Cutaneous Humoral Immunity: Vasculitis and Autoimmune Bullous Dermatoses

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acantholytic diseases like Hailey-Haileys, Grovers, Dariers disease in which the DIF is negative.

Pemphigus foliaceus: The bullae are due to a superficial intra-epidermal cleavage. Commonly, there is no mucosal lesion. The target antigen is DSG-1. It is less present than DSG-3 in mucosae. That is an explanation why the mucosae are usually normal in this type of pemphigus.

IgA pemphigus: Clinically IgA pemphigus is characterized by pruritic vesiculopustular eruptions. Histopathology is useful in differentiating the two major subtypes of IgA pemphigus [12]. In the Subcorneal Pustular Dermatosis (SPD) subtype, there is an increased intensity of IgA autoantibodies in the upper surface of the epidermis. In contrast, in the Intra Epidermal Neutrophilic (IEN) type, the IgA antibodies are located either in the entire or in the lower part of the epidermis. The target antigen type of the SPD subtype is desmocollin-1, which is important for the differential diagnosis with Sneddon-Wilkinson sub-corneal pustulosis in which the DIF is negative. The IEN type involves IgA antibodies directed against desmogleins 1 and 3. IgA pemphigus has been associated with monoclonal IgA gammopathy and multiple myeloma. Other associated diseases include human Immunodeficiency Virus (HIV) infection, Sjogren disease, rheumatoid arthritis, and Crohn's disease.

Drug-induced pemphigus: The lesions are polymorphous in this type of pemphigus. Clinically and histologically, they may be lichenoid or erythema multiforme. The etiologic investigation may find that some cases are drug-Induced. Because of the atypical presentation of the lesions, it may be very difficult to establish the diagnosis of pemphigus. If firstly negative, the DIF must be repeated.

Para Neoplastic Pemphigus: Para Neoplastic Pemphigus (PNP) is a rare muco-cutaneous autoimmune disease associated with neoplasm. Blisters and interface dermatitis sometimes co-appear in the same lesion. In DIF of the muco-cutaneous lesions, IgG autoantibodies and/or complement deposition is observed in the epidermal

is deeper than that of pemphigoid. Clinically, bubbles occur in middle-aged adults, in areas exposed to trauma (feet, hands, buttocks) and in the mucous membranes. The DIF shows a linear IgG deposit at the DEJ. The autoantibody targets the NC1 domain of collagen VII.

Linear IgA disease: In adults, lesions are easily annular or in cockroaches, the newer bubbles have a circinate arrangement around the older lesions [15]. Some forms can manifest as detachments simulating toxic epidermal necrolysis or Lyell syndrome, with mucosal lesions. The differential diagnosis is all the more difficult as drugs are incriminated in linear IgA dermatosis, especially vancomycin. Histologically, a sub-epidermal bulla with an intact roof is observed. The superficial dermis contains numerous neutrophils. The DIF shows a linear IgA deposit at the DEJ.

Dermatitis Herpetiformis (DH): DH preferentially affects young adults and children. DH is associated to coeliac disease in 15%-25% of the patients [16]. Clinically the lesions are herpetic-like small grouped vesicles that are symmetrically arranged. The lesions are preferentially located on the trunk and the bottom. The histological lesions are intra-papillary micro-abscesses located in some dermal papillae. Discrete lesions of leucocytoclastic vasculitis may occur. The DIF shows IgA granular and/or fibrillar IgA and C3 deposits in the dermal papillae, and inconstant IgA deposits in vessels. In the blood, circulating immune complexes may be present.

Associations of humoral immune pathologies

Schematically, the dermatoses related to humoral immunity can be divided into 2 groups of pathologies: Those due to vascular deposits of circulating immune complexes and those due to autoimmune antibodies. In the first group, the main lesions are cutaneous vasculitis. The second group is dominated by Autoimmune Bullous Dermatoses. Nevertheless both types of humoral immunity may be combined, especially in systemic diseases.

Discussion and Conclusion

For instance, patients with lupus erythematosus may have lesions of vasculitis and lesions of AIBD, like linear IgA bullous dermatosis or acquisita bullous epidermolysa. Regarding their cause, infectious and non-infectious agents, environmental agents and drugs, may be invoked as a possible cause of vasculitis as well as autoimmune diseases. For instance, pemphigoid can be observed after scabiosis, vancomycin is a possible cause of linear IgA bullous dermatosis; D-penicillamine can induce lesions of drug-induced pemphigus. Both circulating immune complexes and autoimmune antibodies are possibly found in the same disease: For instance, patients with Dermatitis Herpetiformis may have in the same cutaneous sample granular deposits in the capillaries and fibrillar anti-fibronectin antibodies in the dermal papillae.

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