A 45-year-old male farmer from rural West Bengal, India, presented with a history of darkening of skin color, thickening and tightening of skin, and repeated painful blistering mainly over the photo-exposed areas for the preceding two years. He also had a history of aggravation of symptoms during summer seasons. At times, the patient used to pass dark colored urine. There was no history of any drug intake prior to the onset of the symptoms, no history of alcoholism and there was no family history. Cutaneous examination showed thickening and tightening of skin particularly involving the fingers, toes, and face. On his hands and feet and exposed areas, small vesicles, some crusted lesions, and a few hypopigmented and hyperpigmented macules were visible. He had generalized hyperpigmentation in addition to mottled hypo- and hyper-pigmentation and hypertrichosis on temples, forehead, and pinna (Fig. 1). Multiple milia on the external ears were also seen (Fig. 2). Scarring alopecia was noted in few areas of the scalp. Nails were shiny and glossy and some toenails showed onycholysis. Distal phalanges of hands were shortened (Fig. 3) and screening for HIV infection were negative. Ultrasonography of the abdomen was normal. Routine urine examination was noncontributory apart from darker color (Fig. 4) than a control sample. Examination of urine under Wood’s light, after acidification with hydrochloric acid, revealed a bright pink fluorescence (Fig. 5) consistent with the presence of porphyrins. On the other hand, a control sample of urine did not fluoresce at all. Estimation of urinary porphyrin levels confirmed the diagnosis of porphyria cutanea tarda (PCT), the most common form of porphyria, characterized by photosensitivity causing scarring, bullae, milia formation, sclerodermatous skin thickening and hypopigmentation.  

The prevalence of the disease in India is unknown; it is estimated at 1 case in 25,000-50,000 U.S.-population. The disease is caused by the deficiency of the enzyme uroporphyrinogen decarboxylase. Apart from an autosomal dominant heritable form of the disease, PCT may also follow ingestion of toxins like polyhalogenated hydrocarbons. The most frequent variety however, involving about 80% of affected individuals, is the non-familial, sporadic form. Co-occurrence of PCT with different systemic diseases such as chronic hepatitis C infection, HIV infection, diabetes, systemic lupus erythromatosus, dermatomyositis, hematological malignancies, and sideroblastic anaemia have all been reported in the literature. In symptomatic familial PCT, the common inducing agents may not be discoverable.

Even after extensive search we did not find any systemic association in our patient and he was treated with twice weekly low-dose chloroquine therapy and advised photoprotection. His condition markedly improved within three months of initiation of treatment.

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