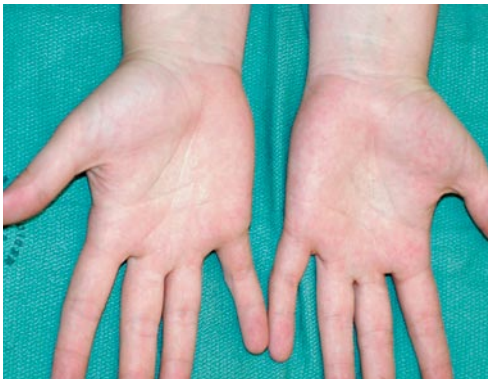
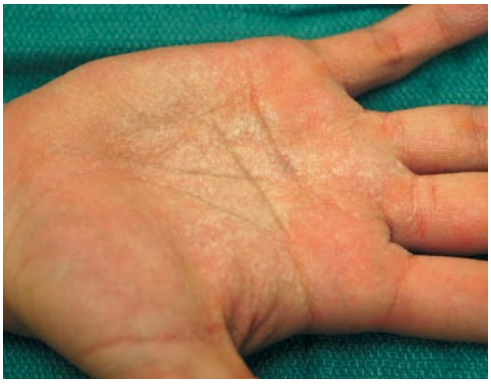
What Is Your Diagnosis?





An otherwise healthy 20-year-old white woman presented with a 1-year history of palmar lesions developing immediately after exposure to water. The initial lesions were small (1–2 mm), white, and papular. After exposure to warm water for 1 to 10 minutes, the lesions became edematous, white, translucent, and pebbly. The small papules coalesced into plaques with central puncta that could be easily peeled off. As the lesions developed, the patient experienced pain and burning. Soon after the hand was removed from water, the lesions regressed. The patient denied having similar lesions on her soles when exposed to water but did admit to a history of mild palmar hyperhidrosis. None of her family members have the same condition.

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The Diagnosis: Aquagenic Syringeal Acrokeratoderma (ASA)

In 1996, English and McCollough[1] documented the first case of transient reactive papulotranslucent acrokeratoderma in twins who presented with a severe hyperkeratotic reaction on their palms after brief exposure to water. After 3 to 5 minutes in the water, the patients developed multiple edematous, translucent, white papules with central puncta on their palms. The lesions regressed to white smooth papules after drying their hands. Since this report, there have been numerous case reports documenting patients with similar symptoms. MacCormack et al[2] termed the disorder *aquagenic syringeal acrokeratoderma* (ASA) because of the severe changes of the palmar surfaces that include keratoderma in the acral sites and the presence of dilated eccrine ducts. Additional terms used to describe this disease in the literature include *aquagenic palmoplantar keratoderma*[3] and *hereditary papulotranslucent acrokeratoderma*.[4]

ASA typically affects women in the third to fifth decades of life.[3] Inheritance either can be autosomal dominant (hereditary papulotranslucent acrokeratoderma) or autosomal recessive.[4] Acquired cases associated with atopy and thin hair also have been documented.[3] All patients with ASA develop an acute onset of palmar lesions after brief immersion in water (Figure 1). Patients may present to their physician with one hand immersed in a cup of water to demonstrate their problem. Thus, the "hand-in-the-bucket" sign may be a clinical clue for this diagnosis.[3]

Clinical symptoms of ASA include pain, burning, and a sensation of skin tightening. While the lesions tend to be nonpruritic, pruritic cases have been documented.[3] The initial lesions tend to be small (1–2 mm), white, and papular. The palms may demonstrate hyperlinearity from the coalescence of the lesions (Figure 2).[5] After 1 to 10 minutes of exposure to warm water (eg, showers, swimming, sweating), the lesions become edematous, white, translucent, and pebbly. The numerous small papules coalesce into plaques with central puncta that can be easily peeled off (Figure 3). Applying pressure to the lesions may express fluid from the puncta. The lesions regress soon after removing the hand from the inciting agent and typically clear within one hour after drying. Hyperhidrosis has been associated in patients with active disease.[2]

