CASE REPORT

Aquagenic Wrinkling of the Palms, two new cases from Saudi Arabia

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Abstract Aquagenic Wrinkling of the Palms (AWP) is a rare dermatologic disease. It was first described four decades ago as a potential test for Cystic Fibrosis (CF) by detecting “skin wrinkling” in response to water immersion. We are reporting two cases of AWP diagnosed at Derma Clinic, Riyadh, Saudi Arabia. The first case was a 9-year-old girl and the second case was a 17-year-old adolescent male. The second case was associated with palmer hyperhidrosis. Both patients denied any medication use. Additionally, signs and symptoms suggesting CF were lacking. Local dermatologists should be aware of rare conditions such as AWP and lack of previous reports from the region may indicate under-reporting rather than lack of cases.

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1. Introduction

Aquagenic Wrinkling of the Palms (AWP), also called aquagenic palmoplantar keratoderma and transient reactive papulo-translucent keratoderma, is a rare dermatologic disease. It was first described in 1974 as a potential test for Cystic Fibrosis (CF) by detecting “skin wrinkling” in response to water immersion (Elliott, 1974). AWP is clinically presented as poorly demarcated edematous white papules and plaques of the palms (and soles) that develop subsequently to water exposure (Tolland et al., 2010). The pathophysiology of AWP is not fully understood; however, it is believed to result from abnormal electrolyte fluxes that result in sodium retention within epidermal keratinocytes and the osmotically induced cell volume increases (Katz et al., 2005). This may explain the apparent association of AWP with conditions with increased sweat electrolyte concentration such as CF and with certain drug administration or with conditions of increased water absorption due to an increased sweat quantity, such as hyperhidrosis (Seitz et al., 2008). Additionally, mutation of the CF gene was thought to predispose to AWP (Berk et al., 2009). A number of reports described the AWP in individual patients, primarily adolescent females from Western countries (Seitz et al., 2008; Syed et al., 2010). To our knowledge, AWP was never described in Saudi Arabia or any Middle Eastern country.

2. Case 1

A nine-year-old girl was brought to the clinic because of persistent skin rash on her palms which becomes more prominent
after exposure to tap water and continues for a few hours afterward. The girl had been experiencing the symptoms for 3 years. To confirm the diagnosis, we asked the girl to immerse her palms in a jar containing tap water kept at comfortable room temperature (at 23 °C). The rash became more prominent within 3 min and more after 5 min. The rash was bilaterally distributed over the palmar surface of both hands as numerous, tiny, soft, coalescent, whitish papules (Fig. 1).

The patient stated that the lesion is painless, non-itchy, but causing mild discomfort. There was no past history of medical or surgical comorbidity. There was lack of signs and symptoms suggesting palmar hyperhidrosis or CF (such as intermittent chronic fatigue, diarrhea, acholic stools, pneumonia, hepatorenal syndrome, nasal polyps, and pancreatic insufficiency). The mother denied any use of medications such as nonsteroidal anti-inflammatory agents and antibiotics and any history of atopic dermatitis. The patient denied history of additional wrinkling complaints in her soles. The mother said that there was no family history of a similar rash in her family members but there was a positive consanguinity, with both parents being first-degree relatives. She refused to let her girl give skin biopsy or to do genetic testing, even after explaining the strong possibility of CF as long as the daughter is asymptomatic. A treatment with topical 20% aluminum chloride solution was prescribed. The medication was tolerable and effective in improving the symptoms.

3. Case 2

A 17-year old healthy male was seeking help for excessive sweating on both palms (palmar hyperhidrosis). Upon examination, the patient had a similar condition as the first patient, on both palms but more prominent on the left than the right (Fig. 2). He denied history of any drug intake and he claimed that none of his family members experienced the same rash. The patient was treated with an intradermal injection of both palms with 100 IU each of botulinum toxin A (BOTOX®, Allergan, Inc., Irvine, CA, USA) that completely relieves the symptoms in a week.

4. Discussion

We are reporting, for the first time in the Middle Eastern region, two cases of AWP in a school-age girl and adolescent male. In a review of the AWP cases published before 2008, approximately half of the cases were diagnosed in children (<18 years) (Seitz et al., 2008). Both genders were shown to be equally affected by AWP in a study that estimated the prevalence of AWP among 45 cases of CF (Berk et al., 2009). However, higher predominance of the AWP among females has been shown in a couple of reports reviewing published case reports (Seitz et al., 2008; Syed et al., 2010). Association of AWP with palmar hyperhidrosis has been reported before (Seitz et al., 2008). It was suggested that AWP is etiologically related to hyperhidrosis that is characterized by abnormal electrolyte fluxes (Kabashima et al., 2008).

Although most of the previous reports used the water immersion test to diagnose AWP, there was no consensus on the cut point duration of the test, the temperature of the tap water, or the percentage of the affected palms (Berk et al., 2009). AWP is treated with medications that decrease the hyperkeratosis associated with the condition, such as 20% aluminum chloride solution or 12% ammonium lactate creams or petroleum jelly (Syed et al., 2010). Additionally, providing a...
water barrier to prevent exposure was shown to be effective (Syed et al., 2010).

Although the association between CF and AWP, that has been suggested since the first report, was confirmed in later reports (Tolland et al., 2010; Berk et al., 2009; Arkin et al., 2012), it is not clear whether AWP is a characteristic and discriminating feature of CF or whether patients without CF who present with AWP should be tested for CF carrier status. Additionally, idiopathic AWP without CF diagnosis has been reported in a number of reports (Davis and Woody, 2004; Trotot et al., 2012; Vale and Adam, 2012). The inability to confirm the CF status in our patients is a limitation of the current report, but it also highlights the difficulty a dermatologist may face to convince the parents with the importance to investigate rare diseases.

In conclusion, we are reporting two new cases of AWP in Saudi Arabia, one of them was associated with palmar hyperhidrosis. Although signs and symptoms suggesting CF were lacking, we could not rule out CF. Local dermatologists should be aware of rare conditions such as AWP and lack of previous reports from the region may indicate under-reporting rather than lack of cases.

Conflict of interest

None declared.

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Figure 2 Image of the AWP before botulinum toxin A injection (A) and after 1 week (B).