Aquagenic Syringeal Acrokeratoderma

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STo the Editor:
Aquagenic syringeal acrokeratoderma, also known as aquagenic palmoplantar keratoderma, is an acquired keratoderma of unknown etiology. Several pathogenic mechanisms have been proposed, including structural or functional defects of the horny layer during adolescence, primary disease of the sweat ducts, increased sodium concentration in the skin thereby increasing the water retention capacity of the horny layer, or a reaction to drugs.1 Since it was first described in 1996 by English and McCollough,18 cases have been published in the literature, 4 of them in Actas Dermosifiliográficas.10-12

We recently diagnosed a new case of this entity in a healthy 28-year-old man who, since childhood, had been complaining of a burning sensation and growing tightness in the palms after only a few minutes of contact with water. The process had progressively worsened, to the point that daily showering was difficult and even dysesthesias were present. There were no concomitant drugs, associated hyperhidrosis, or relevant family history. The disease had been treated for years with various topical preparations usually prescribed for chronic irritative dermatitis or atopic eczema, with no improvement of any kind.

The dermatological examination initially showed mild bilateral palmar hyperkeratosis. After submerging the hands in water for 3 minutes, the palm skin acquired a thickened, crushed, whitish appearance, with accentuation of palm wrinkles and formation of small confluent translucent papules that showed dilated sweat duct openings (Figure 1). The symptoms gradually improved to baseline status within 30 to 45 minutes.

Based on clinical symptoms resembling aquagenic syringeal acrokeratoderma, we performed a histological study that revealed mild hyperkeratosis and discrete dilatation of the acrosyringium, of unclear pathological significance (Figure 2). Because the symptoms were similar to the alterations described at the palmar-plantar level in patients with cystic fibrosis,13 we performed a sweat test, which was normal.

The patient was started on topical treatment with an 18% aluminum hydrochloride water-alcohol solution but showed no significant improvement. He declined to participate in a therapeutic trial with botulinic toxin.14

We describe this case study in order to extend the body of literature on this new entity in Spain and express our opinion that this disease may have been underdiagnosed to date.

References

To the Editor:

We have read with interest the article by Dr Sánchez-Castellanos, Dr Sandoval-Tress, and Dr Henández-Torres entitled, “Persistence of the Omphalomesenteric Duct. Childhood Differential Diagnosis of Umbilical Granuloma” and would like to make a few comments. Although there have been reports of clinical differences between the various neonatal umbilical nodules (umbilical granuloma, persistence of omphalomesenteric duct, urachus remnants, etc) in the past, these were not based on confirmed diagnoses, but rather on the response or lack thereof to a destructive treatment. Making a diagnosis on whether or not the lesion is destroyed by chemical cauterization with silver nitrate is not a very scientific approach.

We agree with the authors’ conclusion that any newborn with an umbilical neoplasm should undergo a study of the lesion to confirm the diagnosis, thus making appropriate treatment possible to avoid potential complications. A greater understanding of umbilical nodules in childhood and their complications would require clinical and epidemiological studies based on histopathological diagnoses. In 2005 we described a simple method for diagnosis by anatomical pathology from biopsies taken by presutured purse-string suture of neonatal umbilical nodules. The painless application of a purse-string suture at the base of the umbilical nodule, a knot to ensure that the lesion was exsanguinated, and immediate removal by cutting with scissors above the suture line allowed histological study of 75% to 90% of the lesion. The presuturing technique with immediate excision for biopsy is minimally traumatic and takes less than a minute. The cosmetic outcome is excellent and we believe the technique is of interest in dermatological surgery.

References


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Reply

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To the Editor:

First of all, we would like to express our appreciation for the comment made by Dr Juan Luis Artola Igarza and coworkers regarding our article entitled, “Persistence of Omphalomesenteric Duct. Childhood Differential Diagnosis of Umbilical Granuloma.”

Regarding the biopsy by presutured purse-string suture that the authors of this comment use for the diagnosis and treatment of umbilical polyps, we should mention that we were not aware of this technique and have not used it. However, after reading the article we feel it is a good option for the diagnosis of umbilical polyps and we hope to be able to use it when our patients present with such a condition in the future.

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