DOI: http://dx.doi.org/10.3315/jdcr.2016.1223

Journal of Dermatological Case Reports

Sporadic and familial cases of aquagenic keratoderma

Hülya Nazik[1], Selçuk Nazik[2], Feride Gül Çoban[3], Betül Demir[4]

1. Bingöl State Hospital, Department of Dermatology, Bingöl, Turkey;
2. Bingöl Maternity and Children's Hospital, Department of Infectious Diseases and Clinical Microbiology, Bingöl, Turkey;
3. Elazığ Education and Training Hospital, Department of Dermatology, Elazığ, Turkey;
4. Fırat University, Department of Dermatology, Elazığ, Turkey.

Corresponding author:

Hülya Nazik, MD

Bingöl State Hospital

Department of Dermatology

Bingöl, Turkey

E-mail: *dr.hulyagul@hotmail.com*

Keywords:

acitretin aquagenic keratoderma, retinoids, treatment

Abstract

Background: Aquagenic keratoderma is a dermatosis characterized by transient whitish and transluscent hyperwrinkling after water exposure. The aim of the current report was to present a sporadic and familial cases of aquagenic keratoderma.

Observation: Sporadic Case: A 38-year-old female patient presented with eruption in the right hand after exposure to water. The patient was placed on systemic acitretin therapy with the diagnosis of idiopathic acquired aquagenic keratoderma. No recurrence occurred during a 6-month follow-up period. Familial Cases: A 55-year-old male patient, who was engaged in fishery, presented to the outpatient clinics of the department of dermatology due to whitish vesicles in the palms of both hands. It was realized that the father, sister, and brother of the patient had similar complaints. The cases were thought to have familial aquagenic keratoderma; however acitretin therapy could not be initiated due to elevated alanine aminotransferase and triglyceride levels. Topical application of salicylic acid 10% and 10% urea containing lotions was effective but did not prevent recurrence.

Conclusion: Systemic acitretin may be an effective agent in the treatment of aquagenic keratoderma, and topical application of 10% salicylic acid and 10% urea-containing lotion did not prevent recurrence. (*J Dermatol Case Rep*. 2016; 10(1): 10-13)

Introduction

Aquagenic keratoderma (AK) was first described in 1974 in a patient with cystic fibrosis (CF).[1] It is a rare form of acquired palmoplantar keratoderma with an unknown etiology and it is also called "aquagenic syringeal acrokeratoderma", "aquagenic wrinkling of the palms" and "aquagenic palmoplantar keratoderma". Transient lesions occur after water exposure.[1,2] The aim of the current report was to present a rare case of unilateral involvement and familial cases of AK and emphasize the efficacy of new treatment regimens.

Case with unilateral involvement

A 38-year-old female patient presented to our outpatient clinics complaining of eruption in the right hand occurring and water exposure. The eruption was white in color, raised from the skin, slightly pruritic, painful and extended from the inner surfaces to the lateral aspects of the fingers. The patient reported worsening of complaints with frequent exposure to water such as during house cleaning and dish washing and the eruptions showed regression and scaling when the hands were dry. The patient did not report any relief with topical steroids having various potency, antiperspirants, oral and topical antifungal therapies. The patient denied similar complaints in the family members and also she did not report atopy, drug use, or hyperhydrosis. Both hands appeared normal on dermatological examination. After water exposure of the patients' hand for five minutes at room temperature, whitish, macerated, translucent and hyperkeratotik plaques with punctuate holes were observed on the palmar and lateral aspects of the third and fourth fingers in the right hand (Fig. 1A, B). The patient did not have palmar involvement and there were no other papular lesions except the above-mentioned plaques. The lesions completely disap-

