Porphyria cutanea tarda with Hepatic Involvement. Nodular Elastoidosis (Favre and Racouchot)

L.

Temmerman

In 1976, this 62-year-old patient started developing pruriginous and slightly painful hyperkeratotic plaques on the back of his hands. On clinical examination, he also exhibited a thickened brownish-grey facial skin with excessive furrowing of the forehead and along the frontal hair margin a few crusts and erosion were present. In addition there was elastoidosis

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with numerous black comedones and follicular cysts around the orbits (fig. 4). In the skin of the face and hands thick, PAS-positive amorphous bands were found around the small blood vessels of the upper dermis. A liver biopsy revealed fluorescent tissue under Wood's light. The urinary porphyrins (uroporphyrins) and plasma porphyrins were markedly elevated. Faecal porphyrins were within normal limits. Further laboratory investigations showed abnormal liver function tests (SGOT, SGPT, AP, GMGT), normochromic, normocytic anaemia, hyper-proteinaemia with hypoalbuminaemia and hypergammaglobulinaemia. Treatment with chloroquine (100 mg/day) was not very successful.

Fig. 4. Elastoidosis with cysts and comedones in a patient with porphyria cutanea tarda.

Anetoderma of Jadassohn

S. Van Brabandt

the surrounding ring of normal skin. Histologically, there was no elastine in the dermis. The serology for syphilis was negative. A. A 16-year-old boy exhibited skin lesions on his neck chest and arms. They consisted of small oval

pink to red papules
0.3–0.5 cm in diameter
evolving to