

Answers to Dermato-venereological Quiz on pages 95-96

1. The diagnosis is porphyria cutanea tarda, acquired type (PCT). Differential Diagnoses include: other types of porphyria, epidermolysis bullosa acquisita, bullous pemphigoid, pemphigus, linear IgA dermatosis, cutaneous lupus erythematosus, and pseudoporphyria.
2. History of drug use, alcoholism, hepatitis status, family history, iron intake, photosensitivity should be explored. This gentleman was found to be a chronic alcoholic and hepatitis C carrier and there was also a positive history for photosensitivity.
3. Urine for porphyrins, blood for liver function, hepatitis status including hepatitis B and C if indicated, iron profile, AFP level to screen for hepatocarcinoma if hepatitis serology is positive. Ultrasound of the liver should be performed.
4. Urinary porphyrin levels are markedly elevated in PCT patients and in visible light, the excess porphyrin pigment can often be seen. A pink fluorescence colour is seen on Wood's lamp radiation of the urine.
5. The patient should be advised to abstain from alcohol and avoid sun exposure which may aggravate the skin condition. Liver function should be monitored and the patient should also be referred to the hepatology department for further assessment. High plasma iron levels inactivate uroporphyrinogen decarboxylase, the enzyme that is deficient in PCT. In cases of iron overload, phlebotomy can remove excessive iron in patients with PCT. Standard phlebotomy for adults consists of removal of 250-500 mL of blood once or twice per week depending on the patient's tolerance and clinical response. Low dose chloroquine or hydroxychloroquine can be considered in cases where phlebotomy cannot be used.