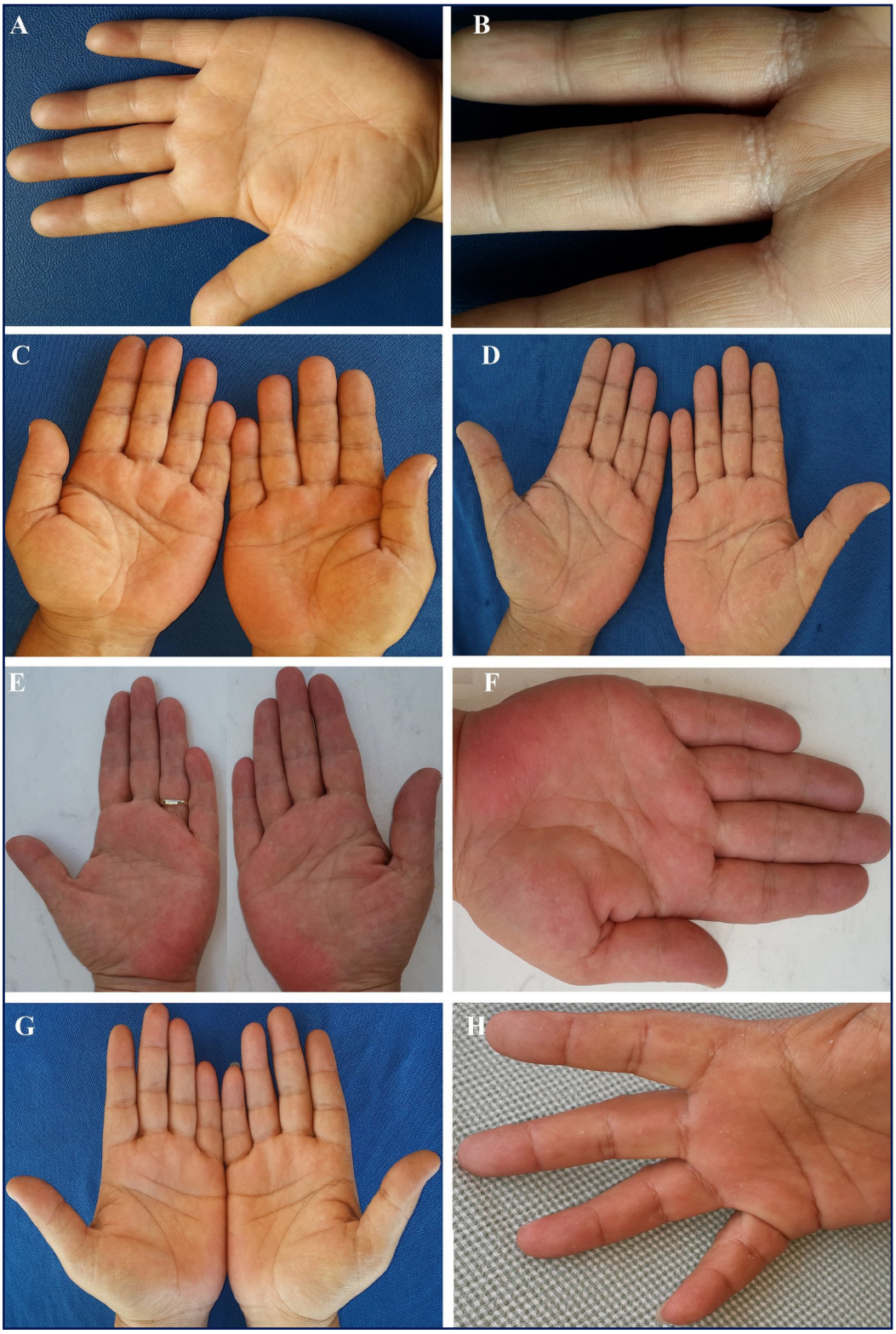

Figure 1

Aquagenic keratoderma photos. (A) Normal appearance of the right hand in sporadic case before water exposure; (B) Whitish and hyperkeratotic macerated plaques on palmar surface of the third and fourth fingers of the right hand after water exposure for five minutes at room temperature in sporadic case; (C) Appearance of whitish and scattered hyperkeratotic papules in a 55-year-old familial case before water exposure; (D) Macerated papules with pebblestone appearance having punctuate holes scattered bilaterally on the palms of the hand after water exposure; (E) Appearance in female familial case before water exposure; there was palmar erythema; (F) Macerated papules with pebblestone appearance having punctuate holes scattered on the palms; (G) Appearance of whitish hyperkeratotic papules before water exposure in a 50-year-old male familial case; (H) Macerated papules with pebblestone appearance having punctuate holes scattered bilaterally on the palms.

peared approximately one hour after when the hands of the patient became dry. Based on these findings, the patient was diagnosed with idiopathic acquired aquagenic keratoderma.

