Case Report

Idiopathic Aquagenic Syringeal Acrokeratoderma

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Abstract

Aquagenic syringeal acrokeratoderma (ASA), also known as transient reactive papulotranslucent acrokeratoderma, is an uncommon disorder that usually affects the palms. It is characterized by transient, translucent, white papules and plaques that develop upon exposure to water. Skin lesions are typically accompanied by itching and burning sensation. While many theories exist in regard to the etiology of this condition, we present a case of ASA that appears to be idiopathic in nature.

Keywords: Acrokeratoderma, aquagenic, hyperwrinkling, syringeal, translucent

Introduction

Aquagenic syringeal acrokeratoderma (ASA) is a rare condition that predominantly affects adolescent and young adult women.[1-3] Over 30 cases of ASA have been reported until 2012.[4] However, in a recent publication in 2015, Erturk-Ozdemir *et al*. presented a case series of ten patients, diagnosed with ASA over a short period of 13 months. Hence, we believe that this disorder has probably been underdiagnosed likely due to the nonlife-threatening clinical presentation of the lesions and its spontaneous resolution. ASA usually presents clinically as bilaterally symmetric pruritic palmar translucent white papules and plaques. Those lesions appear 2–10 min after exposure to water and last for 20–30 min. The soles are involved less frequently than the palms. The etiology of ASA is unknown, but several pathogenic mechanisms and associations have been proposed.[5,6] We present the case of a patient with ASA of unknown etiology.

Case Report

A 22-year-old Caucasian female presented with concerns of dryness, roughness, whitening, and hardening of her palms with handwashing or after showering. She stated that these changes to her palms are usually accompanied by a sensation of intense itch. She denied any known history of inherited disorders or medication use. She stated that she first noticed the unusual appearance of her palms approximately 5 years ago. On physical examination, white rugated plaques were noted on her bilateral palms 2 min after submersion in a basin of water in the office. It took about 30 min for these plaques to resolve after drying.

A clinical diagnosis of ASA was made based on her history of worsening with immersion in water and by observation of the papules on the palms in the office after submersion in a water basin [Figures 1 and 2].

We advised her to apply 20% aluminum chloride to her palms nightly for 1 month. We recommended that her hands should be completely dry before applying the medication to avoid risk of irritation.

After approximately 1 month, the patient returned for follow-up. She reported near-complete resolution of the white plaques and complete resolution of the associated pruritus. At that point, we advised her to slowly taper use until she would be able to successfully discontinue without recurrence. Photographs were re-taken for comparison to her hands pretreatment [Figure 3].

Discussion

ASA is an uncommon acquired dermatologic disorder. The literature describes these patients as presenting with

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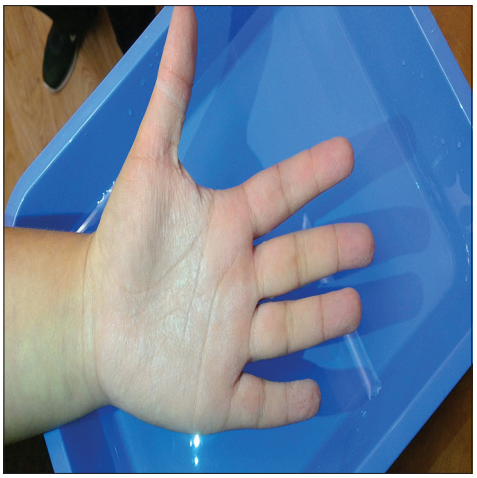

Figure 1: White translucent papules on the left palm

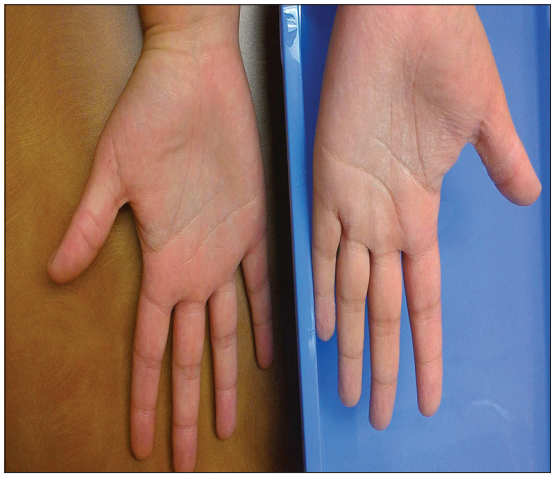

Figure 2: Morphological comparison of the right palm not dipped in a bowl of water with the left palm dipped in a bowl of water

