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.
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.\textsuperscript{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.\textsuperscript{5} 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.

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

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

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

**Fig. 3.** (A-D) Microscopic photos of the lesion illustrating the intraepidermal eccrine ductus (acrosyringium) dilatation and tortuosity (arrows) (Hematoxylin-eosin stain; original magnification ×40, x100, x200, and x400, respectively).
patients with cystic fibrosis, while the rate is approximately 25% in the case of being a carrier. In cystic fibrosis patients, defective chlorine channels create an osmotic gradient and hypertonic sweat secretion results in the dilatation of eccrine channels. 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. 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. 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. 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. 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. 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.

REFERENCES