PHOTOBIOLOGY AND PHOTOPROTECTION

FIVE CASES OF ERYTHROPOIETIC PROTOPORPHYRIA: AN OBSERVATIONAL STUDY WITH OVER 10-YEARS OF FOLLOW-UP

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Background: Erythropoietic protoporphyria (EPP) is an inherited disease of porphyrin metabolism caused by decreased activity of ferrochelatase. Photosensitivity starting from childhood lowers quality of life and liver dysfunction may lead to hepatic failure and death. We describe the clinical and laboratory findings including erythrocyte protoporphyrin (PP) levels on five cases with EPP followed for over 10-years of EPP.

Observation: Five patients of EPP were men and developed the photosensitivity symptoms in childhood. They were diagnosed by an increased erythrocyte PP level. After diagnosed as EPP, the patient had the explanation about protection from sun exposure and received clinical and observations 2 or 3 times per year. Two brother patients (a 28-year old and a 30-year old man) with high PP level which indicated 3500-4500 µg/dL RBC (normal range: 30-86) has not caused liver dysfunction, because sun protection carried without exception was continued. In another two cases who were also brothers with a same genetic abnormality, younger brother (a 23-year old man) showed liver dysfunction with a marked increase of erythrocyte PP level, when sun protection was insufficient. On the other hand, elder brother showed no increase of liver enzyme and erythrocyte. One case (a 32-year old man) showed a fluctuation of more than two times of erythrocyte PP level with a change in life environment. The above showed that liver function of EPP patients have been influenced by life environment and the level of the sun protection.

Key message: These observational studies indicate that protection from sun exposure including clothes and sports is most important management of EPP.