SKIN

BRIEF ARTICLE

Aquagenic Wrinkling and Pain of The Fingertips: A Case Report and Review of Aquagenic Palmoplantar Keratoderma

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ABSTRACT

Aquagenic palmoplantar keratoderma (APPK) classically presents as exaggerated wrinkling of the palms after brief immersion in water that lasts 10 minutes to a few hours. We present a case report of a 19-year-old female who presented peeling, sensitivity, and pain on her palmar hands, most prominently on her fingertips, that persists for several minutes to an hour after touching water. This case highlights a unique presentation of APPK predominantly involving the patient's palmar fingertips, a distribution not described in the literature. Current therapies are unsatisfactory and better therapies are needed to improve the quality of life for patients with APPK.

INTRODUCTION

Aquagenic palmoplantar keratoderma (APPK) is a rare disorder often seen in patients with cystic fibrosis or carriers of the cystic fibrosis gene. Clinically, it presents as small, white, or translucent papules coalescing into plaques, sometimes associated with a painful or burning sensation. The cause is uncertain but thought to be related to sodium chloride imbalance in the epidermis leading to increased water retention.[1] We discuss a case of APPK that presented as aquagenic wrinkling and pain of the fingertips.

CASE REPORT

A 19-year-old female presented to the dermatology clinic with hand sensitivity since childhood that began worsening around the age of 12. She complained that whenever her hands touch water of any temperature, within minutes, she experiences peeling, sensitivity, and pain on her palmar hands. These symptoms occur most prominently on her fingertips and sometimes her lateral palm and thenar eminence, which persists for several minutes to an hour. She frequently moisturizes her hands throughout the day with little improvement. It does not involve her feet. Additionally, she noted excess palmar sweating. She reported a history of eczema as a child but no allergies or asthma. She takes no medications. Her sisters have the same symptoms to a lesser degree. She has no family history of cystic fibrosis.

On exam, the patient had normal-appearing palms with Fitzpatrick type II skin (**Figures 1A and 1B**). After one minute of exposure to running tap water, she developed cobblestone wrinkling of her palmar fingertips with shiny edematous papules and plaques,

which improved by the time she left the office (**Figure 2**). This response suggested the diagnosis of APPK.

Genetic testing was recommended to determine if she was a carrier of the cystic fibrosis gene. She was counseled that she may improve with age and informed of treatment options: 20% aluminum chloride followed by urea cream, dimethicone, iontophoresis, and botulinum toxin injections. She had no improvement with 20% aluminum chloride followed by urea cream or botulinum toxin injections (fifty units in each palm). Currently, the patient is managing her symptoms through frequent moisturization and minimizing palmar contact with water.

DISCUSSION

APPK is a rare condition that classically presents as exaggerated wrinkling of the palms, and sometimes the soles, after brief immersion in water that lasts 10 minutes to a few hours.[2] The wrinkling often resolves within minutes to hours of drying the hands. Concurrent pain, burning, itching, and tingling may often be present. It favors adolescents and females and can be sporadic or inherited. Clinically, the patient's palms can present with small white or translucent edematous papules coalescing into plaques with prominent eccrine ducts. Dermoscopy shows papules at sites of dilated acrosyringeal ostia.[2] APPK most often involves the palms but less often involves the soles. Notably, our patient's involvement is predominantly on the palmar fingertips, a distribution not specifically described in the literature but has been subtly apparent on close inspection of published photographs.[3]

Although the etiology of APPK is not well understood, several mechanisms have been suggested. One theory is that abnormally functioning sweat glands cause an increased sodium chloride concentration in the epidermal cells resulting in an increased capability of the stratum corneum to bind water.[1] APPK is associated with cystic fibrosis and presents with the same mutations found in CF, most often ΔF508 of the CFTR gene.[2] The literature has described that 40-84% of patients with CF have concomitant APPK.[1] Improper expression of aquaporins 3 and 5 have also been theorized to impact the sweating mechanisms.[4] Other explanations involve a defective stratum corneum barrier and autonomic nervous system dysfunction.[5] It can also be idiopathic or drug-induced. Furthermore, APPK can be associated with marasmus, nephrotic syndrome, cardiac abnormalities, and palmoplantar hyperhidrosis.[2]

