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Porphyria cutanea tarda and hepatocellular carcinoma: a case report

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Introduction & Objectives:

Porphyria cutanea tarda (PCT) is a rare skin disorder originating from a deficit of the liver enzyme uroporphyrinogen (UROD) decarboxylase. PCT may be a risk factor for hepatocellular carcinoma (HCC) and other cancers, but the evidence is unclear. We present an uncommon case of PCT induced HCC for patient with no history of risk factors.

Materials & Methods:

67-year-old women had a 3-month history of multiple symmetrical blisters, crust, scarring and milia on face, dorsum of hands, elbows and neck associated with photosensitivity. Patient had a medical history of arterial hypertension, glaucoma, and diverticulosis. There were no risk factors for liver disease, no family history or alcohol abuse or skin blistering disorders.

Results:

Skin biopsy revealed hyperkeratotic epidermal tissue and the formation of subepidermal bulla. In the superficial derma, thickened and PAS-positive vessels were present. During direct immunofluorescence, immunoglobulin G surrounding vessels was detected. The urine test showed elevated total porphyrin level (547.37 nmol) with the predominance of uroporphyrin, and a normal range of the porphobilinogen. Based on clinical, histological and laboratory findings the diagnosis of PCT was confirmed and 200 mg hydroxychloroquine 2 times per week was started.

Additionally, serology tests for hepatitis B and C were negative, and liver enzymes (AST 50 IU/l, GGT 397 IU/l) were increased. Also, raised serum alpha-fetoprotein (AFP) 14.8 kU/l was found. A contrast-enhanced computed tomography (CT) scan showed 10.4 cm x 11.8 cm x 10.9 cm focal liver lesions with central necrosis in S4, S5, S7, S8 and 2.2 cm x 2 cm analogic lesion in S6. Based on CT results, and raised level of AFP, the diagnosis of HCC was confirmed. Angiography revealed S8 12 cm and S6 4 cm well-vascularized hepatic tumor and transarterial chemoembolization (TACE) for tumor size reduction was performed. During the dermatological follow up after 2 months, the patient reported that no new blisters were appearing, and hyperpigmentation occurred instead of old lesions. Treatment with hydroxychloroquine was continued.

Conclusion:

Early diagnosis and timely treatment of PCT and HCC could help to reduce the negative impact on patients' health.

