GENETICS AND GENODERMATOSES

DARIER DISEASE IN TEENAGE FEMALE PATIENT: A CASE REPORT

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Background: Darier disease (DD) is an autosomal dominant genodermatosis which is often under - or misdiagnosed. In approximately 70 % of patients, the disease begins between the ages of six and 20 years, and peak during the teenage years. Both sexes are equally affected. DD presents with keratotic, sometimes crusted, red to brown papules and plaques which favor seborrheic areas such as the trunk, scalp margins, face, presternal, interscapular areas and lateral aspects of the neck.

Observation: A 16 year-old girl without comorbidities and no history of any previous skin disorder presented to dermatology department with a two years history of bilateral simetrical rash on lateral sides of neck and presternal area and also the frontal hairline. The patient was treated in the past with topical antimicotic and antibiotic creams several times without any significant improvement. No nail or oral mucosa involvement were established. She complained of moderate itch and worsening after sweating and sun exposure. Results of a biopsies of lateral side of neck lesions and trunk lesions showed foci of suprabasilar acantholysis and dyskeratosis including both corps ronds and grains, while result of direct immunofluorescence was negative. Typically we started treating patient with adaplen 0.1 % cream once daily every 3 days and slowly increasing the frequency of application.

Key message: The diagnosis of DD is suggested by the clinical finding of a persistent eruption of keratotic papule involving the seborrheic areas. A skin biopsy for histologic examination is needed to confirm the diagnosis and exclude other conditions with similar clinical findings. There is no cure for DD. The goals of treatment are the improvement of the skin appearance, relief of symptoms and prevention or treatment of infectious complications.