

## Blister In The Sun: A Case Of Porphyrria Cutanea Tarda

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(Control ID: 1334532)

**Learning Objective 1:** Discuss the presentation and diagnosis of porphyria cutanea tarda

**Learning Objective 2:** Review the relationship between PCT and hepatitis C

**Case:** A 67 year-old man with a history of hypertension and end stage renal disease on hemodialysis presented with a blistering skin rash on his hands and face. Two weeks prior, the patient noticed blisters on his right and left hands; a blister on his lower lip developed and burst. The lesions were painful but not pruritic. On exam the patient's blood pressure was elevated, but he was afebrile and vital signs were otherwise normal. His non-dermatologic exam was normal. The skin exam revealed a swollen, hypopigmented lower lip, and multiple flesh-toned plaques and ulcers on palmar and dorsal hands. No vesicles were intact, and there were no lesions on his trunk, lower extremities, or genitals. An HIV ELISA was negative, but the hepatitis C (HCV) viral load >100,000/mL. Fractionated plasma porphyrins were elevated with uroporphyrin 262.4 mcg/L (<0.2mcg/L), heptaporphyrin 214.4 mcg/L (<0.2mcg/L), hexaporphyrin 7.8 mcg/L (<0.3mcg/L), pentaporphyrin 81.5 mcg/L (<0.4mcg/L) and coproporphyrin 12.8 mcg/L (<0.8mcg/L). Skin biopsy of the right hand was consistent with porphyria cutanea tarda (PCT). Based on the presence of elevated plasma porphyrins and characteristic skin biopsy, the patient was diagnosed with PCT.

**Discussion:** PCT is a metabolic condition resulting from the deficiency of hepatic uroporphyrinogen decarboxylase, the fifth enzyme in the heme synthetic pathway. The deficiency leading to PCT can be hereditary or acquired and the incidence of PCT is one in 25,000, typically occurring in patients over thirty. PCT has been described in patients on chronic hemodialysis, with the pathogenesis likely due to the lack of clearance of porphyrin precursors and their accumulation in the serum. Multiple studies have also revealed a strong association between PCT and HCV, with significant geographic variation between regions depending on HCV prevalence. In North America, prevalence of HCV in patients with PCT may be as high as 66 percent. The mechanism linking the two is not known, but several have been proposed including release of iron from hepatocytes due to damage from HCV leading to oxidative stress. PCT is associated with accumulation of porphyrins in the liver, which typically result in mild elevations in serum alanine aminotransferase and aspartate aminotransferase. Porphyrins also accumulate in the skin and exposure to ultraviolet or visible light creates oxidized substances that activate collagenases, leading to disruption of cell components in the subepidermis and the formation of bullae on sun-exposed areas. Other dermatological findings include hypo- and hyperpigmentation and scarring also on sun-exposed areas. PCT should be suspected in any patient who presents with blistering on sun-exposed areas, especially if risk factors are present. The diagnosis is confirmed by demonstration of markedly elevated porphyrins in the plasma or urine. Patients diagnosed with PCT should also undergo testing for HCV, HIV and iron overload. In patients with or without iron overload, the treatment of PCT is repeated phlebotomy. Low-dose hydroxychloroquine is an alternative therapy if phlebotomy is not tolerated. Supportive care, such as analgesia, avoiding sun exposure, and wearing protective clothing, is also recommended during therapy.

## **Atrio-Esophageal Fistula; A Rare But Deadly Complication in the Treatment of Atrial Fibrillation**

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**Learning Objective 1:** Recognize atrio-esophageal fistula as a potential risk of pulmonary vein isolation in the treatment of atrial fibrillation.

