SKIN

BRIEF ARTICLE

Coexisting Psoriasis and Anti-P200 Pemphigoid: A Case Report

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ABSTRACT

Psoriasis and autoimmune bullous diseases have been shown to coexist in certain patients. Notably, anti-p200 pemphigoid has been associated with psoriasis. We present a case of a patient who presented with psoriasis and was also found to have anti-p200 pemphigoid. This case highlights the importance of recognizing this association, which may aid in diagnosis but also poses an interesting challenge in co-managing these two entities.

INTRODUCTION

Anti-p200 pemphigoid is a rare, subepithelial immunobullous disease that may be underdiagnosed. It has been reported to occur in patients who also suffer from psoriasis.[1] We discuss a case of a patient with both psoriasis and anti-p200 pemphigoid to emphasize the importance of considering anti-p200 pemphigoid as a diagnosis in patients with a history of psoriasis who present with a new onset bullous disease and to highlight various treatment modalities we found effective.

CASE REPORT

A 39-year-old male presented to the dermatology clinic with a few months' history of erythematous, scaling plaques over his face, torso, and upper and lower extremities. He had been seen in the emergency department previously for a similar rash, which was treated as psoriasis, but he had not followed with a dermatologist for this concern.

Further chart review revealed he had previously seen a dermatologist for a generalized bullous disease three years prior. At that time, he presented with bullae over his torso and bilateral upper and lower extremities as seen in **Figure 1**. Workup with direct immunofluorescence revealed linear staining of IgG and C3 in the basement membrane zone as seen in **Figure 2**, as well as IgG reactivity in a dermal pattern on human split skin on indirect immunofluorescence. Pemphigus panel, anti-type VII collagen antibody, IgA pemphigus panel, and anti-nuclear antibody were all negative. Given these findings, a diagnosis of anti-p200 pemphigoid was favored. The patient was treated with methylprednisolone, doxycycline, and dapsone, but was lost to follow-up.

Upon his presentation three years later, a physical exam revealed diffuse, erythematous plaques with fine scale over his face, torso, and extremities as seen in **Figure 3**. There were no active bullae. A biopsy was performed and revealed findings consistent with psoriasis. The diagnosis of coexisting

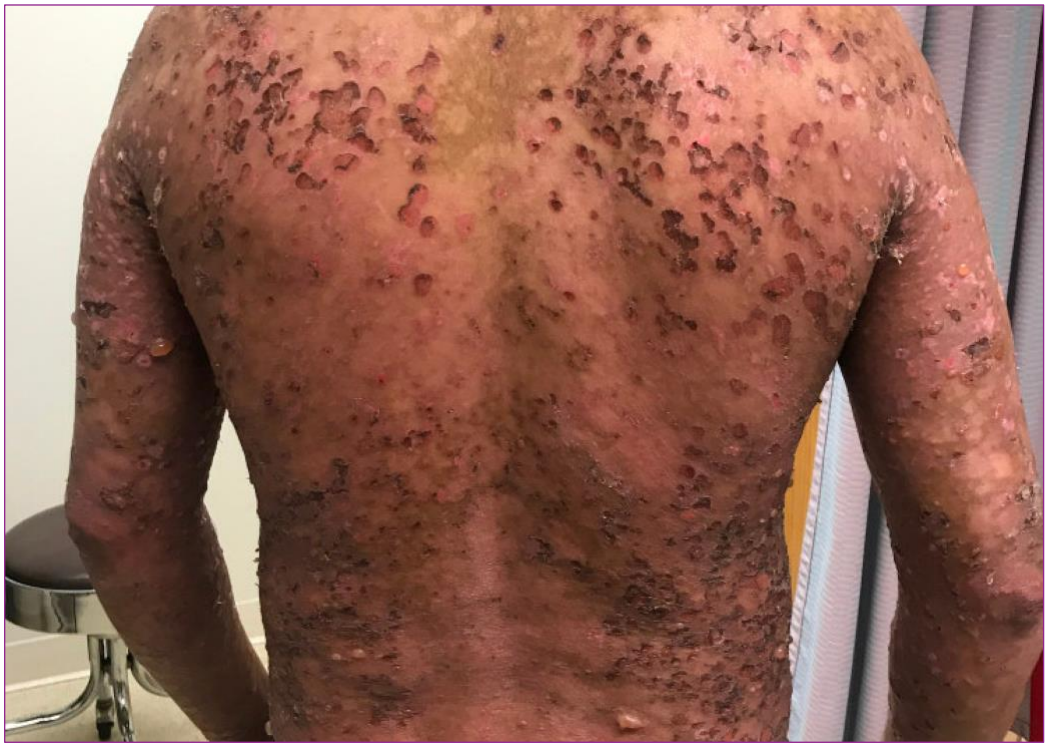

Figure 1. Numerous, scattered, eroded, erythematous plaques and bullae on the back and bilateral upper extremities.

