Seven types of porphyria are recognized. The light sensitivity in the six types with cutaneous lesions is due to the production of porphyrin precursors that are fluorescent at wavelengths of 400 nm.
In *erythropoietic porphyria*, a very rare disease that typically develops during infancy or childhood, recurrent vesiculobullous eruptions in the skin may appear. In an autosomal recessive pattern, hypertrichosis and brown-stained teeth that fluoresce are additional features.

In *erythropoietic protoporphyria*, the usual reaction to light is erythema and edema followed by thickening and superficial scarring of the skin. In rare instances, fatal liver disease develops quite suddenly, usually in persons of middle age but occasionally in patients only in the second decade of life.
In *porphyria variegata*, different members of the same family may have either cutaneous manifestations identical to those of *Porphyria cutanea tarda*. Also, a sharp fluorescence emission peak at 626 nm is specific for the plasma of *Porphyria variegata*.
Three forms of *porphyria cutanea tarda* can be distinguished: sporadic, familial, and hepatoerythropoietic. In the sporadic form, only the hepatic activity of uroporphyrinogen decarboxylase is decreased. Almost all patients are adults, and no clinical ... form can occur without any precipitating factor, in most instances, in addition to the inherited enzymatic defect, an...
In *hereditary coproporphyria*, a very rare disorder, there are episodic attacks of abdominal pain and a variety of neurologic and psychiatric manifestations. There are also cutaneous manifestations indistinguishable from those of porphyria cutanea tarda and porphyria variegata.
Histopathology

The histologic changes in the skin lesions are the same in all six types of porphyria. In mild cases, homogeneous, pale, eosinophilic deposits are limited to the immediate vicinity of the blood vessels.
In severely involved areas, which are most common in erythropoietic protoporphyria, the perivascular mantles may be thickened. The perivascular mantles can be identified by staining with Sudan IV or Sudan black B. In addition, the PAS-positive dermal-epidermal basement membrane zone may be thickened.
In areas of sclerosis, which occur especially in porphyria cutanea tarda, the collagen bundles are thickened. In contrast to scleroderma, PAS-positive, diastase-resistant material is often present in the dermis in perivascular locations.

The bullae, which are most common in porphyria cutanea tarda and least common in erythropoietic protoporphyria, arise on sun-exposed skin at sites of previous trauma and heal with scarring. It is quite characteristic of the bullae of porphyria cutanea tarda that...
Porphyria Cutanea Tarda = اﻠآﺠﻠﺔ اﻠﺠﻠدﻴﺔ اﻠﺒورﻔﻴرﻴﺔ

dermal papillae often extend irregularly from the floor of the bulla into the bulla cavity (104,122). This phenomenon, referred to as festooning, is explained by the rigidity of the upper dermis. Eosinophilic material within and around the capillary walls in the papillae and the papillary dermis is explained by the rigidity of the upper dermis. The epidermis forming the roof of the blister often contains eosinophilic bodies that are elongate and som...
Pathogenesis

The substance around dermal vessels has the appearance of hyalin because it consists of homogeneous, eosinophilic material produced in large amounts in the dermis of lipoid proteinosis and produced by fibroblasts as amorphous material, is absent.

On electron microscopic examination, concentric duplications of the basement membrane around the dermal blood vessels are seen. Intermingled filamentous and amorphous material is seen throughout the upper dermis and even in the mid dermis.
Proof that the perivascular material in porphyria represents excessively synthesized basement membrane material.
In the majority of patients, direct immunofluorescence testing has revealed the presence of immunoglobulins, particularly IgG, in the epidermis. This observation suggests that the immune system may play a role in the pathogenesis of the disease. However, the phenomenon of skin lesions is not merely autoimmune; rather, it is the result of "trapping" of immunoglobulins and complement in the filamentous material.

The enzymatic defect that causes each form of porphyria is known. Enzyme determination
Liver damage is generally mild and chronic in porphyria cutanea tarda. In erythropoietic protoporphyria...
*Pseudoporphyrina Cutanea Tarda*
In patients with chronic renal failure who are receiving maintenance hemodialysis, an eruption indistinguishable from...
Pseudoporphyrria cutanea tarda may also occur following the ingestion of certain drugs, such as furosemide. In patients with pseudoporphyrria, the histologic picture is indistinguishable from that seen in mild cases of porphyria. Histopathology reveals festooned dermal papillae. The blisters usually are situated above the PAS-positive basement membrane zone.
immunoglobulins are often observed in vessel walls and at the dermal-epidermal junction. Complement is
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