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Case Report

Porphyria Cutanea Tarda in a 54-Year-Old Patient with a History of Hepatitis C: A Case Report

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ABSTRACT

Background

Porphyria CutaneaTarda (PCT) is the most common type of porphyria and is caused by a decrease in the activity of the hepatic enzyme uroporphyrinogen decarboxylase. It is expressed in both a sporadic form and genetic form and typically presents with cutaneous manifestations described as skin blisters in sun exposed areas.

Case

A 54-year-old male presented complaining of bullous itchy lesions on his hands and upper extremities that were at different stages of healing. Lab results were consistent with porphyria including elevated serum total porphyrins. He was scheduled for phlebotomy every other week for six weeks, hydroxychloroquine, minimize any sun exposure and to completely stop smoking.

Conclusion

Widespread skin lesions associated with underlying liver disease is a characteristic presentation for PCT. Hepatitis C is an antecedent risk factor for PCT, but can now be treated with antiviral therapy with the expectation of attainment of a sustained virologic response. Improvements in arresting progressive liver disease in Hepatitis C patients may improve PCT symptoms, as well.

Keywords: *Porphyria CutaneaTarda; Hepatitis C; Acquired liver disease.*

INTRODUCTION

Porphyria CutaneaTarda (PCT) is the most common type of porphyria. It is caused by a decrease in the activity of the hepatic enzyme Uroporphyrinogendecarboxylase (UROD), which leads to a disruption in the hemesynthesis pathway.[1] UROD catalyzes the step during which uroporphyrinogen III is converted to coproporphyrinogen III. Thus, a decrease in UROD activity will lead to the accumulation of uroporphyrinogen III, which gets oxidized and forms uroporphyrins. These uroporphyrins, when exposed to sunlight, release free radicals through an immune mediated reaction leading to the formation of skin blisters and lesions in sun exposed areas.[2] Uroporphyrins are also excreted in urine, consequently, urine samples from patients with PCT will turn dark black when exposed to the sun.[3]

PCT is expressed in both a sporadic form and genetic form. The sporadic form, which is found in approximately 80% of cases, is commonly seen in patients with a history of liver damage, alcohol use, iron overload due to hemochromatosis, infections such as Human Immunodeficiency Virus, Hepatitis B, or Hepatitis C, chronic tobacco use, or long-term

estrogen use. The genetic form, which is found in about 20% of cases, is caused by an autosomal dominant mutation of the UROD gene, leading to decreased or complete inhibition of the UROD enzyme activity.[4]

Patients with PCT typically present with cutaneous manifestations described as skin blisters in different stages of healing, itchy bullous skin lesions, scarring, or hypo or hyperpigmentation in sun exposed areas. A history of liver injury due to an underlying infection or alcohol use is usually found in patients with PCT. To diagnose PCT, urine and serum porphyrin levels should be obtained which will show an increase in uroporphyrin and heptacarboxylporphyrin levels.[5]

CASE PRESENTATION

A 54-year-old male with a history of hypertension, coronary artery disease, 19 year history of hepatitis C, depression, and 5 years in prison presented to the outpatient clinic in 2008 complaining of some bullous itchy lesions on his hands and upper extremities that were at different stages of healing. Review of systems was significant for photosensitivity, itchiness, and erythema. He denied having any fever, weight loss, or night sweats. His family history was significant for prostate cancer in his father and coronary artery disease and hypertension in his mother. His social history was significant for smoking one pack of cigarettes per day and intravenous drug use. He had no known drug allergies. At home, patient takes clonidine, enalapril, NitroQuick (Nitroglycerin), and mirtazapine. His temperature was 100.4F, blood pressure 140/68, respiration rate 19, and heart rate 92. On physical exam, about 1-2 inches large bullous lesions were found on his hands and upper extremities. Patient stated that these lesions inflate within a couple of hours to days then burst. The rest of the physical exam was completely normal. Given the patient's history, presentation, and physical exam, the bullous skin lesions were suspected to be Porphyria Cutaneatarda (PCT) and urinary porphyrins were ordered. Lab results are shown in Table 1. Based on these lab results, the patient was diagnosed with PCT and was scheduled for phlebotomy every other week for six weeks. A complete blood count and ferritin were continuously monitored at each draw. He was also prescribed hydroxychloroquine and was advised to minimize any sun exposure and to completely stop smoking.

Table 1. Laboratory values at diagnosis.