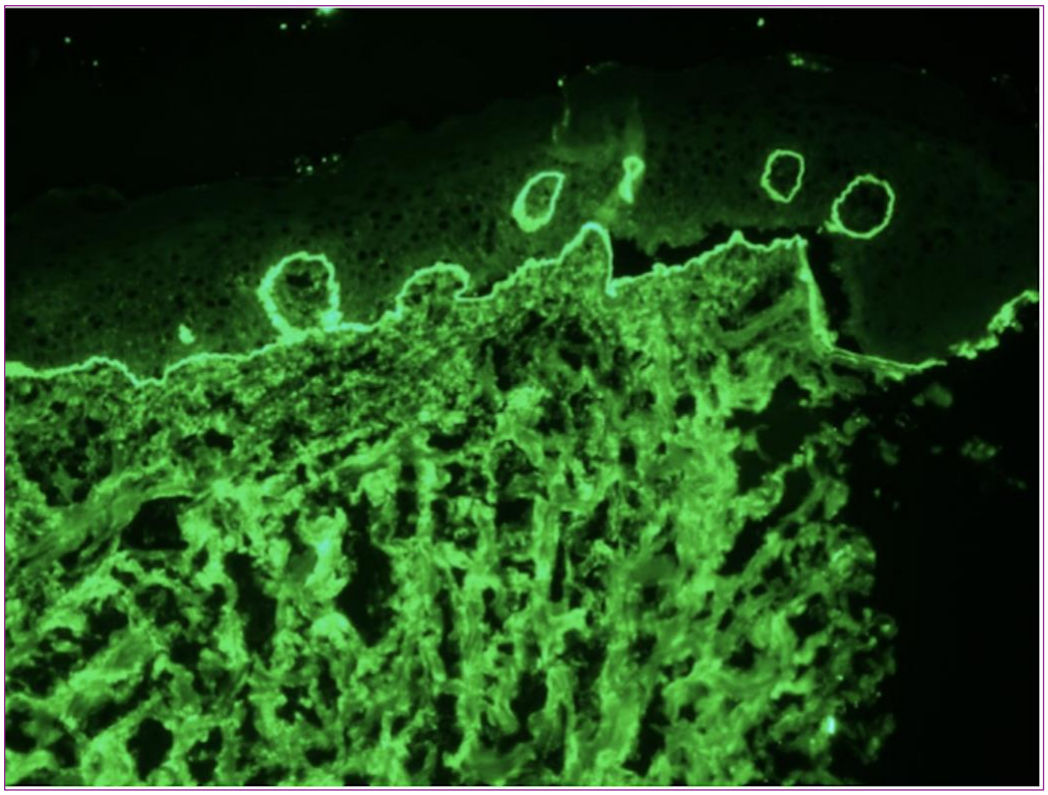

Figure 2. Direct immunofluorescence staining revealing linear IgG reactivity in the basement membrane zone.