The diagnosis of APPK is usually made based on the clinical history and physical exam. The palmar eruption can be reproduced by the "hands in water bucket" test, which consists of immersing the hands in 15˚C water for five minutes.[2] Although a biopsy can be performed to confirm the diagnosis, our patient did not require one. Since the diagnosis is usually clinical, there are not many histopathological observations. Still, histology will reveal normal skin or dilated eccrine ostia and a mildly hyperkeratotic stratum corneum.[2]

This condition should be distinguished from hereditary papulotranslucent acrokeratoderma, which usually presents as asymptomatic persistent, yellowish-white, and translucent papules in girls during puberty.[6] It is an autosomal dominant condition in which the lesions are persistent and not induced by water. Another consideration is symmetrical acrokeratoderma, which presents as symmetrical hyperkeratotic brownish plaques

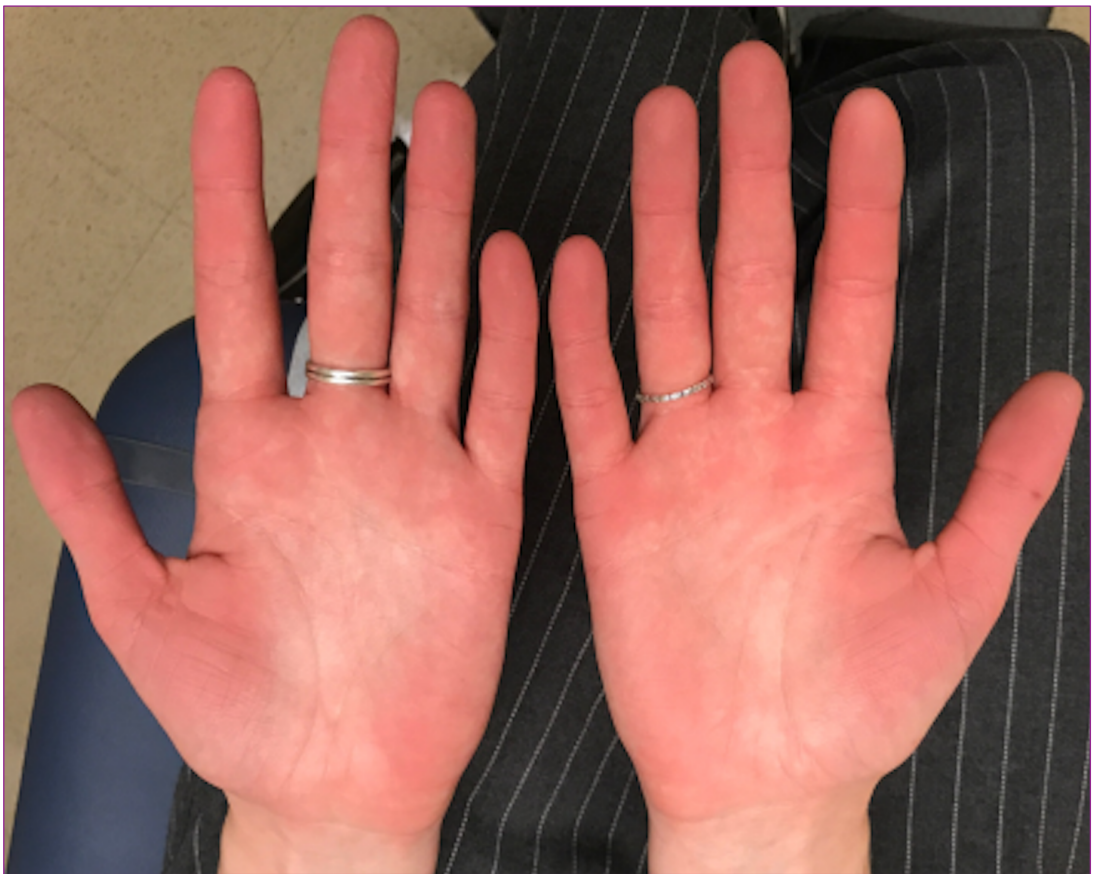

Figure 1A. Normal hands prior to water exposure.

