



Acquired aquagenic acrokeratoderma: A case series

Akkiz akuajenik akrokeratoderma: Olgu serisi

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Abstract

Aquagenic syringeal acrokeratoderma (ASA) is a rare kind of an acquired keratoderma that predominantly affects adolescents and young females. The etiology is unknown. Clinically, ASA is characterized by transient edematous white papules and plaques occurring a few minutes after exposure to water. It is most commonly localized on the palms, but may also affect the plantar area and dorsum of the hand. Despite the female predominance mentioned in the literature, the aim of this study was to investigate the clinical characteristics of six male patients diagnosed with ASA in our clinic.

Keywords: Aquagenic syringeal acrokeratoderma, male, dorsum of the hand

Öz

Akuajenik siringeal akrokeratoderma (ASA); etiyolojisi bilinmeyen, sıklıkla adölesan ve genç erişkin kadınları etkileyen, nadir, edinsel bir keratoderma türüdür. Klinik olarak suyla temastan birkaç dakika sonra ortaya çıkan, geçici ödematöz beyaz papül ve plaklarla karakterizedir. Sıklıkla palmar yüzeylerde lokalizedir, el dorsalleri ve ayak tabanını da etkileyebilir. Bu çalışmada; literatürde kadın predominansı üzerinde durulmasına rağmen, kliniğimizde ASA tanısı alan altı erkek hastanın klinik özelliklerinin incelenmesi amaçlanmıştır.

Anahtar Kelimeler: Akuajenik siringeal akrokeratoderma, erkek, el dorseli

Introduction

Aquagenic syringeal acrokeratoderma (ASA) is a rare kind of acquired palmoplantar keratoderma with unknown etiology characterized with edematous white papules and plaques which occur following contact with water for 2-4 minutes^{1,2}. ASA was described by English and McCollough in 1996 for the first time as "transient reactive papulo-translucent acrokeratoderma"³. Although the lesions are frequently observed on the palmar region, the dorsa of the hand and feet may also be affected^{1,2,4}. ASA usually affects adolescent girls and young adult women. A limited number of male cases has been reported^{1,3,5}.

In this article, we aimed to present the clinical and laboratory findings of the male patients who were diagnosed with ASA and treated in our clinic in comparison with the literature data.

The patients who were clinically and/or histopathologically diagnosed with ASA in our outpatient clinic between 2011 and 2014 were examined retrospectively. At the time of outpatient clinic admission, the patients were informed that data would be obtained from their files and informed consent was obtained. The patients were examined in detail in terms of gender, age at the time of onset of lesions, localization of lesions, persistence of lesions, accompanying symptoms or morbidity, family history and treatment response.

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Case Report

A total of fifty patients who developed whitening in the hands and feet a short time after contact with water and were clinically and/or histopathologically diagnosed with ASA were included in the study. The demographic properties, clinical characteristics and therapies administered are shown in Table 1. All patients were male and they were aged between 24 and 45 years (mean age: 31,6). The disease period ranged between 2,5 months and 145 years. On dermatologic examination, white, keratodermic, macerated plaques were observed symmetrically on the dorsa of the hands, fingertips, volar regions and palms following contact with water for a short period. Dilated spaces were observed inside these plaques. It was found that these symptoms regressed in 30-60 minutes. Four patients (66.6%) were diagnosed clinically as having ASA, while the diagnosis was also supported histopathologically in two patients (33.4%). The lesions were localized symmetrically on the palmar regions in four patients (66.6%) (Figure 1), symmetrically on the dorsa of the hands in two patients (33.4%), symmetrically on the fingertips in one patient (16.6%) (Figure 2) and symmetrically on the volar regions in one patient (16.6%). Burning sensation (50%; n=3) and mild hyperhydrosis (50%; n=3) were the most common accompanying symptoms. The complaints usually occurred 10 seconds-5 minutes following exposure to tap water and regressed in 30-60 minutes after elimination of contact with water. The bucket test revealed macerated, edematous papules, plaques and dilated spaces in all patients. Complete response to 20% aluminium chloride treatment was observed in five of the patients (83.3%) and partial response was observed in one patient (16.7%).