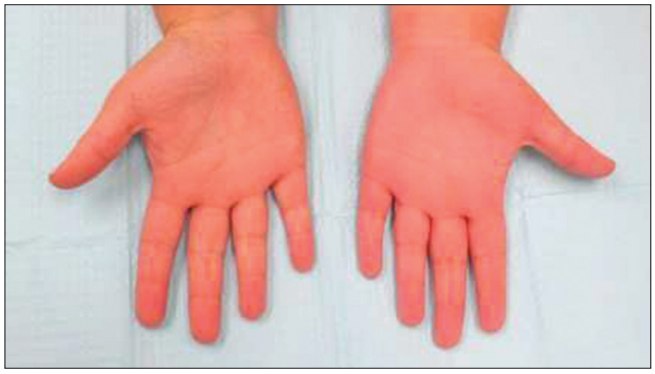

Figure 3: The wet and dried left and right palms after treatment

pitted, flat-topped, or translucent papules, with pebbly or white, prominent eccrine pores that are macerated in appearance and appear on the hands and feet. Due to the appearance of these papules with submersion in water, the disorder is popularly called the hand-in-the-bucket sign.[7] ASA was described for the first time by English and McCollough in 1996 in two sisters.[8] This rare disorder was named by the authors as transient reactive papulotranslucent acrokeratoderma, to emphasize the transient nature of the translucent papules shortly after immersion in water.[7]

ASA is different from the hereditary form, called hereditary papulotranslucent acrokeratoderma (HPA), which is characterized by persistent white papules on the palms and soles, hair abnormalities, and atopic diathesis.[7] HPA is an autosomal dominant disorder and it was first reported in 1973 by Onwukwe *et al*. White papules occur in puberty and are concentrated on the margins of palms and soles.[7] Lesions are usually persistent, and the appearance of papules is not associated with water exposure. ASA has been described under various names, including transient reactive papulotranslucent acrokeratoderma, aquagenic keratoderma, transient aquagenic palmar hyperwrinkling, and early aquagenic wrinkling.[9]

Although the exact etiology is unknown, ASA has many proposed causes. It has been reported to be induced by cyclooxygenase-2 (COX-2) inhibitors, such as rofecoxib and celecoxib. It has also been induced by other drugs including aspirin, acetaminophen, ascorbic acid, and clarithromycin.[6] The proposed mechanism behind the pathogenesis of drug-induced ASA involves increased sodium retention in the epidermal keratinocytes and therefore increased capacity of water uptake in the stratum corneum. This mechanism is similar to that seen with renal side effects of COX-2 inhibitors.[10] ASA has also been associated with hyperhidrosis, palmar erythema, allergic rhinitis, bronchial asthma, cystic fibrosis, and malignant melanoma.[11]

The pathogenesis of ASA remains unclear, but it may be a primary keratoderma or related to an acquired sweat gland abnormality since it has a strong association to cystic fibrosis.[12] Several studies have found that it is present in about 40%–84% of cystic fibrosis patients and also in carriers.[6,13] Improvement of the ASA lesions with sweat gland-suppressing treatment options such as botulinum toxin injection and topical aluminum chloride suggests an etiopathogenesis of aberration of eccrine sweat gland function.[14] The diagnosis of ASA is mostly clinical, but histology and dermoscopy may also be useful. Histopathologically, two case reports on ASA noted clear cell change and vacuolization of epithelial cells. Other histologic findings include mild orthokeratotic hyperkeratosis and dilated eccrine ducts.[9] On dermoscopy, marked enlargement of the sweat duct puncta is seen compared with those in unaffected palmar regions, reflecting the dilated and tortuous acrosyringium.[14] Differential diagnosis for ASA is cutaneous disease associated with water contacts including xerosis, aquagenic urticaria, HPA, aquagenic pruritus, and aquadynia.[15]

ASA causes a burning and tightening sensation that may lead to a considerably impaired quality of life. Fortunately, the condition is characterized by a mild course and a good

prognosis. Spontaneous resolution has been reported in four cases, and some cases have been found to clear after a few years.[16-18] Treatment modalities available include aluminum chloride hexahydrate, urea with salicylic acid, or formalin in alcohol. In cases accompanied by hyperhidrosis, botulinum toxin injections proved to be effective.[7] Endoscopic thoracic sympathectomy has been effectively used for long-term treatment in patients with ASA associated with severe palmar hyperhidrosis refractory to treatment.[14] In data collected by Houle *et al*. in 2010, of 59 reported cases of ASA in the literature, four cases spontaneously resolved, four drug-related cases resolved with withdrawal, and the rest resolved or improved with treatment. There were seven cases which did not improve with treatment with aluminum-based therapy or antihistamine. However, it is very likely that the patients would have responded to treatment if more aggressive measures such as botulinum toxins, salicylic acid, or sympathectomy were employed.[19]

Conclusion

ASA remains a rare but interesting disorder with an etiology that is not completely understood. Our patient's history and physical examination did not suggest any known etiology. Our patient had no family history of cystic fibrosis or personal history of associated conditions or medication use. Genetic testing may be indicated in her case for cystic fibrosis or any other associated diseases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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