COEXISTENCE OF BULLOUS PEMPHIGOID & HYPERTHYROIDISM: A CASE REPORT

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INTRODUCTION

Bullous pemphigoid (BP) is one of the most common autoimmune bullous diseases, although its frequency is less than pemphigus vulgaris (0.0004/100,000 vs 0.1-0.5/100,000 population [1]. In recent prospective studies from Germany and Switzerland, the annual incidence was about 12 to 13 new cases per 1 million population [2, 3]. It is primarily a disease of the elderly with an incidence among patients aged over 90 years being 40 per 100 000 population per year.

The association of bullous pemphigoid with many other diseases has been mentioned before, but no common etio-pathogenic pathway has been demonstrated [4 5, 6]. The most frequent associations are those with primary biliary cirrhosis, psoriasis - 63.4%), and an unusual condition termed multiple autoimmune syndrome (MAS), defined as the combination of at least three autoimmune diseases in the same patient . With multiple autoimmune syndrome, we observe a special frequency of various dermatological autoimmune diseases, such as vitiligo, bullous pemphigoid, and pemphigus vulgaris.

CASE SYNOPSIS

An 89-year-old Filipino man was seen in April 2017 at the Out- Patient Department Clinic of World Citi Medical Center because of a multiple skin blisters. His condition started 2 weeks prior to consult when he noted appearance of multiple, pruritic, tense, vesicles and bullae initially on both legs that gradually spread to the thighs and arms (Fig.1 A & B). He did not complain of fever, difficulty of breathing, abdominal pain or body malaise. His vital signs were within normal range. Examination of the skin showed dry, cool & mottled with multiple vesicles to tense bullae, some erythematous erosions over the arms, axilla, inner thighs, and legs. Nikolsky and Asboe Hansen signs were negative. Mucosal and genital areas were free of lesions. Hematologic tests showed normal CBC (WBC 7 x 10'/ul, Hemoglobin 121 g/L, Hematocrit 0.36, Platelet 210). Serum chemistries are within the normal range. Chest Xray was unremarkable as well. A diagnosis of Bullous Pemphigoid was confirmed by the presence of subepidermal blisters with mild superficial & deep perivascular and interstitial lymphocytic infiltrate admixed with eosinophilia and rare neutrophils and solar elastosis on skin punch biopsy (Fig. 2) as well as linear deposition of IgG & C3 at the dermo- epidermal junction on Direct immunofluorescence (Fig. 3). He was then started on oral Prednisone & erythromycin then discharged improved. Initially, lesions were partially controlled until 3-weeks post discharge, he was readmitted for palpitations with recurrence of multiple revealed Hyperthyroidism. Anthyroid drug was then added to his medications. Presently, his skin lesions were completely controlled.

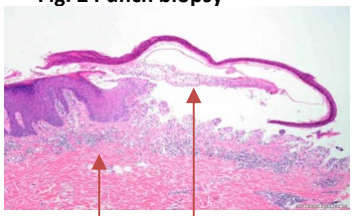


Fig. 1.A



Fig. 1B

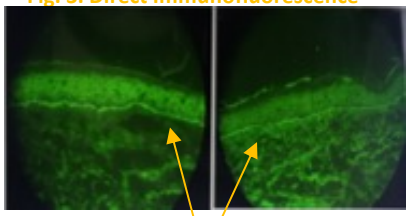
Fig. 2 Punch biopsy



Subepidermal blister with detached overlying epidermis

Perivascular infiltrates predominantly eosinophils

Fig. 3. Direct Immunofluorescence



Linear deposits at the basement membrane

CONCLUSIONS

We conclude that there exists an association of Bullous pemphigoid & Thyroid disease in our case being both autoimmune mediated. Based on Humbert and Dupond classification, they belong to two different types of class of Multiple Autoimmune syndrome (MAS). Although not typical of MAS, a common pathophysiologic mechanism can explain their co-existence. Both Autoimmune thyroid disease (AITD) and Bullous pemphigoid (BP) have autoantibodies belonging to IgG class. Importantly, patients with AITD are known to express HLA-B8 and the risk of its development is increased in patients with haplotypes HLA-DR3 or HLA-DQA 1*501 & HLA-DR4 while BP patients usually show HLA-DQB1, an HLA possibly associated with multiple diseases of the APS group

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