Seven types of porphyria are recognized. The light sensitivity in the six types with cutaneous lesions is caused by a porphyrin molecule. 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 sun-exposed skin are characteristic. Additional features include hypertrichosis and brown-stained teeth that fluoresce.

In *erythropoietic protoporphyria*, the usual reaction to light or ultraviolet 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 precipitating factor is found. In the familial form, an autosomal dominant trait is seen, but this form can occur without any precipitating factor, in most instances, in addition to the inherited enzymatic defect, an acquired factor may play a role.
In hereditary coproporphyria, a very rare disorder, there are episodic attacks of abdominal pain and a variety of neurologic and psychiatric symptoms. Additionally, 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 with cutaneous lesions. Differences may be observed, and bullae are present in some instances. In addition, sclerosis of the collagen is present in old lesions.

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 of the dermis may become prominent. The perivascular收藏 may be stained 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.

The bullae, which are most common in porphyria cutanea tarda and least common in erythropoietic protoporphyria, arise ... membrane zone and thus heal with scarring. It is quite characteristic of the bullae of porphyria cutanea tarda that the
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 induced by 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 amorphous material which is anhydrous and contains 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, which are found in deposits at solar-exposed sites on the skin. Rather than being the result of a direct binding 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 deficiencies are associated with the accumulation of precursors of porphyrins, which are then converted in trace amounts into heme.
Liver damage is generally mild and chronic in porphyria cutanea tarda. In erythropoietic protoporphyria,
Porphyria Cutanea Tarda = اﻠآﺠﻠﺔ اﻠﺠﻠدﻴﺔ اﻠﺒورﻔﻴرﻴﺔ
Porphyria Cutanea Tarda = اﻠآﺠﻠﺔ اﻠﺠﻠدﻴﺔ اﻠﺒورﻔﻴرﻴﺔ
Pseudoporphyría Cutanea Tarda
In patients with chronic renal failure who are receiving maintenance hemodialysis, an eruption indistinguishable from...
Pseudoporphyria cutanea tarda may also occur following the ingestion of certain drugs, such as furosemide, which can exacerbate the condition. Withdrawal of the drug usually results in resolution of the pseudoporphyria.

Histopathology

In patients with pseudoporphyria, the histologic picture is indistinguishable from that seen in mild cases of porphyria. Histologically, the lesions are characterized by blisters with festooned dermal papillae. The blisters are typically 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