retention capacity of stratum corneum. Belli et al.¹⁷ presented a case of ASA secondary to ibuprofen use and have suggested that COX inhibition may play a key role in the pathogenesis of ASA and that common use of NSAIDs may increase frequency of the disease. Our patient had never received COX-2 inhibitors.

Yan et al.² have defined the lesions becoming evident by immersing hands of 3 patients in water as "hand in the bucket" sign which has been suggested to be an important finding for the diagnosis of ASA. Clinical and histopathological findings are often sufficient for diagnosis. Sezer et al.¹⁸ reported that sweat gland openings in the dermatoscopic examination of ASA patients were three times wider than in normal skin areas and they recommended that dermatoscopy be used as a rapid and non-invasive diagnostic method prior to histopathological examination. Dermatoscopy would help to avoid unnecessary biopsies in doubtful cases.

Treatment options include aluminum salts, salicylic acid creams, antihistamines, and botulinum toxin injections.⁴ In our patient, topical 20% aluminum chloride and hexahydrate produced a partial response to treatment.

In conclusion, while ASA is rare, our case presents an extremely rare condition associated with the involvement of periungual region.

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