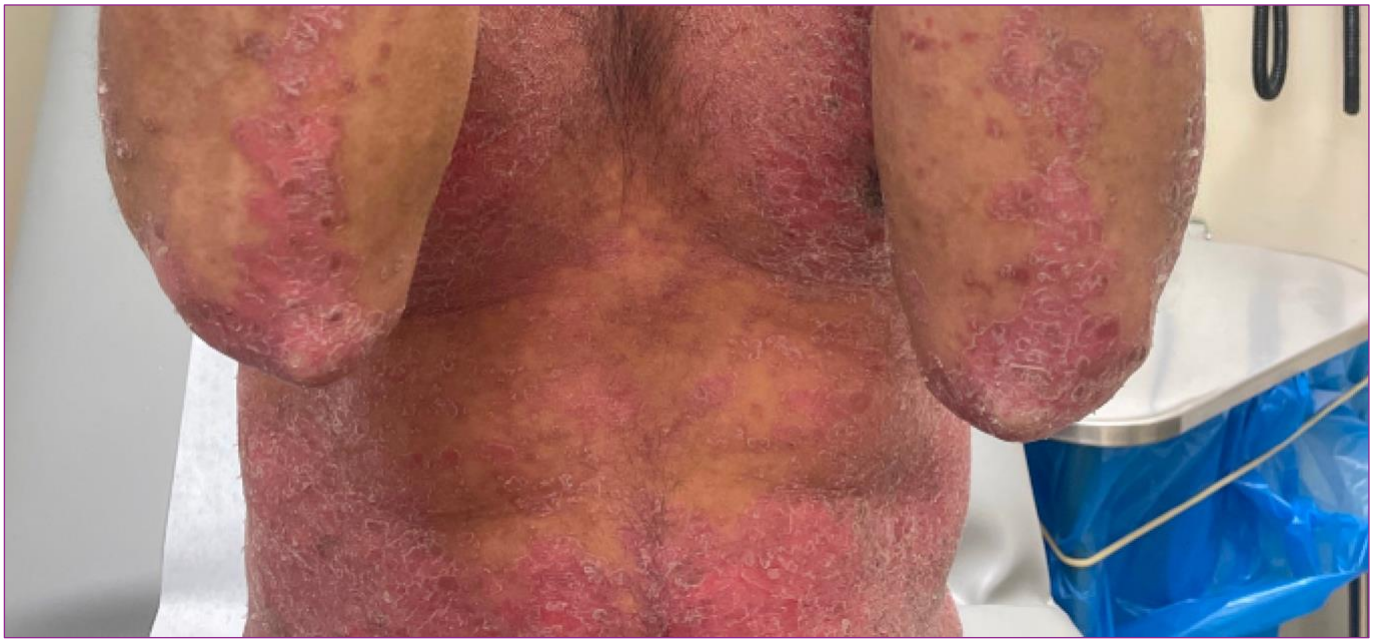

Figure 3. Scattered, pink to erythematous, well-defined plaques with overlying scale on chest, abdomen, and extensor forearm.

psoriasis and anti-p200 pemphigoid was made. He was treated with a course of methylprednisolone and mycophenolate mofetil to prevent a recurrent bullous outbreak and had marked clinical improvement.

DISCUSSION

The coexistence of psoriasis and autoimmune bullous diseases (AIBDs) has been well-reported. The first documented case describing the coexistence of these two entities dates back to 1929.[2] A large-scale study evaluating possible co-morbidities in 51,800 patients with psoriasis found an increase in the prevalence of pemphigoid.[3,4] A case series evaluating 145 cases of patients with coexisting psoriasis and AIBDs found the majority of cases were complicated by bullous pemphigoid (63%), anti-p200 pemphigoid (37%), or a combination (8%).[3]

The first case of anti-p200 pemphigoid associated with psoriasis was reported by Chen et al. in 1996.[5] A patient with a history of psoriasis presented with bullae over their psoriatic lesions as well as on uninvolved skin. Further workup with both direct and indirect immunofluorescence was not consistent with any autoimmune bullous disease previously identified. Through further analysis, Chen et al. were the first to detect a novel autoantibody against a 200-kDa dermal protein in the lower lamina lucida of the basement membrane zone.[5,6] They described this entity, which later became known as anti-p200 pemphigoid, as distinct from other AIBDs and hypothesized an association with psoriasis.[5]

The prevalence of anti-p200 pemphigoid is unknown. This is due, in part, to the fact that tests to detect the pathogenic antibody are often not commercially available.[7]. Unfortunately, in our patient's case and many others, immunohistochemistry testing to confirm the diagnosis is unable to be performed. Therefore, it is hypothesized that many cases of anti-p200 pemphigoid are undiagnosed. Although its prevalence is not

well understood, there has been much data to support anti-p200 pemphigoids association with psoriasis. In fact, psoriasis has been noted in about 30% of published cases of anti-p200 pemphigoid.[1]

The reasoning for this association is not well understood. It is hypothesized that the inflammation within psoriatic lesions themselves may trigger antigen exposure and auto-antibody formation. For instance, the neutrophilic infiltrate in psoriatic lesions releases matrix metalloproteases that degrade multiple targets, including laminins. Exposure of degraded laminin then triggers the production of autoantibodies that eventually lead to anti-p200 pemphigoid lesions.[3]

Patients with anti-p200 pemphigoid present with bullae and urticarial plaques, typically on acral skin.[6] Psoriasis typically precedes the diagnosis of anti-p200 pemphigoid in these patients by a mean time of 15.1 years.[7] The mean age of onset in anti-p200 pemphigoid patients is 65.5 years, however, cases of bullous disorders displaying IgG reactivity to the p200 antigen have been reported in patients ranging from 5 to 94 years old.[7,8] Interestingly, our patient was diagnosed at the lower end of this age range.

A standard treatment regimen for psoriasis and anti-p200 pemphigoid has not been identified. The most utilized monotherapy is systemic corticosteroids at a dose of about 40-60mg of prednisolone daily.[6] However, recurrence of disease has been noted to occur upon steroid tapering.[6] In fact, a systematic review of patients with anti-p200 pemphigoid found that 39.6% had at least one flare over the course of their disease and recommends systemic corticosteroids in conjugation with an adjuvant agent for long-term disease control.[9] Adjuvant therapies noted to be effective include dapsone, tetracyclines, cyclosporine, azathioprine, mycophenolate mofetil, intravenous immunoglobulins, methotrexate, colchicine, plasmapheresis, and rituximab.[9] Our patient was initially treated with dapsone, however, this was stopped due to noted anemia. He was then treated with mycophenolate mofetil and methylprednisolone to prevent the recurrence of bullous outbreaks. At follow-up, he was significantly better and transitioned to methotrexate, a more typical treatment for psoriasis. He remains free of active bullous disease.

CONCLUSION

Patients with a history of psoriasis and new onset bullous lesions may have anti-p200 pemphigoid and, if so, a treatment regimen consisting of systemic corticosteroids plus an adjuvant therapy, notably Dapsone, is an effective form of therapy.

Conflict of Interest Disclosures: None

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