Laboratory Study	Value	Reference Range
Uroporphyrin (umol/mL)	293	0-4
Serum Total Porphyrins (ug/dL)	21.1	<1.1
Heptacarboxyporphyrins (ug/dL)	4.1	<1.1
Coproporphyrins (ug/dL)	1.7	<1.1
Hexacarboxyporphyrins (ug/dL)	5.9	<1.1
Total Bilirubin (mg/dL)	1.4	0.3-1.2
Direct Bilirubin (mg/dL)	0.4	≤ 0.4
Aspartate Aminotransferase (AST) (units/L)	38	May-40
Alanine Aminotransferase (ALT) (units/L)	51	Jul-56

DISCUSSION

Patients with an underlying liver disease, usually hepatitis C, rarely develop PCT but patients with PCT are usually found to have an underlying liver disease.[6] Chronic infection with hepatitis C was shown to cause an oxidative stress and, therefore, unmask the UROD deficiency in patients who are genetically predisposed. It was very important to analyze the patient's previous medical history and utilize the physical exam findings to form a differential diagnosis that includes PCT and, therefore, order the appropriate labs accordingly. Recent treatment recommendations for PCT now include avoiding sun exposure, phlebotomy, hydroxychloroquine, and treating any underlying infection such as Hepatitis C or HIV. Our patient presented 2008 and he was treated with phlebotomy and hydroxychloroquine. He was also advised to avoid any sun exposure and was counseled on smoking cessation. Now with the current pharmacological advancements in treating hepatitis C, a combination therapy of sofosbuvir-ledipasvir (Harvoni) can be added to the regimen to treat the patient's underlying hepatitis C infection that could be triggering his PCT. Treating the underlying hepatitis C infection in patients with PCT has shown to resolve their skin lesions and to normalize their urine porphyrin levels, therefore decreasing the risk for any future complications.[6] PCT is a easily treated, and nonfatal when recognized. However, underlying liver disease and other comorbidities are often present, and relapse rates may vary. In fact, the range of relapse over a median 1 – 11 year observation period after treatment varies from 20 – 35%.[7] Excess alcohol intake should be discouraged.

CONCLUSION

In this case report, we presented a patient who came in complaining of a blister-like skin rash and pruritus in his hands and upper extremities years after being diagnosed with hepatitis C. The current treatment for PCT mainly consists of treating and avoiding possible triggers, phlebotomy, and hydroxychloroquine. Recent evidence suggests improvements of the PCT skin lesions after treating the underlying hepatitis C. Larger studies are still required to determine the effectiveness of treating PCT by treating the underlying skin infections or possibly by minimizing the triggers. These studies will give us more information and data about all the possible treatment options for PCT.

CONFLICTS OF INTEREST

None.

REFERENCES

1. Jin MF, Wieland CN. Porphyria CutaneaTarda. *Mayo Clin Proc*. 2021; 96(5): 1248-1249. doi: 10.1016/j.mayocp.2020.12.025

2. Anderson HB, Storandt MH, Kallamadi R, Bande D, Matta A. Iatrogenic Iron Overload Causing Porphyria Cutanea Tarda in a Patient With a Rare Nonsense Heterozygous UROD Gene Mutation. 2021; 13(7): e16215. doi: 10.7759/cureus.16215

3. Janssens L, Ji H, Greenlund A. Clin Gastroenterol Hepatol . 2021; 19(9): A19. doi: 10.1016/j.cgh.2020.05.028

4. Rudnick S, Bonkovsky HL. Editorial: Hepatitis C and Porphyria Cutaneatarda in 2020. *Aliment Pharmacol Ther*. 2020; 51(12): 1432-1434. doi: 10.1111/apt.15728

5. Lederhandler M, Chen L, Meehan SA, Brinster NK, Neimann A. A Case of Porphyria Cutaneatarda in The Setting of Hepatitis C Infection and Tobacco Usage. *Dermatol Online J*. 2019; 25(12): 13030/qt11p0982m.

6. Lydia Sastre, Jordi To-Figueras, Sabela Lens, et al. Resolution of Subclinical Porphyria Cutaneatarda after Hepatitis C Eradication with Direct-Acting Anti-Virals. *Aliment Pharmacol Ther*. 2020; 51(10): 968-973. doi: 10.1111/apt.15703

7. Salameh H, Sarairah H, Rizwan M, et al. Relapse of Porphyria Cutaneatarda after Treatment with Phlebotomy or 4-Aminoquinoline Antimalarials: A Meta-Analysis. *Br J Dermatol*. 2018; 179(6): 1351-1357. doi: 10.1111/bjd.16741