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Case Study

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IDIOPATHIC AQUAGENIC SYRINGEAL KERATODERMA - A RARE CASE PRESENTATION

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ABSTRACT

Aquagenic keratoderma (AK) is a rare acquired skin condition characterized by recurrent and transient white papules and plaques associated with a burning sensation, pain, pruritus and/or hyperhidrosis on the palms and more rarely, soles triggered by sweat or contact with water. We present the case of a 14-year-old girl with idiopathic aquagenic keratoderma.

KEYWORDS: Aquagenic keratoderma (AK).

INTRODUCTION

In 1996, English and McCollough[1] described a new entity in 2 sisters characterized by a transient and recurrent keratoderma exclusively on the palms after brief water exposure. The condition occurred 3–5 min after exposure to water and resolved within a short time after drying. English and McCollough chose the term transient reactive papulotranslucent acrokeratoderma because they likened the appearance of these lesions to those seen in hereditary papulotranslucent acrokeratoderma (HPA). First described by Onwukwe, Mihm, and Toda, et al.[2] Yan et al. have documented 3 female patients with the same condition, and they coined the term 'aquagenic palmoplantar keratoderma' because the earlier term of 'transient reactive papulotranslucent acrokeratoderma' suggested an inherited keratoderma which might be misleading.[3]

Aquagenic keratoderma (AK), also known as aquagenic palmoplantar keratoderma, aquagenic syringeal keratoderma, aquagenic wrinkling of the palms and aquagenic acrokeratoderma, is a rare condition presenting with recurrent and transient white papules and plaques on the palms and more rarely, soles triggered by sweat or contact with water. AK is predominant in women with a mean age of onset of 21 years. Its etiology is unknown, although about one‐third of cases are associated with cystic fibrosis (CF) and some instances are induced by intake of various drugs, including aspirin, indomethacin, salazopyrin, rofecoxib and celecoxib.[4-6]

CASE REPORT

A 14-year-old girl presented with complaints of roughness dryness whitening and haedening of her hands on the palmar surface within 10 minutes of exposure to water. These changes to her hand associated with mild burning and sensation to itch .Drying of her hands leads to resolution of the symptoms within 60 minutes. The feet were unaffected.

The manifestations were transient and resolved within 30‐60 min after drying his hands. No other family members presented similar findings. Patient also reported a history of hyperhidrosis, while denied relevant personal pathological antecedents or history of drug intake. Physical examination revealed a normal palmar skin surface. After immersing the hands in water at 15°C for 5 min, small white pebble‐like papules with prominent eccrine ostia were evident [Figure 1a and b] and the patient reported a burning sensation. The lesions and symptoms normalized within 40 min after drying (Fig 1). Small white pebble‐like papules with prominent eccrine ostia after immersing the hands in water at 15°C for 5 min. Magnification of the papules after immersion. We prescribe her 20% aluminium chloride for night time application after completely drying her palms.

After 2 month the patient return for followup and reported near complete resolution of white papule, plaque and associated burning and itching .she was advised to slowly taper off the application until she would be able to discontinue the drug.

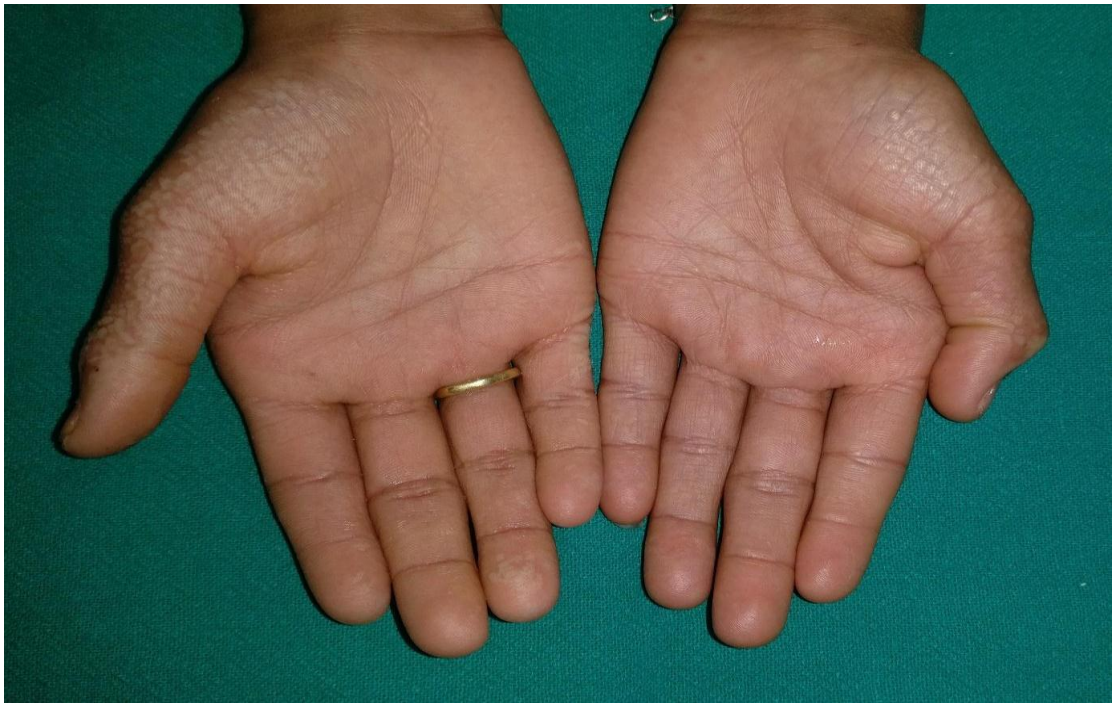

Fig 1: multiple white pebble like papules and plaque.

DISCUSSION

AK continues to be also a focus of research as to its genetic predisposition and association with other diseases including CF, focal hyperhidrosis, and Raynaud phenomenon.[7,8] The diagnosis of AK is mainly clinical and the ”hand‐in‐the‐bucket “sign, when patients submerge their hands in water for a time ranging from 2 to 10 min in order to demonstrate the lesions, is an important diagnostic clue.[9] The main differential diagnosis is hereditary papulotranslucent acrokeratoderma, which appears during puberty with persistent, asymptomatic, yellow‐white, translucent palmoplantar papules and plaques that are unrelated to water exposure.[10] The pathogenesis of AK is still unknown; despite multiple hypotheses have been proposed including eccrine gland or nerve dysfunction, hyperhidrosis, defective stratum corneum barrier function, occlusion of the eccrine duct ostia, or weakness of the eccrine duct wall.[11 12] AK continues to be also a focus of research as to its genetic predisposition and association with other diseases including CF, focal hyperhidrosis, and Raynaud phenomenon.[13,14] Regarding CF, it is estimated that between 44% and 80% of patients with CF have AK.[15,16] That is why, this disease served as a model to study the genetic association and the underlying pathogenic mechanisms of AK. In fact, the cause of development of these associated disorders is a homo‐or heterozygous mutation for ΔF508, which was first discovered in the CFTR gene in CF.[17,18] AQPs may also be involved since they regulate various functions of the skin. AQP3 is expressed by keratinocytes from the basal epidermal layer up to the spinous layer in human skin. AQP 10 is a water transporting AQP first localized in intestinal glycocalyx.[19] It is also expressed in human skin.[20] Because of its capability to open water channels rapidly in addition to carrier‐mediated transport, AQP 10 more than AQP3 would be a possible candidate for the abnormal reaction of palmar skin to water immersion. When expression of AQP3 is increased by all‐trans retinoic acid, none of the features of AK develop[21]

Conflict of interest
Nil

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nil

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