LETTER TO THE EDITOR

[Translated article] Comment on ‘‘Darier Disease: A Case Series of 20 Patients and Review of the Literature’’

Comentario al artículo ‘‘Enfermedad de Darier: serie de 20 casos y revisión de la literatura’’

To the Editor:

I have read the report in the Case and Research Letters section of the July–August issue on a retrospective study between 2008 and 2019 of 20 cases of Darier disease, described as the first Spanish case series. In 1998, we published in this journal – the results of a clinical-genetic study in Spain of 3 families (not interrelated) with a total of 70 members, of whom 25 were affected by Darier disease (13 males and 12 females), with ages ranging from 12 to 71 years at the time of the study and onset ranging from 9 and 20 years. In that article, we reported on the type and distribution of the cutaneous manifestations observed, similar to those very well described in the recent article. We also described other clinical features of the disease already reported in other studies: papular lesions in the oral mucosa of the palate resulting in cobblestone plaques; dental abnormalities including gaps, notching on the free edge of the incisors, and conical canines; nail changes including onycholysis and white or red longitudinal bands on the nail plate with nicking on the distal end. Palmoplantar keratoderma with characteristic pitting – present in 12 of our patients together with nail alterations – has been reported to be an early manifestation of the disease.

Several studies have reported on the importance of the neurological and psychiatric disorders associated with Darier disease. We observed these disorders in 15.3% of our patients in the form of multiple sclerosis, epilepsy and depression as well as suicidal ideation and behavior. We also noted the docile attitude and social and emotional withdrawal found in young and middle-aged patients with this disease, which had a negative effect on their social relationships.

An association with bone disease, a controversial issue in this setting, was observed in 2 of our patients from the same family, both of whom had very marked cutaneous expression of the disease in the form of bone cysts in the upper extremities and jaw.

References


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