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A Presentation of Cutaneous Photosensitivity Diagnosed as A Case of Porphyria Cutanea Tarda in Hepatitis C

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Abstract

BACKGROUND: Porphyria is a rare enzyme metabolic disorder in heme biosynthetic pathway, resulting in accumulation of porphyrins or porphyrin precursor. Porphyria cutanea tarda (PCT) is the most common type of porphyria. PCT is due to deficiency of enzyme uroporphyrinogen decarboxylase (UROD). It can either be sporadic (type1) or familial (type2) and presents typically with cutaneous manifestations.

CASE: A 36-year-old male came with complaints of rash at the back of hand which heals with scar for the past 3 months, it increases on sun exposure. He is a known smoker and alcoholic. Lab findings revealed hepatitis C infection, elevated porphyrins. Patient was started on phlebotomy, low dose chloroquine and treatment for hepatitis C.

CONCLUSION: Extensive lesions on hands in the background of hepatitis C and alcohol history is characteristic of PCT. Treatment of HCV with antivirals reduces the viral load and symptoms of PCT. Phlebotomy to be done. Other preventive measure like reducing sun exposure, minimalize alcohol intake and avoiding possible trigger should be tried.

1. Introduction:

Porphyria Cutanea Tarda (PCT) is most common type of porphyria. It is caused by decrease in the activity of uroporphyrinogen decarboxylase (UROD) [1], which catalyses the conversion of uroporphyrinogen III to coproporphyrinogen III, leading to accumulation of uroporphyrinogen III and gets oxidised to uroporphyrins. These uroporphyrins on sun exposure gets oxidised and leads to formation of blisters, itchy lesions and heals with scar formation. Uroporphyrins will also be excreted in urine and on sun exposure, urine color will be changed to dark black [5]. It can be due to sporadic (type 1) in 80% or familial (type 2) in 20%. It is more common in females. Risk factors include: liver disorders, alcohol consumption, iron overload stage like hemochromatosis , HIV,

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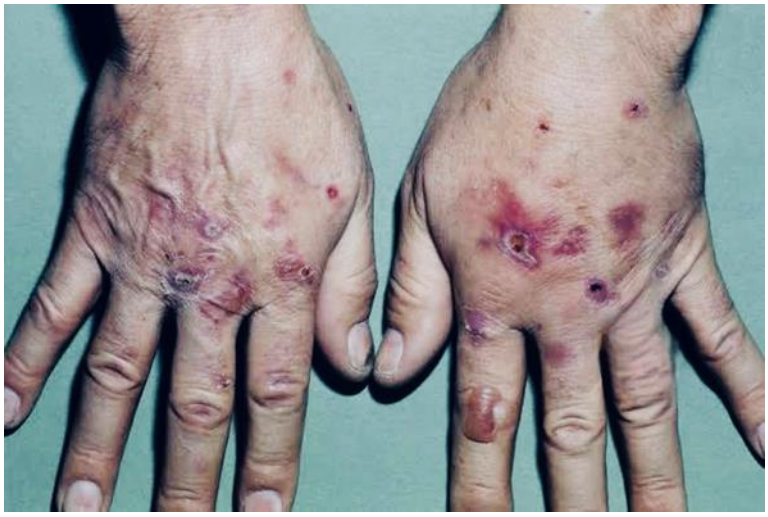

Figure 1: Dorsum of hand showing lesions of various stages surrounded by hyperpigmentation with few lesions showing scabs.

Hepatitis B, Hepatitis C, long term tobacco and estrogen use. The genetic form of PCT is, autosomal dominant, due to mutation of UROD gene leading to inhibition of uroporphyrinogen decarboxylase. PCT patients will have cutaneous presentation of skin blisters which will have different stage of healing, itchy, pigmentation of hypo/hyper, scarring in sun exposed ares [4]. Diagnosis include measuring urine and serum porphyrins level, which shows increase in uroporphyrins. Coproporphyrin and isocoproporphyrin excretion will be increased in feces [2].

Hepatic uroporphyrinogen decarboxylase activity measurement can be done to diagnose [7].

2. Case Presentation:

A 36-year-old male patient, known alcoholic and smoker went to dermatology OPD with complaints of rash on back of hands since 3 months, he explains that the rash starts as boil, ruptures and heals with scar (Figure 1). Rash itches more on exposing to sun. No history of fever, body ache. No known drug allergies were present. Nil significant family history. On general examination, the patient was vitally stable. Local examination of hand: 1-2cm bullous lesion present, along with it different stages of lesion were found. Biopsy of the lesion was taken showing subepidermal bullae with dermal papillae[1]. The patient was diagnosed as epidermolysis bullosa of acqusita type and started on steroids. But the rash did not improve and flare ups were present. On further evaluation, liver function test showed hepatitis like picture with total bilirubin-2.5mg/dl, direct bilirubin-1.2mg/dl, indirect bilirubin-1.3mg/dl, AST-112 units/L, ALT-70 units/L. General medicine was referred. Viral serology was sent and Hepatitis C came as positive. In view of patients general appearance of hypertrichosis and milia which started few months back, serum Iron levels and porphyrins were sent and found to be elevated with uroporphyrin- 260 umol/ml, serum total porphyrins-24 ug/dl. Bullous lesions in the background of hepatitis C infection with elevated porphyrins, Porphyria cutanea tarda was diagnosed. Patient was started on interferon and ribavarin; for hepatitis C, low dose hydroxychloroquine, phlebotomy was done. Other protective measures like sun screen usage, avoidance of alcohol were adviced. Patient is been regularly followed up in out patient basis.

3. Discussion:

PCT patients have been found to have underlying hepatitis C infection more commonly. Chronic hepatitis C infection increases the oxidative stress resulting in PCT in genetically predisposed individuals. It is important to differentiate PCT from other bullous lesion like epidermolysis bullosa acqusita, pseudoporphyria, bullous lupus erythematosus. We diagnosed our patient as PCT with history of alcoholic, lesion increased on sun exposure and investigations like deranged liver function, hepatitis C positivity, biopsy showing

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picture of PCT (subepidermal blisters, which lack inflammatory infiltrate, with preservation of dermal papillae in the lesion's floor ‘festooning') [8] with elevated porphyrins. However, genetic testing was not done. Our patient was started on phlebotomy (400ml/ every second week), hydroxychloroquine (HCQ) for PCT. HCQ works by increasing porphyrins excretion and their synthesis [6]. Interferon, ribavarin for hepatitis C. Treatment of underlying hepatitis C in PCT patients reduces the lesion and normalise urine porphyrins level [3]. Other measures to be advised like using sun protection, identifying and avoiding any triggers. Porphyria can be treated easily once it is diagnosed on time. PCT predispose the patient to cirrhosis or liver cancer [2], hence it is important to educate the patient to avoid alcohol. Patient was regularly followed up and showed improvement in symptoms and normalisation of liver functions.

4. Conclusion:

In the above presented case report, patient came with complaint of blister like lesion in hands in the background of hepatitis C which was undiagnosed until he presented. PCT should be suspected and kept in differentials in patients with classic biopsy picture and deranged liver function. Treatment measures mainly include phlebotomy, hydroxychloroquine, hepatitis C treatment. Recent updates shows treating the underlying hepatitis C infection reduces the severity of PCT lesions.

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