**Learning Objective 2:** Identify the signs and symptoms of atrio-esophageal fistula.

**Case:** A 57-year-old man with a history of chronic atrial fibrillation on anticoagulation and previous right parietal ischemic stroke was admitted with acute-onset fevers, vomiting, and disorientation. His surgical history was notable for mitral valve replacement, PFO closure five years prior, and radiofrequency pulmonary vein isolation for restoration of sinus rhythm 30 days prior to admission. On presentation he was hypotensive, tachycardic, febrile, and complaining of a headache. He was volume resuscitated, pancultured, and empirically started on intravenous antibiotics for meningitis. Physical exam revealed equal and reactive pupils with mild periorbital edema and pale conjunctiva, bibasilar rales, and no focal neurologic deficit or abnormal cardiovascular findings. Laboratory studies revealed an INR of 1.5 and lactic acid of 3, but otherwise unremarkable complete blood count and metabolic panel. CSF was unrevealing. CT head showed a subacute left frontal infarct and an old right parietal infarct. Cultures and chest plain film were negative. Initially his symptoms improved with antibiotics, but overnight he developed dark blood per rectum, vertigo, weakness, and shock, requiring intubation and fluid resuscitation. His clinical signs strongly indicated a left atrio-esophageal fistula formation with septic embolic infarct. A CT chest confirmed a fistulous connection between the esophagus and left atrium. MRI brain revealed innumerable, acute, bilateral cerebral and cerebellar embolic infarcts. The patient was immediately taken to the operating room and underwent successful closure of the left atrium and esophageal tear. Despite early surgical intervention, the patient's mental status failed to recover and his family withdrew care.

**Discussion:** Left atrial catheter ablation to encircle the pulmonary veins is becoming increasingly recognized as an effective treatment for atrial fibrillation in cases where the arrhythmic activity originates in the muscle sleeves of the pulmonary vasculature. Less recognized is the small but potentially lethal risk of esophageal injury caused by creating a radiofrequency-induced deep tissue injury to the left atrial posterior wall. While left atrio-esophageal fistula formation occurs in fewer than 1% of cases, it should be considered in any patient presenting with symptoms of meningitis, endocarditis, stroke, or gastrointestinal bleed with a history of pulmonary vein catheter ablation within the last 30 days. Mortality is greater than 50% and those who survive are usually recognized early and have few co-morbidities. Treatment is immediate surgical correction of the defect in the tissues walls, antibiotics, and supportive care. Research in risk reduction is limited, due to the relative infrequency of the condition. Low frequency radiation and continuous monitoring of esophageal temperatures is thought to reduce risk of injury, however, these have not been shown to significantly decrease mortality. Left atrio-esophageal fistula is a rare but often fatal complication of pulmonary vein isolation in the treatment of atrial fibrillation and should be considered in any patient presenting subacutely with sepsis, stroke, or GI bleed.

## An Unusual Case of Meningitis

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**Learning Objective 1:** Diagnose and distinguish medication-related etiologies of community acquired aseptic meningitis.

**Learning Objective 2:** Recognize the role of taking a thorough medication history in the assessment of a patient with suspected meningitis.

**Case:** A 58-year-old woman presented with a severe headache with onset three hours after taking prophylactic amoxicillin for a routine dental cleaning. She described a constant bifrontal headache with mild photophobia, multiple episodes of emesis, diarrhea, fevers, chills, and myalgias. She denied any phonophobia, visual changes, neck stiffness, or other neurologic symptoms. She described similar symptoms five months prior, 6 hours after amoxicillin use for a dental cleaning, which resolved spontaneously in less than 24 hours without medical intervention. Past medical history was remarkable for a right total hip replacement one year ago after a traumatic fracture and anaphylaxis to soy, nuts, peanuts, beans and legumes. She was married, a non-smoker, with no history of alcohol or illicit drug abuse and employed as a health care lawyer. Her only medications were amoxicillin and epinephrine 1:1000 pen. On exam, she was febrile to 103 degrees Fahrenheit, but otherwise non-toxic. Physical exam and detailed neurologic exam were normal. Cerebrospinal fluid was obtained by lumbar puncture. There were 611 nucleated cells in tube 1 (92% neutrophils, 370 red blood cells) and 624 nucleated cells in tube 4 (90% neutrophils, 17 red blood cells). Total protein was 228 mg/dL and glucose 67 mg/dL. The patient received meningitic dosing of vancomycin, ceftriaxone, and acyclovir. Culture, gram stain, and herpes simplex virus DNA amplification of her cerebral spinal fluid were all negative. Her symptoms rapidly resolved within 12 hours. An allergy consultation was obtained. Given the recurrent pattern of symptoms with amoxicillin exposure, she was felt to have amoxicillin-induced aseptic meningitis. She was discharged home with instruction to avoid all penicillin-based products.