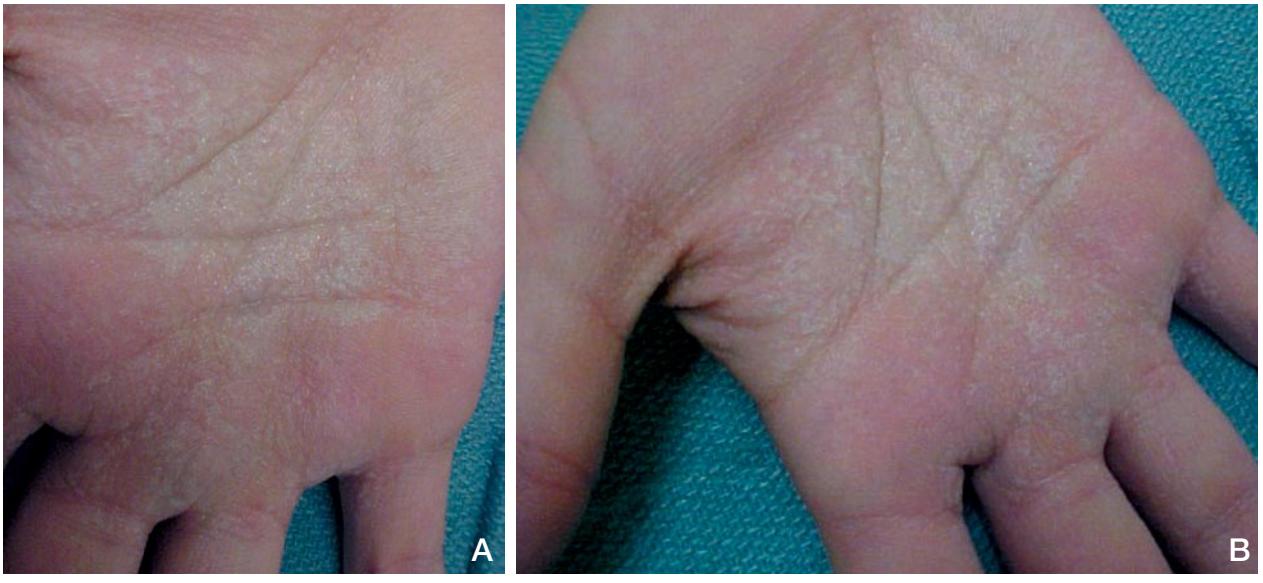

Figure 1. The patient's right palm after submersion in water (A and B).

Several cases have reported substantial plantar involvement in patients with ASA.[3]

On histopathology, these lesions typically demonstrate numerous dilated eccrine ostia and signs of orthohyperkeratosis of the epidermis.[1] There also may be hypergranulosis and acanthosis.

While the exact pathogenesis of this disorder is unknown, there have been several hypotheses. MacCormack et al[2] proposed that the condition may be related to an underlying aberration of the eccrine sweat duct. They suggest that external factors such as occlusion or friction may contribute to the dilatation of the eccrine ostia seen in this condition. Lowes et al[5] hypothesized that patients with ASA tend to have an inherent defect of their palmar and plantar stratum corneum, which is exacerbated after brief immersion in water.

