GENETICS AND GENODERMATOSES

AN UNUSUAL CAUSE OF PERINEAL INTERTRIGO!!!

K Belhareth [(1)] - M Korbi [(1)] - M Lahouel [(1)] - Y Soua [(1)] - H Belhajali [(1)] - M Youssef [(1)] - J Zili [(1)]

University Hospital Of Monastir, Fattouma Bourguiba Hospital, Department Of Dermatology, Monastir, Tunisia [(1)]

Background: Acrodermatitis enteropathica (AE) is a rare disease affecting the uptake of zinc. It occurs generally during the first months of life after breastfeeding withdrawal. We report a case of a perineal intertrigo revealing an AE in a child of 6 years.

Observation: A6 year-oldchild, first child of a non-consanguineous marriage, was referred to our department for perianal lesions evolving for 9 months and resistant to various symptomatic treatments. On dermatological examination, he had perianal erythematous lesions with an erosive intertrigo. There was no mucosal involvement. The rest of the clinical examination was unremarkable. Laboratory evaluation revealed low plasma zinc level. There was no immunodeficiency. Zinc supplementation at a dose of 15 mg/day was followed by complete healing of the lesions within 3 weeks. The evolution was marked by a clinical relapse as soon as the treatment was stopped. The reintroduction of zincled to rapid healing of the lesions. The diagnosis of AE was made. Thegenetic report is not available yet.

Key messages: AE is a rare autosomal recessive disorder. The responsible gene is on chromosome 8q24.3. The clinical presentation of AE is very characteristic with symmetrically distributed erythematous squamous, sometimes vesiculobullous or pustular lesions, around perioral, anogenital, and acral areas.Other clinical symptoms were described, including: growth retardation, alopecia and recurrent infections. Mucous lesions may include gingivitis, stomatitis and glossitis. To the best of our knowledge, an erosive chronic perianal intertrigo has not been described in association with AE. The diagnosis is based on clinical aspect and is confirmed by low plasma zinc levels and a good response to zinc supplementation. Our observation is original by not only the late onset of the disease but also by the confusing clinical presentation with a resistant perineal intertrigo.