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[Pansclerotic porphyria cutanea tarda after chronic exposure to organic solvents]

[Article in German]

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Abstract

A 63 year old man developed generalized scleroderma with massive sclerotic areas, particularly in the abdominal region, four years after being diagnosed with porphyria cutanea tarda (PCT). He had almost daily exposure to organic solvents (benzene, trichlorethylene) for many years. The cutaneous fibrosis progressed dramatically leading to a pansclerosis, even though the uroporphyrin levels were borderline and the liver enzyme values were normal. Organic solvents are among those substances which can cause a cutaneous fibrosis. The unusually complicated clinical development in our patient was a combination of the two initial factors, the PCT and the long term exposure to organic solvents. The pansclerotic PCT was differentiated from a systemic sclerosis, a disabling pansclerotic morphea and a generalized morphea by means of histological examinations, the absence of a Raynaud phenomenon and the non-involvement of additional organs. Auto-antibodies typical for systemic sclerosis were negative. Using a medium dosage of UVA1 phototherapy and intensive physiotherapy, the progression of the skin disease was stopped and the sclerosis improved.

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