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Case Report

Aquagenic Syringeal Acrokeratoderma: Case Report

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Abstract: Aquagenic syringeal acrokeratoderma is a rare, acquired and transient type of keratoderma that may occur after a few minutes of exposure to water, usually occurs in adolescents and young female adults and it is characterized by the sudden appearance of translucent papules that converge on the palms and occasionally on the soles, macerated and with accentuation of the finger lines. We present the case of a 36-year-old female, with a history of long evolution of contact dermatitis. She presented a dermatosis characterized by maceration, roughed skin with wrinkling appearance and slight desquamation especially over thenar area after contact with water for a few minutes. After received general skin care, second generation antihistamine drugs, and emollients with 10% urea for a week, she reports improvement of the lesions. Aquagenic acrokeratoderma is a rare disease that can adversely affect the patient's life quality, however the etiology and pathogenesis are not clear; until date, there is no consensus on clinical treatment; for this reason it's important to identify the cases and report which treatments were effective, and achieve unifying the management of this pathology.

Keywords: Aquagenic, keratoderma, syringeal, dermatosis, dermatology, wáter.

INTRODUCTION

Aquagenic syringeal acrokeratoderma palmoplantaris, also known as aquagenic wrinkling, transient reactive papulotranslucent acrokeratoderma transient aquagenic palmar hyperwrinkling, is a rare acquired disorder, that predominantly affects young woman in a relation 2:1 with man; mostly localized on the palms. It is characterized by translucent papules in palms and soles, edematous plaques and keratoderma developing after a brief exposure to water and resolving shortly after stopping contact with water [1, 2].

CASE PRESENTATION

A 36-years-old female, no chronic degenerative diseases, with a history of long evolution of contact dermatitis, as well as allergies to pollen and dust, she presented with a 3 month history of dermatosis localized and symmetrical, characterized by maceration, with wrinkled appearance after contact with water for a few minutes, she also denied palmar hyperhidrosis, discomfort feeling or sensitivity loss when the lesions appeared; She received general skin care, second generation antihistamine drug, and emollients with 10% urea; after 1 week of treatment she express improvement of the lesions.

