## 2.3.5 Porphyria cutanea tarda

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| **Definition** | Metabolic disorder with disturbed hemoglobin synthesis and accumulation of phototoxic metabolites (porphyrins) |
| **Clinic** | Increase photosensitivity with dense blisters, milia and scars in sun-exposed areas, especially backs of hands |

### Histopathology

- *Subepidermal blister with little inflammation*
- *Retained dermal papillae which extend into blister lumen (festooning, naked papillae)*
- Deposits of eosinophilic *PAS-positive material* around small vessels in superficial dermis
- Apoptotic keratinocytes. Accumulations of degenerated basement membrane type IV collagen in the spinous layer (caterpillar bodies)

### Additional studies

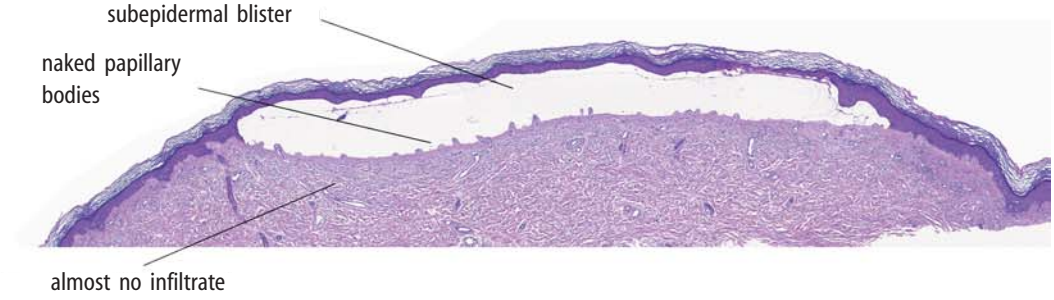
DIF: IgG (rarely IgM and C3) are trapped at DEJ and around papillary dermal vessels. Type VII collagen at base of blister, type IV on roof. Diagnosis confirmed by elevated urine porphyrins, blood examinations, genetic and enzymatic studies

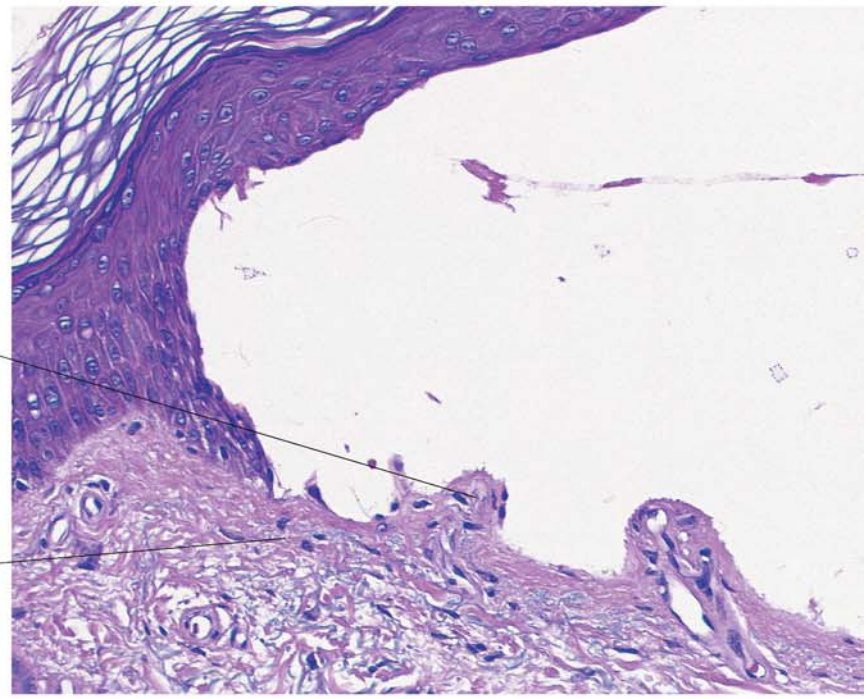
### Differential diagnoses

- **Bullous pemphigoid and epidermolysis bullosa acquisita**
  Broad-based subepidermal blister. Usually infiltrate with eosinophils in bullous pemphigoid, but cell-poor types exist. EBA can be histologically identical to PCT and only be separated with biochemical studies. DIF: Deposits of IgG and C3 at DEJ
- **Epidermolysis bullosa**
  Congenital mechanobullous disorder with many forms. EB simplex forms have degeneration in basal layer and blisters but no scarring. Junctional and dystrophic forms have subepidermal blisters. Blisters usually with minimal infiltrate; often scarring. IF examination negative
- **Bullous disease of diabetes**
  Subepidermal blister with little infiltrate. No apoptotic epidermal keratinocytes

### Comments

Erythropoietic protoporphyria (EPP) features marked photosensitivity early in life and distinctive facial scarring. Biopsy reveals more massive deposits of eosinophilic PAS-positive material in dermal papillae and around dermal vessels


subepidermal blister
naked papillary bodies
almost no infiltrate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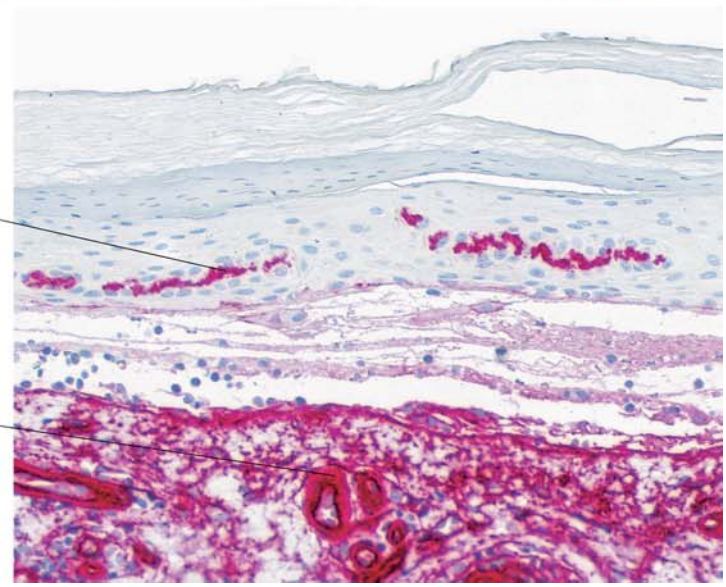
caterpillar bodies – collagen IV-positive basement membrane fragments
PAS-positive, thickened vessel walls

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