The clinical and immunopathological manifestations of anti-epiligrin cicatricial pemphigoid, a recently defined subepithelial autoimmune blistering disease

Cicatricial pemphigoid (CP) is a rare, acquired, autoimmune, subepithelial blistering disease. It primarily affects mucous membranes but it also may involve the skin. Morbidity is associated with the propensity for scar formation and may be especially severe when mucosal surfaces such as the conjunctivae, larynx, esophagus, or urethra are involved. Over the last 10 years, studies in a number of laboratories have shown that circulating autoantibodies in patients with CP may target one of several different autoantigens. One subset of patients with the CP-phenotype have IgG anti-basement membrane autoantibodies against laminin 5 (α3β3γ2) (i.e., patients with anti-epiligrin CP [AECP]). This review discusses recent advances in the understanding of CP and emphasizes salient features of AECP pathophysiology.


16 Rahbar R, Shapshay SM, Healy GB. Mucous Membrane Pemphigoid (mucous membrane [cicatricial] Pemphigoid, cicatrical pemphigoid, anti-epiligrin cicatricial pemphigoid, benign mucous membrane pemphigoid, benign mucosal pemphigoid). Aimee S. Payne. Share on Facebook. Mucous membrane pemphigoid (MMP) encompasses a group of rare autoimmune blistering diseases that can be relatively difficult to diagnose and manage, reflecting the potential diversity and severity of organ system involvement. The patient may need to consult with several doctors (dermatology, oral medicine, ophthalmology, and otolaryngology most frequently). Anti-epiligrin cicatricial pemphigoid (AECP) is a chronic, mucous membrane-dominated, subepithelial blistering disease characterized by circulating anti-basement membrane zone auto-antibodies to laminin 5. Recent studies have shown that people with AECP have an increased relative risk for malignant tumours. In this report we describe two patients with AECP. We report the clinical and immunopathologic findings in a cohort of 35 patients with anti-epiligrin cicatricial pemphigoid (AECP). These patients have a mucosal predominant subepithelial blistering disease that is clinically indistinguishable from other forms of cicatrical pemphigoid. Anti-epiligrin cicatricial pemphigoid is an autoimmune subepithelial blistering disorder of mucous membranes and skin.