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Acquired aquagenic papulotranslucent acrokeratoderma

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Abstract

Aquagenic papulotranslucent acrokeratoderma is a rare condition with the development of white-to-transluscent papules and plaques after exposure to water. While the first report was described as an autosomal dominant hereditary condition, there have since been acquired cases reported in association with cystic fibrosis, with prior exposure to a drug, or as an idiopathic condition. We present a 24-year-old man with acquired aquagenic papulotranslucent acrokeratoderma that has been present since infancy, without a family history, without prior drug exposure, and without any personal or family history of cystic fibrosis. Thus far treatment with urea cream, calipotriene ointment, vitamin E cream, and clobetasol ointment has been ineffective. Our patient will be treated with botulinum toxin.

Case Presentation

PATIENT: 24-year-old-man
DURATION: Since infancy
DISTRIBUTION: Hands and feet

HISTORY: A 24-year-old man presented to Dermatologic Associates for evaluation of a pruritic skin condition that had affected his hands and feet since infancy. The patient notes that after exposure to water the skin of his hands and feet become white and scaly. The use of soap worsens the condition and causes more scale and more pruritus. The patient does note excessive sweating of the palms. He denies a family history of a similar condition. There is no family history of cystic fibrosis. There is no personal or family history of eczema, asthma, or rhinitis. The patient takes vitamin B and has no known medication allergies.

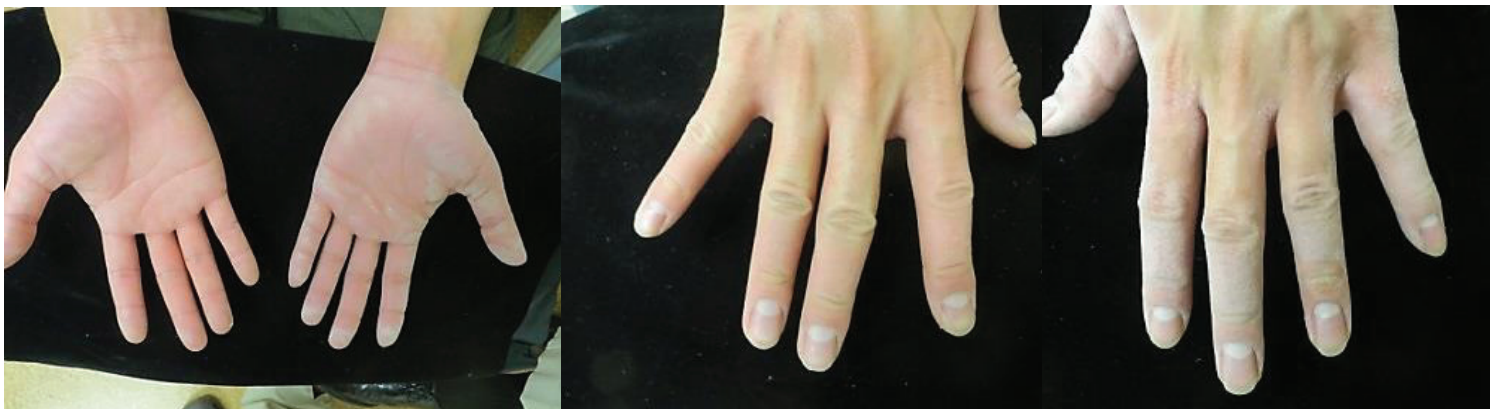

Figure 1. Erythema and scale of the dorsal and palmar aspects of the hands and dorsal and plantar aspects of the feet. Immersion of the left hand in water resulted in white papules and plaques with pruritus.

The patient was treated with urea cream and vitamin E cream in the past without improvement. Use of calcipotriene ointment provided some relief from pruritus in the past, so the calcipotriene ointment 0.005% was continued twice daily with ammonium lactate 12% daily. One month later the patient still had scale and pruritus. Clobetasol ointment twice a day was started with some further improvement in one week. Botulinum toxin injections decreased the hyperhidrosis, pruritus, and scale.

PHYSICAL EXAMINATION: There was erythema and scale of the dorsal and palmar aspects of the hands and dorsal and plantar aspects of the feet. Immersion of the left hand in water resulted in white papules and plaques with pruritus (**Figure 1**).

LABORATORY DATA: Cystic fibrosis DNA analysis for 32 CF mutations on the Universal Array Platform Regions of the CTFR gene were negative.

HISTOPATHOLOGY: None

DIAGNOSIS: Acquired aquagenic papulotranslucent acrokeratoderma

Discussion

Acquired aquagenic papulotranslucent acrokeratoderma is a rare disorder, which was first reported as part of an autosomal dominant hereditary condition in 1973 [1] and as an acquired condition in 1996 [2]. Recurrent, symptomatic keratoderma with translucent papules of acral skin develop after immersion in water with resolution shortly after drying. As the etiology is yet to be elucidated, the condition may be referred to in the literature by the descriptive names of causative factor or affected site: transient reactive papulotranslucent acrokeratoderma, aquagenic syringeal acrokeratoderma, watersport hands, aquagenic palmar wrinkling, and aquagenic palmoplantar keratoderma [3]. Thus far, there have been fewer than 50 cases reported in the literature. It is thought that due to the transient nature of the condition it is often underreported [4].

One of the difficulties in diagnosis lies in the heterogeneity of cases. While initially reported to predominately affect young females [3], a more recent case series collected in only a 13-month period showed six men were affected and four women [4]. While some patients experience burning and pain, others experience pruritus, such as our patient. Some patients are neurologically asymptomatic and are only bothered by the appearance [2-4].

While most cases are thought to be acquired and have onset in the second and third decade of life, several cases have reported a hereditary nature with onset in infancy [1, 4-8].

While the pathogenesis of this condition is not yet elucidated, many theories have been postulated from both the demographics and the histopathologic features of biopsied skin. Histopathologic features range from normal skin to dilated eccrine ducts with hyperkeratosis, compact orthokeratosis, and keratolysis of the cornified layer [3, 9]. However, the clinical presentation is considered sufficient to make the diagnosis, and therefore a biopsy was not done in our patient.

Owing to the dilation of the sweat ducts on histopathologic examination and association with hyperhidrosis, it has been postulated that there is abnormal electrolyte exchange in the affected skin when exposed to water in a parallel manner with electrolyte abnormalities in cystic fibrosis (CF). In one study of 27 patients with CF, 11 patients had aquagenic papulotranslucent acrokeratoderma (41%) although no specific mutation was identified [10]. There may be abnormal sodium chloride concentration of the sweat as reported in the case of 10-year-old boy without the common CF mutations [11]. One case report linked papulotranslucent syringeal acrokeratoderma to a mutation in the CFTR gene with F508del mutation in one allele. The patient had no family history and no other relevant symptoms; however, this is the first clue we have to true genetic etiology of the condition [12]. An early theory related the development of this condition to drug exposure with COX-2 that may induce the condition by causing an elevated skin sodium level, which also suggests that an electrolyte abnormality

in the skin is critical to this condition [13].

The response to treatment is variable. General treatment is symptom directed, such as antihistamines, topical glucocorticoids, urea, and salicylic acid, or else targets the commonly associated hyperhidrosis, such as aluminum hydroxychloride or botulinum toxin or tap water iontophoresis [3, 4, 14, 15]. There has been a case report of failure of treatment with all of the above mentioned therapies [16]. So far our patient has failed all treatment modalities tried, but we are hoping that botulinum toxin treatment of the hyperhidrosis will provide some relief.

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