Porphyrin Metabolism Abnormalities and Its Treatment in a Uremic Patient with Porphyria cutanea tarda

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Dear Sir,

Porphyrin metabolism abnormalities and manifestation of porphyria cutanea tarda (PCT) have been reported in several cases of uremic patients on maintenance hemodialysis so far [1]. To diagnose PCT, porphyrin analysis in the urine is the standard method. However, in most uremic patients, urinalysis is not available because of oliguria or anuria. Some clinicians have tried to diagnose and treat PCT by measuring porphyrin concentrations in the dialysate [1,2] or feces [3] but have failed to make a diagnosis since there were no high amounts of porphyrins in samples. We report herein the successful treatment and the recognition of improvement in abnormal porphyrin metabolism by determining fecal and dialysate porphyrin concentrations.

A 37-year-old female uremic patient with maintenance hemodialysis for 7 years had shown bullous dermatosis, pigmentation, small scar and skin fragility in her face, forearms and hands (fig. 1). She had no history of alcohol abuse or taking contraceptives. She was the only one to have symptoms in her family. The skin biopsy suggested characteristic findings of PCT. In this patient and 4 other age-matched female uremic patients on maintenance hemodialysis who did not have PCT-like dermatosis, as the control group, porphyrin analysis was performed in the feces and dialysate by spectrofluorometer and high-pressure liquid chromatography. The patient showed higher amounts of fecal uroporphyrin (URO), 559 $\mu\text{g}/24\text{ h}$, coproporphyrin (COPRO), 914 $\mu\text{g}/24\text{ h}$, and heptacarboxyporphyrin (HEPTA), 1,785 $\mu\text{g}/24\text{ h}$, than those (15.8 ± 5.3 , 227.8 ± 65.6 , 22 ± 14.7 , respectively) of the control group. There was not much difference in the protoporphyrin concentration between the patient and the control group (267 versus $233.5 \pm 114.7\text{ }\mu\text{g}/24\text{ h}$). Each ratio of URO/COPRO and HEPTA/COPRO in the patient was 0.61 and 1.95 and was higher than that of 0.08 ± 0.04 and 0.03 ± 0.01 in the control group (fig. 2). With the hemodialysis procedure using ethylene vinyl alcohol membrane (1.2 m²), the concentrations of URO and COPRO in the dialysate were 8 and 4 $\mu\text{g}/\text{liter}$ 1 h after the beginning of hemodialysis and 18 and 2 $\mu\text{g}/\text{liter}$ 3 h after that. Meanwhile, the values in the control group

were 3.3 ± 0.6 and 1.3 ± 0.3 (after 1 h) and 3.5 ± 1.2 and 1.3 ± 0.3 (after 3 h), respectively. These results indicated the abnormal porphyrin metabolism and confirmed that the patient had PCT. Several months after the discontinuation of supplements of aluminum hydroxide gel and ferrous sulfate which had been medicated intermittently for years and a minimized chance to have dialyzer and hemodialysis tubing exposed to daylight while hemodialysis was performed with a sunscreen, the patient had no more bullous lesions or erosion. Each ratio of URO/COPRO and HEPTA/COPRO came close to the control value (fig. 2).

Fig. 1. Small scar and pigmentation in a face exposed to sunlight.

Hemodialysis has not been considered to be a useful treatment in PCT associated with uremia because porphyrins are poorly dialyzable. However, on several occasions, porphyrins

permeated into the dialysate could be measured like in our case and others [4,5]. We think that porphyrin analysis in the dialysate is of benefit for the diagnosis of PCT although hemodialysis is not effective as a therapeutic tool. Uremic patients have to expose their blood to daylight during hemodialysis. We think this fact may be one of the precipitating factors in the association of PCT with chronic renal failure. In our case, we have tried to minimize the photosensitizing effect of circulating porphyrins on skin lesions. Consequently, several months later, no additional symptoms appeared. We consider that avoiding blood from exposure to sunlight and daylight is a simple but effective method to prevent the occurrence of PCT in uremic patients with long-term hemodialysis.

Fig. 2. URO/COPRO and HEPTA/COPRO ratio before and after the treatment.

2.0 -
1.5
1.0 -

II

before after treatment Control Patient

URO/COPRO

before after treatment Control Patient

HEPTA/COPRO

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