Red, white and blues: Darier disease and mood disorder

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DESCRIPTION

A middle-aged woman presented with a progressively worsening rash for 25 years. The rash was foul smelling and tender, occurring symmetrically in the folds of her abdomen and lower legs. Her medical history was significant for major depressive disorder. She had been receiving care from multiple psychiatrists over the last 10 years, and she has been hospitalised in inpatient psychiatric facilities for acute psychotic episodes. She admitted to having two of five siblings with similar skin problems, both additionally having been diagnosed with mood disorders including bipolar disorder and major depressive disorder. The patient stated the other three siblings were healthy. On examination, the patient had plaques of confluent papules and foul-smelling, weeping patches which were distributed over the folds of her chest and abdomen, as well as the flexure surfaces of her bilateral lower extremities (figure 1). Examination of her nails revealed red and white longitudinal striations and v-shaped nicking (figure 2). Two punch biopsies were performed in the involved skin which demonstrated acantholytic dyskeratoses. The patient was accordingly diagnosed with Darier disease (DD) and treated with appropriate wound care, topical corticosteroids and doxycycline with an intention to begin topical retinoids once the affected skin had epithelialised. The patient was also advised to continue following up regularly with her mental health specialists.

This patient and her family compellingly showcase a genetic inheritance of both mood disorder and skin disease in DD. In a previous study, 100 unrelated individuals with DD were assessed for using standardised neuropsychiatric measures and the findings concluded that neuropsychiatric symptoms in DD are not simply a psychological reaction to having a skin disease but are consistent with the pleiotropy hypothesis that mutations in the ATP2A2 gene, in addition to causing DD, confer susceptibility to neuropsychiatric features.[1] It is critical for clinicians to understand these ATP2A2 genotype-phenotypes in order to appropriately address both the neuropsychiatric and cutaneous diseases in patients with DD.

Figure 1 Popliteal fossa of left lower extremity showing a severely denuded and weeping patch.

Figure 2 Fingernails demonstrating red and white longitudinal striations with v-shaped nicking.

Learning points

- Individuals with Darier disease (DD) often have both neuropsychiatric disease and skin disease.
- The co-occurrence of neuropsychiatric disease and skin disease in individuals with DD is likely due to pleiotropy.
- Appreciation for the variety of ATP2A2 genotype-phenotypes that exist in DD may help clinicians to address both the neuropsychiatric and cutaneous diseases in patients with DD.

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1 Gordon-Smith K, Jones LA, Burge SM, *et al*. The neuropsychiatric phenotype in Darier disease. *Br J Dermatol* 2010;163:515–22.

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