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IMAGES IN PAEDIATRICS

Can I touch water if I have cystic fibrosis?

Tengo fibrosis quística: ¿no puedo tocar el agua?

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A girl aged 4 years with cystic fibrosis (CF, heterozygous F508del/G542X genotype) was brought in for a checkup and it was reported that she developed papular whitish lesions in palms and soles upon contact with water associated with hyperwrinkling (Figs. 1 and 2) and associated pain if she stayed immersed in water for a long time. The symptoms disappeared after drying.

The patient was referred to the dermatology department, where she received a diagnosis of aquagenic keratoderma[1,2] (AK) and prescribed treatment with emollient creams,[2,3] which did not alleviate the symptoms.

At age 6 years, she started treatment of CF with elexacaftor-tezacaftor-ivacaftor. After 3 months of treatment, the family reported complete resolution of AK (Fig. 3).

Aquagenic keratosis is a little-known cutaneous disease, but its association with CF has been described extensively,[2,3] and it has been hypothesised that sweat hypertonicity could lead to to a higher ratio of flux from eccrine glands in the palms.[2] It is a clinical diagnosis (whitish papules with a central depression that appear after contact with water, possibly in association with pain[1]), and the treatment is symptomatic. Although it is a benign disease, it can have an impact on the patient's quality of life. Therefore, the use of novel CF conductance regulator modulators could be the answer for the treatment of AK.

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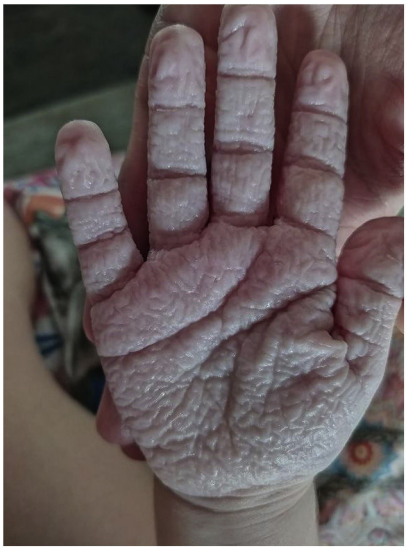

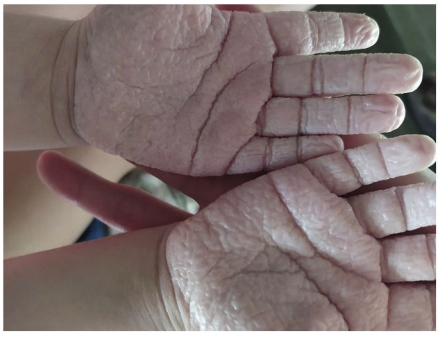

Figures 1 and 2 Whitish lesions in the palms of both hands and hyperwrinkling developed during bathing.

Figure 3 Resolution of lesions after treatment with elexacaftor-tezacaftor-ivacaftor.

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