

Immunobullous Skin Conditions

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Diagnosis	Antigen (Weight, kDa)	Other
BP	BPAg1 (230), NC16A domain of BPAg2 (180)	BPAg2=BP180=collagen XVII; linear IgG and C3 at DEJ on DIF; epidermal staining on salt-split skin
Cicatricial pemphigoid (mucous membrane pemphigoid or benign mucosal pemphigoid)	C terminal of BPAg2 (180), laminin-5, BPAg1 (230)	Oral involvement most common; laminin-5=laminin-332=epiligrin; linear IgG and C3 at DEJ on DIF
Ocular cicatricial pemphigoid	β 4 subunit of α 6 β 4 integrin (200)	Linear IgG and C3 at DEJ on DIF
Lichen planus pemphigoides	BPAg2 (180)	Linear IgG and C3 at DEJ on DIF
Pemphigoid gestationis	BPAg2 (180)	Sometimes BPAg1; linear IgG and C3 at DEJ on DIF
Pemphigus vulgaris	Dsg3 (130), Dsg1 (160)	Dsg3 (mucosal); Dsg1 (cutaneous); intercellular IgG on DIF; IIF using monkey esophagus
Paraneoplastic pemphigus	Dsg3 (130), Dsg1 (160), plectin (500), desmoplakin I (250), desmoplakin II (210), BPAg1 (230), envoplakin (210), periplakin (190)	Common associations include (in order of frequency) NHL, CLL, Castleman disease, thymomas; IgG intercellular and linear at DEJ on DIF; IIF using rat bladder
Pemphigus foliaceus	Dsg1 (160)	Fogo selvagem endemic in Brazil, vector <i>Simulium</i> black fly; intercellular IgG on DIF; IIF using guinea pig esophagus
IgA pemphigus	Desmocollin (100–110)	IEN, SPD; intercellular IgA on DIF
Pemphigus vegetans	Dsg3 (130), Dsg1 (160)	Neumann disease (severe); Hallopeau disease (milder); intercellular IgG on DIF
Pemphigus erythematosus (Senear-Usher syndrome)	Dsg1 (160)	Lupus and pemphigus foliaceus overlap; IgG intercellular and linear at DEJ on DIF
Chronic bullous dermatosis of childhood	BPAg2 LAD-1 (97)	Linear IgA at DEJ on DIF
LAD	BPAg2 LAD-1 (97)	Vancomycin can be a drug-induced cause; linear IgA at DEJ on DIF
Dermatitis herpetiformis	Epidermal transglutaminase 3 (77)	Antiendomysial and antigliadin antibodies; granular IgA in dermal papillae on DIF
Epidermolysis bullosa acquisita	Collagen VII (290)	Linear IgG at DEJ on DIF; dermal staining on salt-split skin
Bullous eruption of systemic lupus erythematosus	Collagen VII (290)	Linear IgG at DEJ on DIF; dermal staining on salt-split skin

Abbreviations: BP, bullous pemphigoid; BPAg, bullous pemphigoid antigen; DEJ, dermoepidermal junction; DIF, direct immunofluorescence; Dsg, desmoglein; IIF, indirect immunofluorescence; NHL, non-Hodgkin lymphoma; CLL, chronic lymphocytic leukemia; IEN, intraepidermal neutrophilic; SPD, subcorneal pustular dermatosis; LAD, linear IgA disease.

Practice Questions

- 1. What is the antigen targeted in ocular cicatricial pemphigoid?**
 - a. $\alpha 6$ subunit of $\alpha 6\beta 4$ integrin
 - b. $\beta 4$ subunit of $\alpha 6\beta 4$ integrin
 - c. desmocollin
 - d. laminin-5
 - e. laminin-6

- 2. Which of the following conditions is most commonly associated with paraneoplastic pemphigus?**
 - a. Castleman disease
 - b. chronic lymphocytic leukemia
 - c. multiple myeloma
 - d. non-Hodgkin lymphoma
 - e. solid organ adenocarcinomas

- 3. A patient develops multiple vesicles and bullae arranged in a crown of jewels configuration after receiving an antibiotic in the hospital for sepsis. Which of the following drugs is the most likely cause?**
 - a. chloramphenicol
 - b. ertapenem
 - c. meropenem
 - d. piperacillin-tazobactam
 - e. vancomycin

- 4. What is the suspected vector for fogo selvagem?**
 - a. *Anopheles*
 - b. *Glossina*
 - c. *Lutzomyia*
 - d. *Phlebotomus*
 - e. *Simulium*

- 5. Chronic bullous dermatosis of childhood is characterized by autoantibody production against:**
 - a. BPAg2 LAD-1
 - b. C terminal of BPAg2
 - c. collagen VII
 - d. collagen IV
 - e. NC16A domain of BPAg2

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