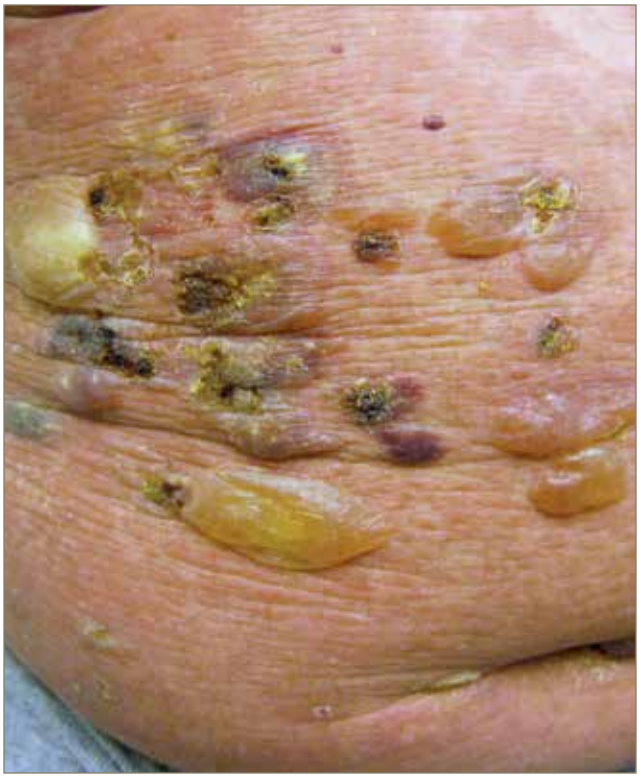
Answer to Dermacase *continued from* **page 963**

5. Bullous pemphigoid

Bullous pemphigoid (BP) is a chronic autoimmune subepidermal blistering disease of the skin with infrequent mucous membrane involvement.[1] The estimated annual incidence is 6 or 7 new cases per million population. Bullous pemphigoid predominantly affects elderly patients, with typical onset after the age of 60 years; however, individuals in any age group can be affected, including children.[2,3] There is no documented sex, racial, or geographic predilection.[4]

Bullous pemphigoid is caused by autoantibodies against the hemidesmosomal antigens BP180 (BPAG2) and BP230 (BPAG1) located in the dermal-epidermal junction of the skin.[1] The classic clinical presentation is a widespread pruritic eruption consisting of multiple tense vesicles and bullae (usually serous and, less often, blood-tinged) arising on both normal-appearing and erythematous skin. Erosions and crusts are observed in areas where blisters have ruptured. Although any skin surface can be affected, sites of predilection include the flexural aspects of the extremities and intertriginous areas.[5] The mucosal surfaces might also be involved, although this is less common. Blisters and erosions can occur on the oral mucosa and, less frequently, on the eyes, nose, pharynx, esophagus, and anogenital region.

In many instances, patients with BP experience a nonblistering phase of the disease that precedes the onset of the bullous eruption by several weeks or months.[6] During this prodromal phase, pruritus can be mild to severe and skin findings are variable, ranging from none to urticaria, eczematous plaques, or excoriated papules and nodules.



Diagnosis

The diagnosis of BP is confirmed by histopathologic and immunofluorescence studies. Two skin biopsies are beneficial in confirming the diagnosis—one from the edge of a blister for routine hematoxylin and eosin staining and the other from perilesional, uninvolved skin for direct immunofluorescence.[1] The patient's serum can also be collected for detection of the pathogenic autoantibodies via indirect immunofluorescence, immunoblotting, or enzyme-linked immunosorbent assay.

The differential diagnosis of BP includes other blistering disorders such as cicatricial pemphigoid, epidermolysis bullosa acquisita, linear immunoglobulin A bullous dermatosis, dermatitis herpetiformis, pemphigus vulgaris, bullous lupus erythematosus, bullous drug eruption, bullous impetigo, and staphylococcal scalded skin syndrome. Cicatricial pemphigoid and epidermolysis bullosa acquisita are the most difficult diseases to differentiate from BP. In addition, erythema multiforme, Stevens-Johnson syndrome, and toxic epidermal necrolysis should be considered for more acute and severe presentations, whereas allergic contact dermatitis and arthropod bites can resemble localized BP.[7,8]

Treatment

The goal of treating BP is to heal existing disease and prevent the development of new blisters. Choice of therapy depends on the extent and severity of the disease as well as the patient's age, medical comorbidities, and medication profile. Bullous pemphigoid can be associated with substantial morbidity, especially in the elderly population. One-year mortality rates range from 6% to 40%.[9]

Systemic corticosteroids remain the mainstay of treatment in severe cases of BP.[10] Therapy with prednisone is initiated at a dose of 0.5 to 1 mg/kg daily until control of disease activity is achieved, with subsequent tapering until resolution. If long-term therapy is required, physicians should ensure that patients are receiving adequate calcium and vitamin D supplementation with or without bisphosphonate therapy to prevent osteoporosis.

High-potency topical corticosteroids (such as 0.05% clobetasol propionate) are frequently used in combination with prednisone, but can also be utilized as monotherapy. In fact, one study demonstrated that ultrapotent topical corticosteroids appeared to be as effective as prednisone.[11] Thus, for patients with mild or localized disease, multiple medical comorbidities, or contraindications to prednisone, ultrapotent topical corticosteroids are the treatment of choice.

Additional therapeutic options for BP include other immunosuppressive agents such as azathioprine, methotrexate, mycophenolate mofetil, cyclophosphamide, cyclosporine, and chlorambucil, as well as anti-inflammatory agents such as dapsone and tetracycline antibiotics with or without nicotinamide. Intravenous immunoglobulin, plasmapheresis, and rituximab also have demonstrated efficacy in treating BP, although these are off-label indications.[5,12]

The intense pruritus frequently associated with BP can be addressed through the use of topical or oral antipruritic agents (ie, antihistamines). Daily oatmeal baths might also provide symptomatic relief of pruritus. Dressings and topical antibacterial agents should be applied to eroded areas to prevent secondary infection.

Most patients with BP will experience complete resolution within several months of treatment, although some individuals might have disease that persists for years. Recurrence is not uncommon.[13,14]

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Competing interests
None declared

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