ACUTE INTERMITTENT PORPHYRIA COMPLICATED BY PREGNANCY

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Summary: The case of a woman with acute intermittent porphyria complicated by two pregnancies is described.

Both pregnancies gave an exacerbation of the disease. Premature delivery were necessary for both children due to intrauterine growth retardation. The first child died from respiratory distress syndrome, but the second child was healthy apart from transient abstinence symptoms due to pethidine given to the mother during pregnancy.

The effect of pregnancy on acute intermittent porphyria is discussed.

Acute intermittent porphyria (AIP) is an inborn error of metabolism characterized by a marked over-production of porphyrin precursors such as delta-aminolaevulinic acid and porphobilinogen. It is a rare disease and the prevalence in Sweden has been estimated at only 2-20 per 100.000 with an accumulation of cases in the northern part of the country (Zilliacus, 1967). The most frequent clinical symptoms are abdominal pain, vomiting, constipation and paralysis or paresis. Psychological symptoms are also common.

CASE REPORT

A 34 year old Caucasian woman presented at 6 weeks in her second pregnancy with acute abdominal pain.

She had her first episode of acute abdominal pain combined with dark urine at an age of 23 years. She had 3 more attacks during the next year after which she received a positive diagnosis of AIP. After that she had two or three episodes annually. It has not been possible to identify any precipitating factor.

In her first pregnancy at an age of 32 years she presented with acute abdominal pain 9 weeks pregnant. She remained at the hospital for 4 days during which time the symptoms disappeared. At 36 weeks gestation she was admitted with severe non-specific, abdominal pain and vomiting. Blood pressure was slightly elevated (140/100). Treatment with pethidine and clorpromazine was necessary until delivery. A caesarian section was performed at 38 weeks because of a suspected intra-uterine growth retardation and decreasing serum estriol.

A male baby was delivered weighing 1340 g. Apgar scores were 7 and 8 at 1 and 5 minutes respectively. The baby was treated with continuous positive airway pressure due to insufficient oxygenation but died at an age of 33 hours from respiratory distress syndrome.

The intervals between the attacks after the first pregnancy was shorter than previously. The first attack during this pregnancy subsided after 3 days but she was re-admitted at 10 and 14 weeks at which time she also suffered from vaginal bleeding. At 20 weeks she was finally admitted with abdominal pain, vomiting and paralysis of her right forearm, to remain in hospital until delivery. She received repeated daily injections of pethidine (150-400 mg/day) as well as chlorpromazine (150 mg/day). Treatment with concentrated carbohydrate solution was also instituted. The paresis in her forearm gradually disappeared during her stay in the hospital. Repeated cardiotocography was normal. A contraction stress test performed at 36 weeks showed no signs of fetal distress although serum estriol levels were low.

Ultrasonography at 35 and 37 weeks showed no increase in the biparietal diameter (86 mm) and the fetus was judged small. On these grounds together with the mode of delivery at the first pregnancy it was decided to deliver the baby with a caesarian section. The operation was performed in epidural analgesia.

A female baby weighing 2150 g was delivered. Apgar scores were 9 and 10 at 1 and 5 minutes. The maturity scores gave a fetal age of 38 weeks and the baby was diagnosed as small for date. The baby developed tremor, irritability, vomiting and diarrhoea at an age of 5 days. These symptoms were considered due to abstinence after the pethidine given to the mother and successfully treated with intramuscular diazepam. Mother and child were discharged from hospital in good condition 14 days after delivery.

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DISCUSSION

This case illustrates the classical signs of AIP such as acute abdominal pain, vomiting and paresis.

It is probable that pregnancy is a precipiting factor for AIP (Brodie *et al.,* 1977). Considerable clinical evidence show that female sex hormones activate AIP (Tschudy *et al.,* 1975). It has also been shown that urine porphyrins and their precursors increase during the last month of gestation in non-porphyric women (Lyberatos *et al.,* 1972).

AIP was earlier associated with a poor maternal and fetal outcome but the large review presented by Brodie recorded only one maternal death in 129 pregnancies and a fetal loss of 9 per cent (Brodie *et al.,* 1977).

The treatment is primarily to avoid precipitating factors. Chlorpromazine alone or in combination with opiates like pethidine are often effective for symptomatic treatment. If high doses of opiates are used as in the last pregnancy of this patient it is necessary to consider the effects on the fetus (Calabrese and Gulledge, 1985). In this case the baby developed signs of abstinence. Treatment of the disease with a high carbohydrate diet may lower the excretion of porphyrin precursors (Tschudy *et al.,* 1975).

Although AIP is rare it is necessary for the obstetrician to be aware of this disease especially because of its protean nature.

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