A case of bullous pemphigoid who achieved a long-term remission by a single course of high-dose intravenous immunoglobulin monotherapy

Running title: BP treated with a single course of IVIG

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Letter to the editor

The high-dose intravenous immunoglobulin (IVIG) is considered as an additional option in the treatment of steroid-resistant bullous pemphigoid (BP) (1, 2). However, to treat BP with corticosteroid is sometimes difficult due to complications. We report a case of BP with severe infectious complications who achieved a long-term remission by a single course of IVIG monotherapy.

A 76-year-old female presented with itchy erythema and blisters which had lasted for 4 weeks. Physical examination revealed edematous erythema with multiple coin-sized tense blisters and erosions all over the body (Figure 1A). Anti-BP180 antibody (aBP180ab) titer was high (2730 U/mL) and eosinophilia was observed (43.4 % of 8000/μL white blood cells). A skin biopsy demonstrated a subepidermal blister with eosinophilic infiltration (Figure 1B). Direct immunofluorescence showed the linear deposition of IgG and complement C3 along the dermal-epidermal junction (Figure 1C). On the basis of these findings, BP was diagnosed. Three weeks of topical mometasone furoate (5 g/d), oral tetracycline (200 mg/d) and nicotinamide (900 mg/d) were not sufficient for disease control. BP Disease Area Index (BPDAI) was 26 for erosions/blisters and 23 for urticaria/erythema. Although systemic corticosteroid was considered, the patient was suffering from septicemia and septic arthritis of the right knee. IVIG (400mg/kg for 5 days) was thus started along with 5 g/d of topical mometasone furoate. Then, the erythema and bullas significantly reduced in a week and the skin lesions almost cured in 8 weeks with the reduced aBP180ab titer (104 U/mL) and BPDAI (1 for erosions/blisters and 0 for urticaria/erythema). The topical

treatment was tapered off. The patient developed septic vertebritis and intervertebral discitis 10 weeks after IVIG. However, BP did not recur during the infectious disease. Since then, she has been in good condition without systemic treatments for 3 years.

There have been BP cases successfully treated with IVIG monotherapy. However, those cases were either treated with multiple courses or infantile cases (3, 4). Of note, the disease activity in our case was rapidly controlled by a single course of IVIG monotherapy and the remission lasted for 3 years without other treatment modalities. While the class 3 topical corticosteroid would also have helped the disease control, the contribution is assumed to be limited considering the strength and amount.

IVIG is regarded to be therapeutic by several functions such as saturating the IgG-protective receptor FcRn and accelerating IgG degradation, neutralizing pathogenic IgG, and negatively modulating cytokine profiles (5). The mechanism of long remission by IVIG in our case is not clear, but at least IVIG induced reduced production of aBP180ab from plasma cells, suggesting a long-term suppression of B cell activities. Bacterial infection may also have accelerated a short-term disease activity. However, since the disease activity of BP was stable during vertebritis, infection is not the sole factor for the development of BP.

In our BP case, a single course of IVIG monotherapy induced the rapid and long-term disease control. IVIG might serve as a good treatment option even as a monotherapy for BP patients with complications.

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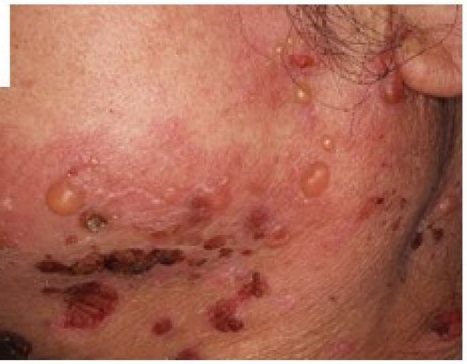
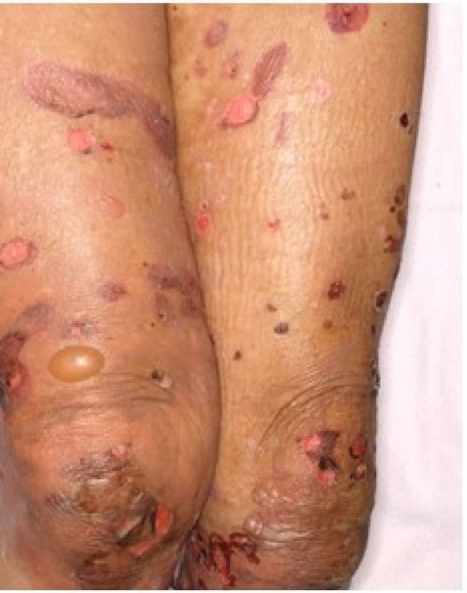
Figure legends