Familial cases

A 55-year-old male patient, who was engaged in fishery, presented to the outpatient clinics of the department of dermatology due to whitish vesicles in the palms of both hands. The complaints of the patient disappeared approximately one hour after when the hands became dry, and there was a family history in the father, sister, and brother. The patient's father had died of myocardial infarction eight years before, and other relatives of the patient were invited to the outpatient clinics. 57-year-old female patient had complaints for 40 years, while 50-year-old male patient had complaints for 35 years. All three patients had a history of hypertension and alcohol use, and there was no history of hyperhydrosis or atopy. Dermatologic examination in three patients revealed palmar erythema. Water tank test was performed in these patients (2). Three patients developed macerated papules with pebblestone appearance having punctuate holes scattered bilaterally on the palms of the hand after water exposure (Fig. 1C, D, E, F, G, H). The lesions showed regression after one hour when the hands of the patients were dry. It was realized that the patients did not receive an efficency therapy until that day.

Lipid profile, aspartate aminotransferase (AST), alanine aminotransferase (ALT), creatinine, and blood urea nitrogen were tested in all patients before administering any therapy. Serum biochemistry parameters were within normal ranges in the first patient. ALT and triglyceride levels were at the upper limit in three familial cases. The first patient was placed on therapy with systemic acitretin (20 mg/day). This patient was asked to come for control visit one month after. Control blood tests were normal, and all complaints disappared. Acitretin dose was titrated to 10 mg/day and the patient was followed for six months, and no recurrence was observed. No clinical side effects or laboratory abnormalities were observed other than dry lip. Hepatotoxic acitretin therapy was not considered in familial cases due to significant history of alcohol use in these cases, and they were placed on a therapy involving topical salysilic acid 10% and 10% urea-containing lotions. At two-month control visit, the patient that was involved in fishery reported longer periods for the occurrence of the lesions and he reported partial relief with the therapy. The other two familial cases reported relief treatment-on periods but the lesions showed recurrence in treatment-off periods.

Discussion

Aquagenic keratoderma is a dermatosis characterized by transient whitish and transluscent hyperwrinkling occurring bilaterally in the palms, fingers, and rarely soles of the feet after water exposure.[3,4] Although the etiology of the disease is unknown, some cases are associated with CF while others are associated with the use of certain drugs such as aspirin, indomethacin, salazopyrin, rofecoxib, and celecoxib.[4-6] The first patient did not have a history of drug use, while all familial cases were using antihypertensive drugs and alcohol. None of the patients exhibited any findings consistent with CF. Familial cases were not considered to be associated with antihypertensive drugs and alcohol use due to disease onset after adolescence.

The condition often occurs sporadically in young females.[7] The complaints of the first patient started one year before admission and there were no family members having similar symptoms. Onwukwe *et al*. defined this disease as atopy-related condition with autosomal dominant inheritance pattern occurring after adolescence.[8] In familial cases reported in the literature, the lesions often occur during pubertal period. The fact that father and all children are affected by this condition suggests autosomal dominant inheritance. Aquagenic keratoderma occurs bilaterally in the majority of the cases and rarely occurs unilaterally in the palms and fingers of the hands and also involves soles of the feet. The lesions distributed unilaterally in the first case and involved inner and lateral aspects of the fingers without involving the palms. There was a bilateral and symmetric involvement in familial cases. The condition can be asymptomatic in familial cases or display a pruritic and slightly painful course as in the first case.

The diagnosis is established by observing the lesions occurring within a couple of minutes after immersing the hands of the patient in a water tank.[2] Histopathological examination may show orthokeratotic hyperkeratosis and dilated eccrine channels; however, none of these findings are pathognomonic.[9] Medical history and clinical findings often lead to the diagnosis of aquagenic keratoderma. Many possible reasons have been put forward to explain the pathogenesis of the condition including eccrine gland and nerve dysfunction, hyperhydrosis, barrier defect in stratum corneum, obstruction of eccrine channel opening, and eccrine channel wall weakness. Despite all these hypotheses, the cause of the disease has not been fully understood. [2,7,10,11]

Various preparations have been used in the treatment of aquagenic keratoderma such as aluminum salts, antihistamines, botulinum toxin injection, 5% salicylic acid ointments, solid vaseline mixture containing 5% salicylic acid, mixture of mometasone furoate ointment and solid vaseline, and 20% urea-containing creams.[12,13] Retinoids and acitretin are known to be effective in patients with defective keratinization.[14,15] Systemic acitretin therapy provided favorable outcomes in the sporadic case, whereas topical 10% salicylic acid and 10% urea-containing lotions provided only temporary relief in regular use; however, recurrence was observed with interruption of topical therapy.

