Bullous pemphigoid

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ESSENTIALS Autoimmune bullous diseases of the skin are a heterogenous group of blistering diseases that affect the skin and/or mucosal membranes. They are associated with significant morbidity and mortality and may present to several different specialists. They are broadly divided into two groups depending on the location of the blisters formed in the skin, which may be subepidermal (pemphigoids, linear IgA disease, dermatitis herpetiformis, epidermolysis bullosa acquisita) or intraepidermal (pemphigus group). Pathogenic autoantibodies (IgG, IgA) target either the proteins that provide keratinocyte adhe- sion (intraepidermal disease) or the proteins of hemidesmosomes that attach the basal cell layer to the basement membrane zone (subepidermal disease). Blisters are often firm in subepidermal dis- ease but burst easily in intraepidermal disease, leading to erosions. Systemic steroids are the mainstay of treatment and may be required at high doses (e.g. 0.5–1 mg/kg), with the pemphigus group often requiring higher doses. Topical steroids and oral tetracyclines are especially beneficial in the pemphigoid group. Over the past years rituximab (monoclonal anti-CD20 antibody) has shown promising results in the treatment of pemphigus. Other immunosuppressant drugs are often needed as steroid sparing agents to control the disease. Introduction Autoimmune bullous diseases are caused by IgG and/or IgA auto- antibodies to the components of the epidermal structures that pro- vide cell-to-cell (desmosome) or cell to basement membrane zone adhesion (hemidesmosome). Antibodies to desmosomes lead to intraepidermal disease and to hemidesmosomes, subepidermal dis- ease. Table 23.4.1 shows the immunopathological characteristics of these diseases including the targeted structures. Clinically, they af- fect a wide age range and can present as tense blisters (subepidermal) or erosions/flaccid blisters (intraepidermal) with or without mu- cosal involvement and scarring. Clinical characteristics and differ- ential diagnoses are summarized in Table 23.4.2 and Table 23.4.3, respectively. Subepidermal diseases Firm blisters are often seen when antibodies target components of hemidesmosomes (Fig. 23.4.1) leading to separation of basal cells from the basement membrane zone and dermis. There might be mu- cosal involvement or scarring. Antibodies can be visualized in the form of a line along the basement membrane zone with immuno- fluorescence studies. Subepidermal diseases include bullous pem- phigoid, mucous membrane pemphigoid, pemphigoid gestationis, linear IgA disease, epidermolysis bullosa acquisita, and dermatitis herpetiformis. Bullous pemphigoid Aetiology and epidemiology This is the most common autoimmune blistering disease in the West, affecting 7–40 people per million per year, with a female predilection. The exact cause is unknown but trigger factors such as trauma, radiation, and drugs have been suggested. There is evidence of asso- ciation with loop diuretics and bullous pemphigoid is more preva- lent in patients with neurological diseases such as cerebrovascular disease, dementia, Parkinson’s disease, motor neuron disease, and multiple sclerosis. The incidence is on the rise with increase in the ageing population. Pathogenesis There are two main target antigens, a

transmembrane protein known as BP180 (collagen XVII, BPAg2) and an intracellular protein, BP230 (BPAg1). Animal models have shown antibodies to these struc- tures to be pathogenic. Blisters are caused by IgG antibody–antigen binding, complement activation, recruitment of inflammatory cells, and secretion of proteolytic enzymes that separate the basal cells from the underlying dermis. Clinical features Erythematous plaques and tense blisters on trunk and limbs often present with intense itch (Fig. 23.4.2). In some cases blisters might not be apparent and only an inflammatory, excoriated, and eczema- like rash can be seen. Involvement of mucosal membranes is rare 23.4 Autoimmune bullous diseases Kathy Taghipour and Fenella Wojnarowska

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