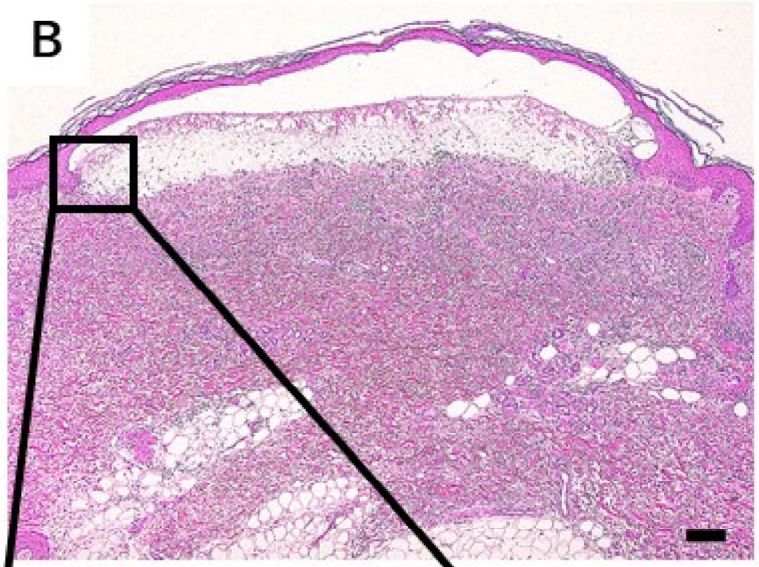
A. Edematous erythema, thumb nail-sized tense blisters and erosions on the face (top) and thighs (bottom).

B. A subepidermal blister (top) with the infiltration of eosinophils (bottom). H-E staining, the bar = 100 μm.

C. The linear deposit of C3 along the basement membrane zone by direct immunofluorescence.



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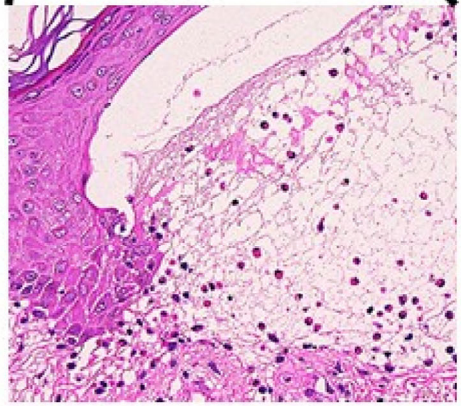

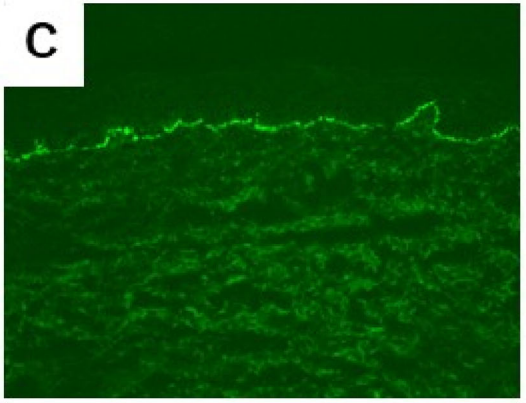
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