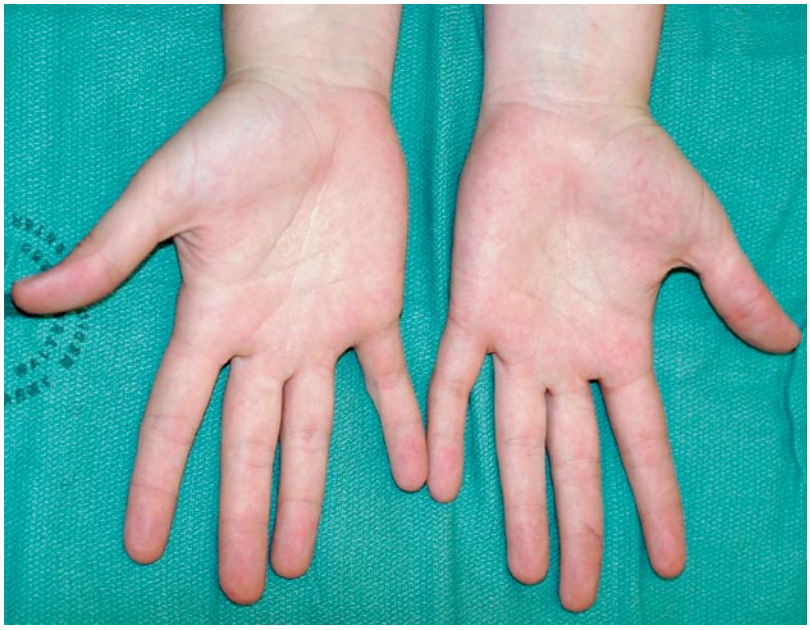

Figure 2. The patient's palms prior to submersion in water.

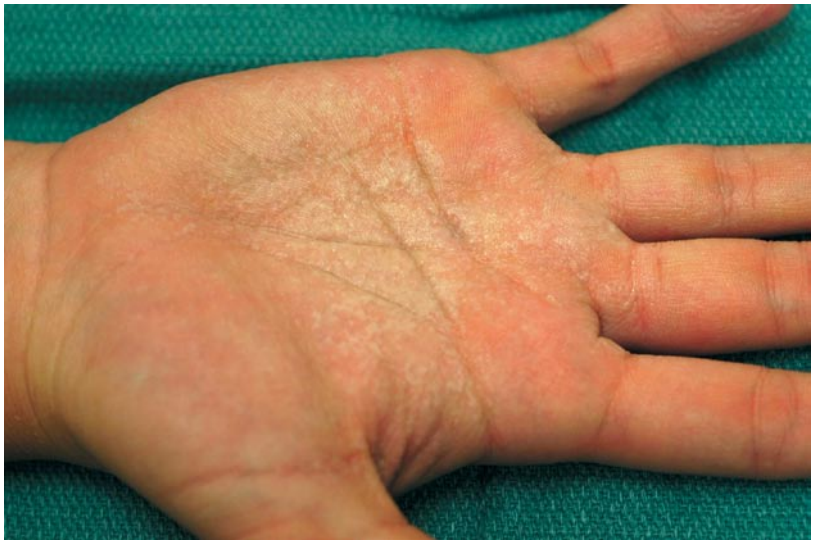

Figure 3. The patient's right palm after submersion in water for 5 minutes.

Although our patient did not present with any signs or symptoms associated with cystic fibrosis, current literature has implicated a possible association between cystic fibrosis and ASA. According to Sweeney et al,[6] patients with ASA may have a defective chloride channel, similar to cystic fibrosis, leading to a decrease in sodium chloride absorption. Because the sodium chloride concentration in the sweat is increased in patients with ASA, the hypertonic sweat creates an osmotic gradient that pulls isotonic fluids into the eccrine duct, subsequently dilating it. Lowes et al[5] reported a case in which a patient presented with both ASA and cystic fibrosis. Because many patients afflicted with ASA have exhibited hyperhidrosis, especially during periods of active disease, a potentially inherent connection between ASA and cystic fibrosis may merit further study.

There have been several suggested treatments for ASA. Yan et al[3] found aluminum chloride hexahydrate successful in treating this disorder because of the possible association with hyperhidrosis. Three patients who applied aluminum chloride on their palms showed no recurrences of their disease upon exposure to water, and the patients all exhibited remission after several months of continuous application.[3] Additionally, Yan et al[3] found partial success using a barrier topical cream containing petrolatum; however, this preparation was not well-tolerated because of the greasy nature of the application.

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