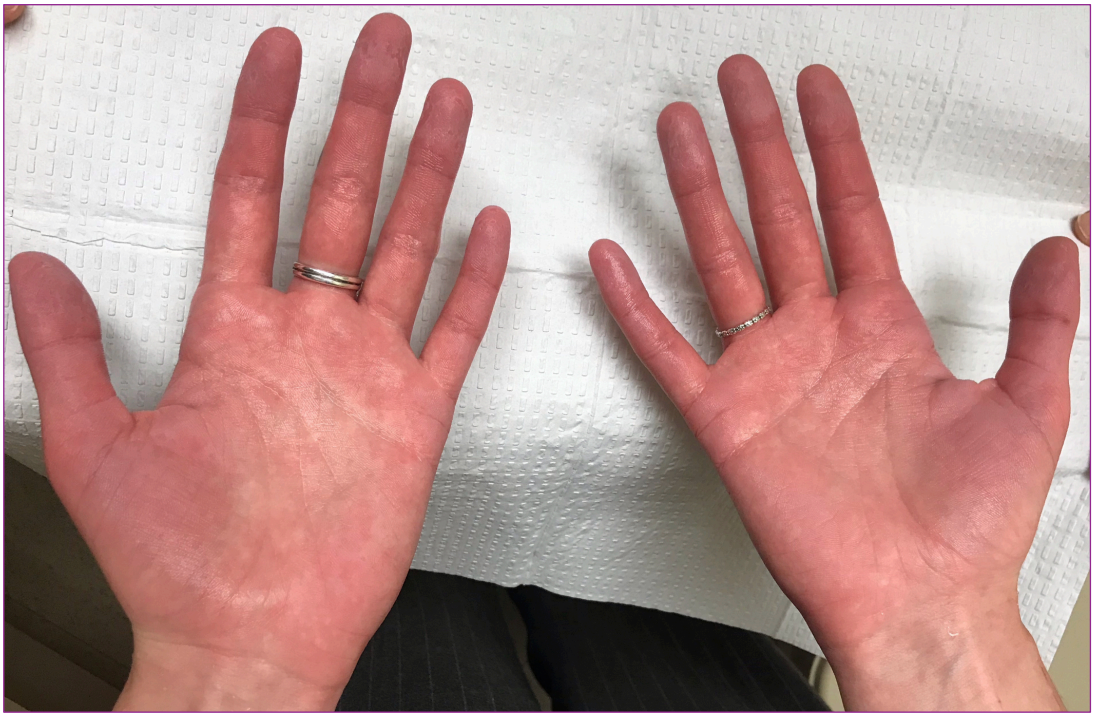

Figure 1B. Hands after one minute of water exposure.

