Seven types of porphyria are recognized. The light sensitivity in the six types with cutaneous lesions is caused by the production of porphyrin derivatives in the skin. These wavelengths lie in the 400-nm range, representing long-wave ultraviolet light (UVA) and visible light.
In *erythropoietic porphyria*, a very rare disease that typically develops during infancy or childhood, recurrent vesiculobullous eruptions 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*.
Three forms of *porphyria cutanea tarda* can be distinguished: sporadic, familial, and hepatoerythropoietic.
In hereditary coproporphyria, a very rare disorder, there are episodic attacks of abdominal pain and a variety of neurologic and psychiatric symptoms. 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 the papillary dermis. These deposits are best visualized with a PAS stain, being PAS positive and diastase resistant.
In severely involved areas, which are most common in erythropoietic protoporphyria, the perivascular mantle...
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 from the epidermis and extend through the dermal-epidermal membrane zone and thus heal with scarring. It is quite characteristic of the bullae of porphyria cutanea tarda that...
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 due to the presence of eosinophilic material within and around the capillary walls in the papillae and the papillary dermis.

The epidermis forming the roof of the blister often contains eosinophilic bodies that are elongate and sometimes intracellular or extracellular; and (c) electron-dense material thought to be of basement membrane origin (124).
Pathogenesis. The substance around dermal vessels has the appearance of hyalin because it consists of homogeneous, eosinophilic deposits. In large amounts in the dermis of lipoid proteinosis and produced by fibroblasts as amorphous material, it 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 observed 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 superficial dermis. This finding is inconsistent with the permanent photochemical phenomenon; rather, they are the result of "trapping" of immunoglobulins and complement in the filamentous material.

The enzymatic defect that causes each form of porphyria is known. Enzyme determinations may be carried out on cultured skin fibroblasts, erythrocytes, or liver tissue.
Liver damage is generally mild and chronic in porphyria cutanea tarda. In erythropoietic protoporphyria, liver damage can be more severe and can lead to liver failure. In patients with normal liver function tests, biopsy of the liver may or may not show portal and periportal fibrosis.
Porphyria Cutanea Tarda = اﻠآﺠﻠﺔ اﻠﺠﻠدﻴﺔ اﻠﺒورﻔﻴرﻴﺔ
Porphyria Cutanea Tarda = اﻠآﺠﻠﺔ اﻠﺠﻠدﻴﺔ اﻠﺒورﻔﻴرﻴﺔ
Pseudoporphyria Cutanea Tarda
In patients with chronic renal failure who are receiving maintenance hemodialysis, an eruption indistinguishable from porphyria cutanea tarda may not be representative of the porphyria metabolism, and the plasma and fecal porphyrins should always be measured.
Pseudoporphyria cutanea tarda may also occur following the ingestion of certain drugs, such as furosemide, nalidixic acid, and other antibiotics. In drug-induced cases, withdrawal of the medication is curative.

**Histopathology**

In patients with pseudoporphyria, the histologic picture is indistinguishable from that seen in mild cases of porphyria. The histologic examination will show hyperkeratosis, acanthosis, and an increase in the number of basal melanocytes. 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
Porphyria Cutanea Tarda = اﻠآﺠﻠﺔ اﻠﺠﻠدﻴﺔ اﻠﺒورﻔﻴرﻴﺔ
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