Pregnancy and Porphyria cutanea tarda

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A 31-year-old woman developed typical clinical and laboratory signs of PCT at the end of her second pregnancy coincident with the summer season. She had elevated liver function values without history of alcoholism, hepatitis or chemical liver damage. She had taken oral contraceptive only before her first pregnancy which was normal. Her hormone analytic values including estrogens corresponded to normal values in pregnancy. Venesections had a beneficial affect on her condition. *Key words: Endogenous estrogens; Oral contraceptives; Venesection.* (Received February 15, 1984.)

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Estrogen drugs including contraceptive pills are known as possible elicitors of Porphyria cutanea tarda (PCT), according to literary reports and several basic works (1, 2). This is in agreement with our clinical observations including a 22-year-old woman who was given Trionetta® (Schering AG) 3 years ago and developed a classical PCT after 18 months.

On the other hand, correlations between pregnancy and PCT is only discussed in few reports. In a 28-year-old woman PCT was diagnosed after she had taken oral contraceptives during 2 ½ years. Remission occurred after discontinuation and phlebotomies. Thereafter she became pregnant and in the first trimester she had again shown clinical and chemical signs of PCT which decreased steadily during the second and third semesters. Her second pregnancy 10 months later was uneventful (3). In a 29-year-old woman pregnancy had no effect on her PCT which began one year earlier. Previously she used two kinds of oral contraceptives which she stopped taking 2 years before the onset of the disease. It was also mentioned that the patient totally abstained from alcoholics during her pregnancy (4). It seems therefore of interest to report a case in whom pregnancy led to the clinical expression of PCT.

CASE REPORT

This patient was a 31-year-old housewife with a negative family history for skin diseases including porphyria, hepatitis or chemical liver damage. Her alcohol intake was very low (an occasional glass of wine once a month). She had urticaria on unclear basis in 1971 and complaints similar to cholinergic urticaria during the summer of 1980. She had taken the oral contraceptive Eugynon® (Schering AG) during 1975–1980. The patient had her first pregnancy in 1981 without any pathologic events, and became pregnant a second time in the first days of January 1983. During her later pregnancy she discovered that her urine was sometimes reddish-brown, but her physician, who took only a few simple blood tests, could not find out the cause. In July, although avoiding sun, she noticed blisters on her hands and was sent to me for consultation in August 1983. She had not taken any drugs and abstained from alcohol during her pregnancy.

On closer examination she showed typical signs of PCT including milia and slight facial hypertrichosis, which latter started according to her history 1–2 years ago (!?). Her initial uroporphyrin values were 5725 nmole/l day and koproporphyrins in urine 960 nmole/l day. Haemoglobin was 14.3 g, serum-iron 27.7. She also had elevated ASAT (66 U/l) and ALAT (137 U/l). Liver scintigraphy showed normal values respectively no presence of hepatic tumour; liver biopsy was not performed. Other laboratory tests, including those for diabetes, were normal. Histology of her lesions showed mostly fibrosis but the DIF was

highly characteristic for PCT: a homogeneous deposition of IgG in small dermal vessels (and also fibrinogen here and at junction area). Hormone analysis including estradiol, progesterone and testosterone in serum as well as estriol/u and HPL/s were normal (corresponding to pregnancy). Venesection of 400 ml was initiated and repeated every fourth week with a rather marked effect on clinical symptoms (no more bullæ) and a slow improvement of pathologic values. She gave birth to a normal boy on September 28, 1983.

DISCUSSION

A young woman is thus reported in whom her second pregnancy has lead to PCT. It can however not be excluded that her PCT occurred earlier as a latent condition: she had taken oral contraceptives, noticed a slight hypertrichosis (since first pregnancy) and had some complaints attributed to sun (although of presumably cholinergic character), before her pregnancies. The mechanism of estrogen drug effect on the expression of PCT is unknown (1), except that they can cause hepatopathy (2). In the actual patient the somewhat elevated liver function test indicates slight liver damage, the cause of which is unclear. Among relevant factors in her case there are the following: oral contraceptives were taken by the patient during 5 years without any clinical side effects and stopped 3 years before the first pregnancy, which was normal, a fact indicating that estrogens could not be blamed for causing a grave liver damage. On the other hand subclinical alterations of liver function cannot be excluded. As a hypothesis, on basis of a preexisting liver damage (due to estrogen-containing contraceptives?) PCT was elicited by the summer sun (despite the patient's effort to avoid direct sun exposure). Her endogenous estrogens were not more elevated than normally in pregnancy. It is therefore improbable that they played a role in eliciting her symptoms in the light of the fact that no PCT developed during her first pregnancy.

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