AUTOIMMUNE BULLOUS DISEASES

LINEAR IGA BULLOUS DERMATOSIS:A CLINICOPATHOLOGIC REVIEW IN A SERIES OF 38 PATIENTS FROM MILAN

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Background: Linear immunoglobulin A (IgA) bullous dermatosis (LABD) is an autoimmune subepithelial vesiculobullous disease due to IgA autoantibodies directed against different antigens of the basement membrane zone of the skin and/or mucosae. It has a biphasic course affecting young children and adults after their 5th decade. Although drug-induced LABD has been recently regarded as a separate entity, its clinical and immunohistopathological features are highly heterogeneous and completely indistinguishable from the idiopathic form. Literature is lacking retrospective studies on LABD from Italy.

Objective: To characterize the clinical presentation, pathological features, management, and course of an Italian cohort of LABD patients.

Methods: A retrospective review of 38 patients with LABD at our department from November 2006 to September 2018, was performed.

Results: Of 38 LABD patients, 11 were children. Mean age at diagnosis was 45.7 years. The overall male to female ratio was 1.2, while it was 2.7 in the paediatric population. Limbs were the most common site of skin lesions (68.4%), followed by trunk (55.3%), head (36.8%) and buttocks (13.2%). Perioral and genital involvement were prevalent in children. Mucosal involvement was seen in 9 (23.7%) patients and the most frequent mucosal localisation was oral cavity (15.8 %). Most cases were idiopathic (94.7%) and only two cases were regarded as probably drug-related. Linear IgA deposits along the BMZ were observed in 29 patients (76.3%), while linear/granular IgA deposits were observed in 7 patients (18.4%). Dapsone was the most commonly used drug and complete response was achieved in most cases (76.3%).

Conclusions: Our epidemiological and clinical findings relative to an Italian cohort of patients are mostly consistent with data present in the literature. As already reported in

previous studies, we confirm that very few cases may be regarded as drug-induced LABD.