GENETICS AND GENODERMATOSES

VARIABLE RESPONSE TO NALTREXONE IN PATIENTS WITH THERAPY REFRACTORY DARIER DISEASE

D Boehmer [1] - K Eyerich [1] - U Darsow [1] - A Zink [1] - T Biedermann [1]

Technical University Of Munich, Dermatologie Am Biederstein, Munich, Germany [1]

Introduction: Darier disease is a rare autosomal-dominant genodermatosis (mutation ATPA2A-gene), characterized by red-brown keratotic papules mainly in seborrhoic areas, which have the tendency to coalesce and form large, macerated, warty areas. Severity varies markedly amongst individuals. Treatment of Darier disease is often frustrating and limited.

Objective: The objective of the study was to figure out, whether a systemic therapy with low-dose Naltrexone, can present a new therapy option for Darier disease.
Recent publications had documented a good response of Hailey-Hailey-disease to Naltrexone. As Darier disease also has a genetic mutation coding for a Calcium-pump, we decided to initiate this therapy in patients with therapy refractory Darier disease.

Materials & Methods: Six patients (4 patients with a severe clinical presentation, 2 patients with a mild clinical presentation) with biopsy-proven Darier disease were initiated on an off-label therapy with Naltrexone 5mg and Magnesium 200mg. They were followed up four weekly. Upon clinical presentation the clinical response, subjective pain and itch scores were assessed.

Results: Results showed that the four patients with a severe form of Darier disease initially improved and then worsened dramatically. The two other patients with a mild clinical presentation improved dramatically and nearly showed a complete remission. The keratotic papules flattened completely.

Conclusion: In summary, results were disappointing, as the patients with the moderate to severe forms initially improved and then dramatically worsened. According to our experience, Naltrexone is currently not a valid therapy option for patients with moderate to severe Darier disease. The patients with a very mild form did show a very good response. To confirm this, additional studies are needed.