Conclusion

In conclusion, systemic acitretin is an effective agent in the treatment of AK, and topical application of 10% salicylic acid and 10% urea-containing lotion did not prevent recurrence.

References

1. Elliott RB. Letter: Wrinkling of skin in cystic fibrosis. *Lancet*. 1974; 2: 108. PMID: 4137036.
2. Houle MC, Al Dhaybi R, Benohanian A. Unilateral aquagenic keratoderma treated with botulinum toxin A. *J Dermatol Case Rep*. 2010; 4: 1-5. PMID: 21886737.
3. Yan AC, Aasi SZ, Alms WJ, James WD, Heymann WR, Paller AS, Honig PJ. Aquagenic palmoplantar keratoderma. *J Am Acad Dermatol*. 2001; 44: 696-699. PMID: 11260552.
4. Luo DQ, Li Y, Huang YB, Wu LC, He DY. Aquagenic syringeal acrokeratoderma in an adult man: Case report and review of the literature. *Clin Exp Dermatol*. 2009; 34: e907-909. PMID: 20055864.
5. Gündüz O, Ozsaraç KÇ, Ercin ME. Aquagenic palmar wrinkling induced by combined use of salazopyrin and indomethacin. *Case Rep Dermatol*. 2013; 5: 21-26. PMID: 23466824.
6. Garçon-Michel N, Roguedas-Contios AM, Rault G, Le Bihan J, Ramel S, Revert K, Dirou A, Misery L. Frequency of aquagenic palmoplantar keratoderma in cystic fibrosis: A new sign of cystic fibrosis? *Br J Dermatol*. 2010; 163: 162-166. PMID: 20302572.
7. Syed Z, Wanner M, Ibrahimi OA. Aquagenic wrinkling of the palms: A case report and literature review. *Dermatol Online J*. 2010; 16: 7. PMID: 20673535.
8. Onwukwe MF, Mihm MC Jr, Toda K. Hereditary papulotranslucent acrokeratoderma: A new variant of familial punctate keratoderma. *Arch Dermatol*. 1973; 108: 108-110. PMID: 4716729.
9. Rongioletti F, Tomasini C, Crovato F, Marchesi L. Aquagenic (pseudo) keratoderma: A clinical series with new pathological insights. *Br J Dermatol*. 2012; 167: 575-582. PMID: 22512866.
10. Coelho-Macias V, Fernandes S, Lamarão P, Assis-Pacheco F, Cardoso J. Aquagenic keratoderma associated with a mutation of the cystic fibrosis gene. *Rev Port Pneumol*. 2013; 19: 125-128. PMID: 23602165.
11. Xia Q. Aquagenic acrokeratoderma: Case report with no involvement of the palms. *Int J Dermatol*. 2012; 51: 1391-1393. PMID: 23067094.
12. Errichetti E, Piccirillo A. Aquagenic keratoderma treated with tap water iontophoresis. *Indian J Dermatol*. 2015; 60: 212. PMID: 25814730.
13. Berna Aksoy B, Hapa AA. Idiopathic Acquired Aquagenic Keratoderma Localized to Dorsum of the Hands. *Turkderm*. 2010; 44: 224-228.
14. Capella GL, Fracchiolla C, Frigerio E, Altomare G. A controlled study of comparative efficacy of oral retinoids and topikal betametasone/salisylic acid for chronic hyperkeratotic palmoplantar dermatitis. *J Dermatolog Treat*. 2004; 15: 88-93. PMID: 15204158.
15. Thestrup-Pedersen K, Andersen KE, Menné T, Veien NK. Treatment of hyperkeratotic dermatitis of the palms (eczema keratoticum) with oral acitretin. A single-blind placebo-controlled study. *Acta Derm Venereol*. 2001; 81: 353-355. PMID: 11800144.