**Discussion:** Drug-induced aseptic meningitis (DIAM) is an uncommon cause of community-acquired aseptic meningitis. Its true incidence is unknown. DIAM has been associated with use of nonsteroidal anti-inflammatory drugs, Cox-2 inhibitors, antibiotics, anticonvulsants, and immunomodulation therapies, e.g, IVIG and OKT3 antibodies. It typically presents with a neutrophilic pleocytosis, and can be mistaken for infectious meningitis. DIAM appears to be more common in patients with autoimmune disease. There are only eight other case reports of amoxicillin-induced meningitis in the literature. The mechanism of DIAM is unknown, but hypersensitivity and immune complex formation have been postulated. Resolution occurs within days of antibiotic cessation. This report adds to the evidence-base and emphasizes the importance of taking a thorough medication history in individuals with suspected meningitis.

## One More Way That Smoking is Bad For Your Health

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**Learning Objective 1:** Recognize acute pulmonary eosinophilia as a cause of fever, dyspnea, and cough especially in the setting of a recent airborne exposure

**Learning Objective 2:** Understand the differences in treatment of acute pulmonary eosinophilia versus the treatment of infectious pneumonia

**Case:** A 22 year-old man on military deployment presented to a remote clinic in Central Africa. He had experienced two days of worsening dyspnea on exertion, fever, and fatigue; he denied a cough. His symptoms worsened despite treatment with azithromycin by the field medic. He was a non-smoker, however, over the last two weeks, he had been smoking local cigarettes to help him stay awake during night patrols.

On physical examination, his temperature was 103.9oF, his heart rate was 120 beats/minute, and his respiratory rate was 32 times/minute; his oxygen saturation was 80%. He had decreased air movement bilaterally, in addition to bilateral inspiratory crackles best heard at the bases of the lungs. There was a tactile fremitus on the right side. Laboratory capabilities were limited, but rapid malaria and rapid influenza tests were negative. His chest X-ray demonstrated diffuse alveolar infiltrates and homogenous opacification of the right hemithorax.

He was placed on continuous oxygen by ventimask and started on intravenous ceftriaxone and vancomycin. He was given normal saline, which had a modest effect on his heart rate. Attempts to wean his oxygen consumption were accompanied by an immediate desaturation to 80%. Because of the limited supplies and minimal improvement, he was transported to a facility with greater capabilities. His CBC revealed a leukocytosis with no eosinophils. He underwent bronchoalveolar lavage (BAL) which showed 30% eosinophils. A diagnosis of acute eosinophilic pneumonia was established, and he was started on prednisone. He experienced a rapid resolution of symptoms and was completely weaned off oxygen two days later. The following week he was released from the hospital and able to return home to his unit.

**Discussion:** Acute eosinophilic pneumonia (AEP) is part of a heterogeneous group of disorders known as the eosinophilic lung syndromes. The syndrome typically consists of an acute febrile illness, severe hypoxia, pulmonary infiltrates, increased eosinophils on BAL, and an absence of infection or other cause. The most commonly documented presenting signs and symptoms are dyspnea, fever, cough, and crackles on inspiration. The diagnosis is established based on pulmonary eosinophilia and exclusion of chronic causes of eosinophilic pulmonary disease. It is important to recognize, as was the case in our patient, that pulmonary eosinophilia can exist in the absence of a peripheral eosinophilia due to a pulmonary eosinophil sequestration.

Although idiopathic causes have been described, patients usually develop the syndrome following an airborne toxin exposure. One key exposure that has been well described is new onset smoking, which was seen with our patient. Typically, patients are in their mid-20s and develop symptoms consistent with AEP within one month of initiation of smoking.

The treatment of AEP is steroids, typically IV methylprednisolone. Dosages vary, but the consensus is 60-125mg of methylprednisolone every six hours followed by an oral prednisone taper. Relapses of AEP have not been described in the literature, and the prognosis is typically excellent if identified rapidly and treated appropriately.

## **Mystery Case**

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**Case:** 54 year old woman with bipolar disorder was sent to the emergency room for worsening behavior, not bathing and placing bags of feces around her room.