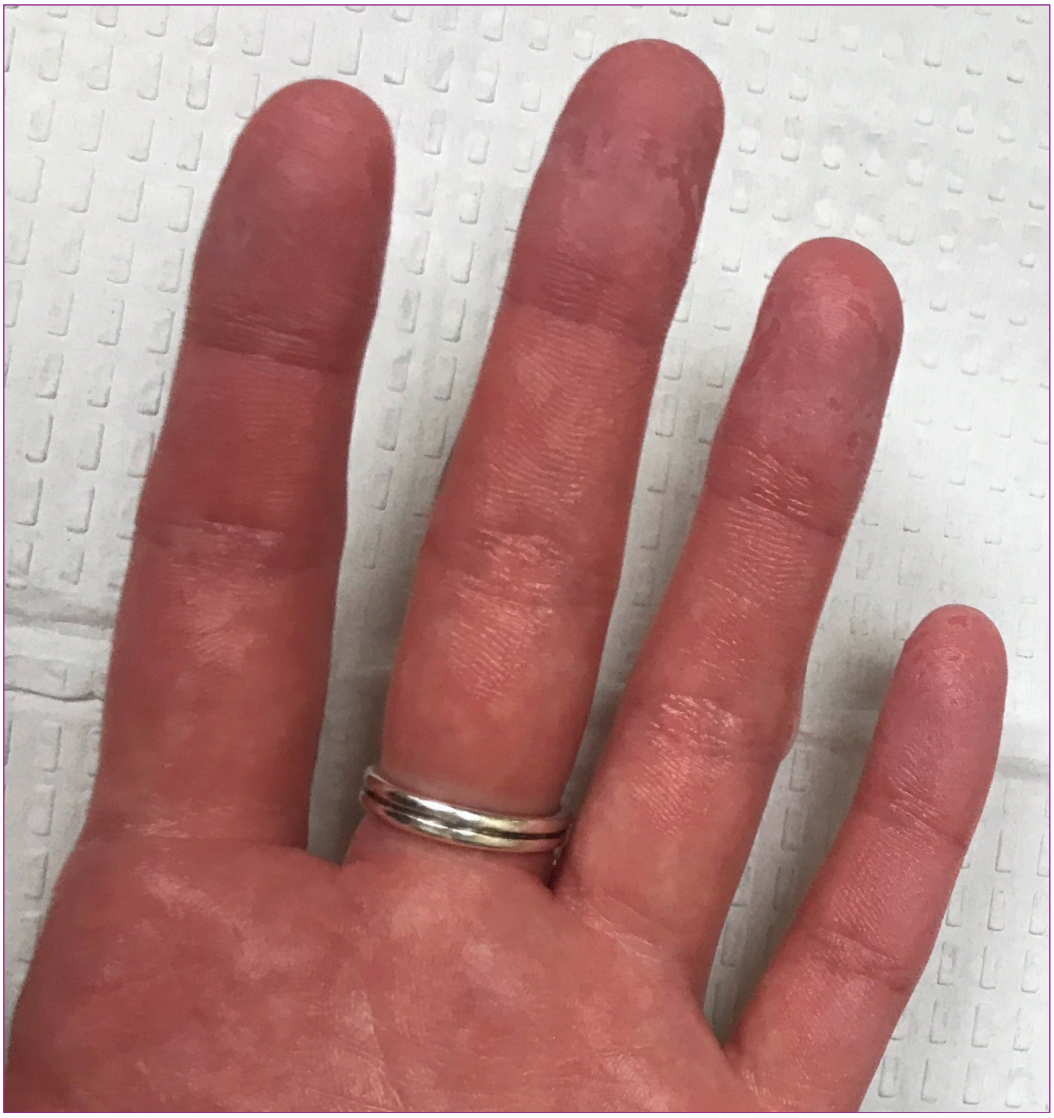

Figure 2. Close up of hands after one minute of water exposure showing cobblestone wrinkling of her palmar fingertips with shiny edematous papules coalescing into plaques.

on the acral areas that exhibit transient and recurrent white maceration after contact with water. Contrary to APPK, it does not affect palmoplantar areas and predominantly occurs in young males.[7]

Currently, there is no consensus treatment for APPK; however, several therapies have been suggested. Topical 20% aluminum hydroxide with or without a keratolytic preparation like topical urea or salicylic acid is the most frequently used treatment.[1] Botulinum toxin treatments have shown a good response in the literature, suggesting that eccrine glands and sweating are linked to the pathogenesis of APPK.[2,8,9] Alternative therapies with little benefit include antihistamines and topical steroids.[3,10] In our case, the patient failed to improve with topical aluminum hydroxide, urea cream, and botulinum toxin. Other cases have reported success using different treatments. Brazilian researchers treated a case of APPK with 5 mg of daily oxybutynin and observed

substantial improvement in symptoms within three weeks.[11] Errichetti and Piccirillo had a case where the patient was found to be resistant to topical 20% aluminum chloride but improved significantly with tap water iontophoresis.[12] Nonetheless, current treatments are often unsatisfactory, and better therapies are needed to improve the quality of life for patients with APPK.[3,10]

CONCLUSION

Ultimately, APPK is a rare condition that adversely impacts the patient's quality of life. This case of APPK presented predominantly in the fingertips and failed to improve with multiple treatments, including botulinum toxin treatment. Additional research is needed to determine the precise pathophysiology of the condition, which can help formulate an efficacious treatment for patients resistant to traditional therapy.

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