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AUTOIMMUNE BULLOUS DISEASES

## DUPILUMAB FOR THE TREATMENT OF RECALCITRANT BULLOUS PEMPHIGOID

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Background: Bullous pemphigoid (BP) is an autoimmune disorder that causes subepidermal bullae. It typically presents in the elderly, and has a wide spectrum of severity. We report a case of bullous pemphigoid successfully treated with dupilumab.

Observation: 83-year-old woman with a history of chronic lymphocytic lymphoma, severe congestive heart failure (CHF), and Alzheimer's dementia presented with 8 weeks of pruritic tense bullae on her trunk and extremities involving 40% of body surface area and uninvolved mucosae. She had not taken any medications for six months. Punch biopsies were performed on the trunk of lesional and perilesional skin. Histopathology revealed subepidermal bullae with eosinophils and neutrophils, and direct immunofluorescence revealed linear IgG reactivity along the dermal-epidermal junction. BP was diagnosed, and prednisone was begun. The patient failed three courses of prednisone taper over 8 weeks and bullae continued expanding. Laboratory testing revealed latent tuberculosis. Given comorbidities including CHF, malignancy, and infection, dupilumab was started at conventional dosing. Within 3 weeks, itch improved and no new bullae appeared. A complete resolution of disease was noted in 15 weeks.

Key Message: Treatment of recalcitrant bullous pemphigoid in patients with multiple medical co-morbidities is difficult. The most common initial treatment of choice for severe disease is oral glucocorticoids, but they can result in several adverse effects. Other options include mycophenolate mofetil, methotrexate, azathioprine, cyclosporine, cyclophosphamide, rituximab, dapsone, intravenous immunoglobulin, and doxycycline. These medications were contraindicated in our patient due to co-morbidities (CHF, infection, malignancy).

Patients with BP have increased circulating IL-4 and IL-13, increased Th2 cell activity, and increased IL-4 induced B-cell class switching. Dupilumab targets both chemokines and inhibits Th2 cells, and we believe the medication is thereby effective for this disease.

This case highlights the effective use of dupilumab for the treatment of BP, but large-scale, randomized studies are needed.





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