Discussion

ASA is a rare form of palmoplantokeratoderma characterized with transparent, white papules which occur as a result of short-term contact with water with bilateral and symmetrical localization⁶. Seventy patients diagnosed with this condition which may be sporadic or familial have been reported in the literature up to the present time and the majority of these patients are female⁷⁻⁹. In the clinical series reported by Rongioletti et al.¹⁰ in 2012, nine of twelve patients were female. In the series composed of three patients reported by Yan et al.² in 2001, all patients were female. In our study, male predominance was found in contrast to the literature. ASA is frequently located in the palmar region. Up to the present time, few cases with dorsal localization have been reported. One of these cases is the case with localization in the hand and dorsum of finger reported by Aksoy and Hapa³ from our country. In accordance with this literature, localization in the dorsum of hand and fingertips was observed in our three patients. Histopathologically, orthokeratotic hyperkeratosis, akantosis, marked eccrine ducti, dilated acrosyringium, focal spongiosis around the acrosyringium, eccrine sweat gland hyperplasia, changes in the eccrine glandular cells and papillary dermal perivascular lymphocytic infiltration are frequently observed. Histopathologic findings have been found to be nonspecific in 20% of the cases in which diagnostic biopsy was performed³. Histopathology is not necessary for the diagnosis. Diagnostic biopsy was performed in two of our cases and spongiotic changes in the stratum corneum, orthohyperkeratosis accompanied by acanthosis and dilatation in the eccrine acrosyringium were found histopathologically.

Table 1. Clinical and demographic properties of the patients

Patient number	Age	Gender	Disease period	Localization	Comorbidity	Accompanying symptom	Permanence	Biopsy	Treatment
1	45	M	6 months	Palmar region, lateral parts of the fingers and dorsal part of the hand, symmetrical	-	Burning, mild hyperhydrosis	Transient	Spongiotic changes in the stratum corneum, orthohyperkeratosis accompanied by acanthosis and dilatation in the eccrine acrosyringium	20% aluminium chloride
2	35	M	7 years	Palmar region, fingertips, symmetrical	-	Burning	Transient	-	20% aluminium chloride
3	26	M	14 years	The dorsal part of the fingers, symmetrical	-	Mild hyperhydrosis	Transient	-	20% aluminium chloride
4	29	M	2,5 months	Palmar region, symmetrical	Alopecia areata	Mild hyperhydrosis	Transient	Spongiotic changes in the stratum corneum, orthohyperkeratosis accompanied by acanthosis and dilatation in the eccrine acrosyringium	20% aluminium chloride
5	31	M	6 months	The dorsal part of the hand and volar part of the wrist, symmetrical	-	-	Transient	-	20% aluminium chloride
6	24	M	4 months	Palmoplantar region, symmetrical	-	Burning	Transient	-	20% aluminium chloride

M: Male

It has been proposed that this condition with unknown etiology may be related with genetic predisposition or conditions including cystic fibrosis, focal hyperhidrosis and Raynaud phenomenon^{7,11,12}. Mild hyperhidrosis was present in three of the subjects (50%) included in our study (Table 1). In the literature, hyperhidrosis accompanying ASA with palmar localization was found in a 28-year old male patient in the case series of Pastor et al.⁹ and in a 24-year old male patient reported by Baldwin et al.⁵ in 2006. Increase in the capacity of epidermal keratinocytes to bind water as a result of increased epidermal sodium concentration has been blamed in the pathogenesis of this condition⁷. It has been thought that drugs including rofekoksib and selekoksib leads to development of ASA by increasing the concentration of electrolytes in sweat by way of inhibition of coxygenase 2 enzyme with a similar mechanism^{7,11,13,14}.

In our study, there was no history of accompanying morbidity and use of medication except for alopecia areate found in on patient.

There is no efficient option in treatment. The most commonly recommended treatment option among the ones preferred in the literature is application of topical aluminium salts³. The other treatment



Figure 1. White keratoderma plaques and maceration in the palmar region



Figure 2. Desquamation and white keratoderma plaques in the periungual regions

options which are found to be efficient and are being applied include antihistaminic drugs, botulinum toxin, 5% salicylic acid and urea^{3,15}. We applied topical 20% aluminium chloride treatment in our patients. Complete response was observed in five (83.3%) of the patients in two weeks and partial response was observed in one patient (16.7%). Recurrence was not observed in the follow-up. Here, male predominance of this rare form of keratoderma has been reported in contrast to the literature which describes female predominance and the demographic properties have been reviewed.

Ethics

Informed Consent: Informed consent was obtained from all the patients included in our study.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Nebahat Demet Akpolat, Fadime Kılınc, **Concept:** Nebahat Demet Akpolat, Fadime Kılınc, **Design:** Nebahat Demet Akpolat, Fadime Kılınc, **Data Collection or Processing:** Nebahat Demet Akpolat, Fadime Kılınc, Ayşe Akbaş, Ahmet Metin, **Analysis or Interpretation:** Nebahat Demet Akpolat, Fadime Kılınc, **Literature Search:** Nebahat Demet Akpolat, Fadime Kılınc, **Writing:** Nebahat Demet Akpolat.

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