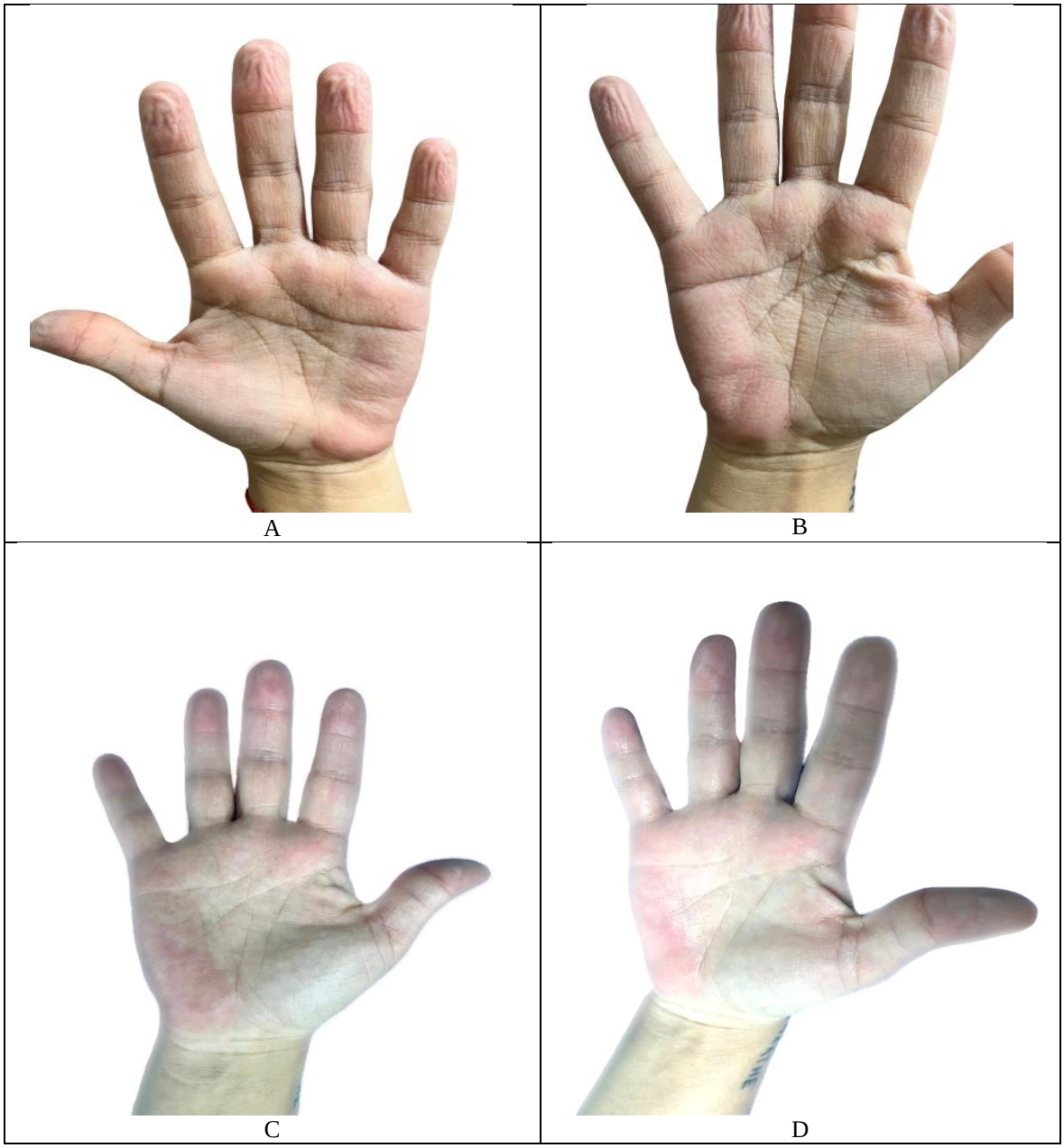

Figure 1: A) Left palmar hand which presents roughed skin with excessive wrinkling and slight desquamation especially over thenar area, after immersion of hands in water for 10 minutes. B) Right palmar hand with the same lesions. C & D) Right palmar hand before water exposure without lesions and after a week of treatment

DISCUSSION

Aquagenic palmoplantar keratoderma is a rare acquired aberrant function of sweat glands, patients develop skin thickening of palmar skin and the appearance of translucent papules after immersion in water [3, 4] usually presented bilaterally and symmetric; patient complain of tingling and pain in the hands, abnormal feeling after touching water, or only the local skin has a sense of tightness starting a few minutes after exposure to water and lasting for 20-30 minutes after removal [5, 6]. Several pathogenic mechanisms have been proposed, including structural or functional defects of the stratum corneum, increased sodium concentration in the skin, thereby increasing the water-retention capacity of the stratum corneum, abnormal regulation of transmembrane channels such as aquaporin 3 and a role of weakness of eccrine duct walls have also been considered in its etiology [6, 7]. The typical skin lesions of aquagenic acrokeratoderma are transient reactive skin keratosis papules caused by water stress, focal skin swelling with translucent white or yellow edematous papules or patches and whitish hyperkeratosis. The lesion can decrease or disappear soon after drying [1, 2, 6]. Histopathological changes include orthohyperkeratosis with increased thickness and abnormal staining of the stratum corneum, dilated acrosyringia, and dermal eccrine ducts with hyperplasia of eccrine glands, clear cell changes and vacuolations, and increased capillaries around and adjacent to the eccrine glands [8]. Its association with atopic dermatitis, Raynaud's

phenomenon, focal hyperhidrosis, and use of cyclooxygenase enzyme (COX)-2 inhibitors (aspirin, rofecoxib) remains anecdotal [9]. Cystic fibrosis, an autosomal recessive disease caused by mutations in cystic fibrosis transmembrane conductance regulator (CFTR) gene, remains the most described association [8, 9]. Improvement of the aquagenic syringeal acrokeratoderma lesions with sweat gland-suppressing treatment options include topical aluminum chloride, 12% aluminum lactate cream, 3% formalin in alcohol, 5% salicylic acid ointment, petroleum-based barrier creams and botulinum toxin injection have been described, all of them showed mixed and transient result; Sezer *et al*., proposed permanent treatment with endoscopic thoracic sympathectomy in patients with with aquagenic syringeal acrokeratoderma, especially if it is associated with severe palmar hyperhidrosis, however more studies are needed [7]. Nevertheless the etiology and pathogenesis are not clear, thus efficacy of related treatment methods are different, until date, there is no consensus on clinical treatment [6].

CONCLUSION

Aquagenic acrokeratoderma is a rare disease that can adversely affect the patient's life quality. The etiology, pathogenesis, and corresponding treatment of aquagenic acrokeratoderma is worthy of further research and exploration.

Conflict of Interest: The authors declare that there are no conflicts of interest at the time of publication of this article.

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