
a series of fact sheets written
by experts in the field of liver
disease

Extrahepatic Manifestations:

Porphyria Cutanea Tarda (PCT)

Alan Franciscus, Editor-in-Chief

Porphyrins are complex molecules in the body that combine with iron to produce heme, which is responsible for giving blood its red color. They combine with globin to form hemoglobin. **Porphyria** is the name of a group of diseases caused by an excess of porphyrins in the blood. **Porphyria Cutanea Tarda (PCT)** is one of the most common types of porphyria, triggered by a deficiency of an enzyme called uroporphyrinogen decarboxylase (UROD). The reduced activity of UROD results in an overproduction and buildup of the protein uroporphyrinogen in the blood and urine of patients. This results in an abnormal production of heme, a compound found in all body tissues and especially in the liver, bone marrow, and red blood cells.

Causes of PCT

PCT can be caused or triggered by hemochromatosis (accumulation of iron in the liver), heavy alcohol use, estrogens (oral contraceptives, and prostate cancer treatment), and viral infections (HIV and HCV), and possibly smoking. Hepatitis C is the most common infection associated with viral infection. In fact, some studies have found that, in people who have PCT that can be traced to a viral infection, greater than 50% are the result of the hepatitis C virus. An inherited deficiency of UROD is responsible for about 20% of cases of PCT.

The number of people in the United States who have been diagnosed with all forms of PCT is unknown but it is estimated that about 1 in 25,000 have PCT. The rates of PCT are higher in some European populations. PCT occurs in both sexes and it usually strikes in adulthood and can affect any racial or ethnic groups.

Symptoms

The symptoms of PCT are mostly confined to the skin. Skin lesions or blisters are most often seen on the hands, forearms, back of the neck and face and areas exposed to the sun. The skin may become red, blister and peel after exposure to the sun or minor trauma. PCT can also cause either darkening or lightening of the skin, increased facial hair, scarring, alopecia (hair loss), thickening of the skin, itching and premature aging of the skin. In severe cases calcium may be deposited in the skin causing non-healing ulcers.

Liver function enzymes can be abnormal although enzymes are usually only mildly elevated. A liver biopsy is generally performed to assess iron stores and to check for any damage caused by PCT.

Diagnosis

Diagnosis is made based on the presence of skin lesions on physical examination, as well as by the measurement of UROD in blood, urine, a skin biopsy and stool samples.

Management

The signs and symptoms of PCT can be managed, but there is no cure; however it is one of the easiest of the porphyria conditions to manage and treat. The most common management techniques include:

- Phlebotomies (the removal of blood) to reduce iron in the liver. Phlebotomies are given until the level of serum ferritin is reduced to about 20ng/ml. Phlebotomies can also reduce the porphyrins to normal levels in the blood. Once levels of ferritin and porphyrins have normalized PCT does not usually recur.
- Avoiding the sun when possible, use sunscreen and protective clothing such as gloves, hats, pants, and long-sleeved shirts when outside.
- Low doses of chloroquine or hydroxychloroquine (drugs that are used to treat malaria).
- Restriction of foods that contain iron.
- Treatment of the underlying disease (HCV) with interferon plus ribavirin has also been found to decrease skin lesions as well as the amount of UROD's found in urine.

American Porphyria Foundation:

<http://www.porphyrifoundation.com/index.html>

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- Extrahepatic Manifestations (overview)
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- Essential Cryoglobulinemic Vasculitis
- Fibromyalgia
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- Lichen Planus
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- Peripheral Neuropathy (PN)
- Porphyria Cutanea Tarda (PCT)
- Pruritus (Itching)
- Raynaud's Phenomenon
- Sjögren's (Show grins) Syndrome
- Systemic Lupus Erythematosus
- Waldenstrom Macroglobulinemia

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The information in this fact sheet is designed to help you understand and manage HCV and is not intended as medical advice. All persons with HCV should consult a medical practitioner for diagnosis and treatment of HCV.

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