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A Rare Case Report of Acute Intermittent Porphyria Complicating Pregnancy

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ABSTRACT

The porphyrias are a group of metabolic disorders due to deficiency of one of the eight enzymes involved in porphyrin-haem biosynthetic pathway and are more common in women than men. Acute intermittent porphyria is the commonest type. It is therefore important for the obstetrician to know about this entity as many women present with abdominal pain during pregnancy. It is a challenge for the obstetrician as few of the drugs commonly used in labour management is contraindicated in these patients. We hereby present a case of acute intermittent porphyria which was complicated by IUGR and abnormal Doppler. She underwent emergency LSCS with good maternal and fetal outcome.

KEYWORDS Pregnancy, Porphyria

INTRODUCTION

Porphyrias comprise a heterogenous group of rare, mainly hereditary diseases caused by partial deficiency in one of the eight enzymes involved in haem biosynthesis.Porphyria and effect on pregnancy has seldom been investigated, and the body of evidence only consists of case reports and small case series which shows the association with low birth weight, preterm delivery, preeclampsia, congenital malformations and perinatal deaths.

CASE REPORT

23 year old primigravida with 36 weeks 5 days of gestation, previously diagnosed to have acute intermittent porphyria was referred to our institution following detection of IUGR, oligamnios, abnormal Doppler and preeclampsia.

She presented with acute onset of severe abdominal pain and vomiting along with brownish discoloration of urine. On examination, pallor was present. She had pedal edema and blood pressure of 160/100. Cardiovascular, respiratory and nervous system examination was normal. Abdominal examination revealed fundal height of 30 weeks with clinically decreased liqour, cephalic presentation with good fetal heart rate. Abdomen was soft and without peritoneal signs. On vaginal examination, cervix was unfavorable.

Her lab reports revealed urine albumin of 3plus and presence of porphobilinogen.Blood investigations was suggestive of anemia and hyponetremia (Hb 9g/dl,Sodium 120meq/l).Renal and liver function tests were normal. Obstetric Ultrasound was suggestive of IUGR (AC 29 weeks, EFW 1.2kg), oligamnios (AFI 3) and reversed diastolic flow on umbilical artery Doppler. NST was pathological.

Emergency physician consultation was done and patient was managed symptomatically with opiate analgesics and antiemetics. Hyponatremia was corrected with normal saline infusion. Emergency LSCS was done in view of IUGR and fetal distress, delivered male baby of weight 1.39kg with Apgar 7.Intraop and post operative period was uneventful. Baby was admitted in NICU in view of low birth weight and was discharged on 11th post operative day.

DISCUSSION

Acute intermittent porphyria results from half normal activity of the enzyme uroporphyrinogen 1 synthase.It is charecterised clinically by acute neurovisceral attacks of severe abdominal pain without peritoneal signs, often accompanied by nausea,vomiting,tachycardia and hypertension. Attacks may be complicated by convulsions and hyponatremia.Acute attack may be provoked by certain drugs, alcohol, endocrine factors, calorie restriction, stress and infection. Acute attack is associated with increased urinary concentration of porphobilinogen. Exclusion of other porphyrias are done by analysis of porphyrin in stool and plasma. Mild attacks are managed by symptomatic treatment, increased calorie intake and fluid replacement. Specific treatment is by injection of human haemin.

The incidence of acute porphyria attack is increased in pregnancy and both estrogen and progesterone are known to provoke attack in non pregnant women. Acute intermittent porphyria, variegate porphyria and hereditary coproporphyria are of significance in pregnancy. Each of these may present acutely with abdominal pain, fever and leucocytosis which may mimic other intraabdominal conditions. A review in 1971(221) described 72 cases of acute porphyria in pregnancy with a 27% mortality rate. Another study of 50 women with acute porphyria recorded only 1 death with a fetal loss rate 13%. More than half of women with acute intermittent porphyria had an attack during pregnancy and the babies from these pregnancies weigh 800grams less than their normal counterparts.

Erythromycin,frusemide,halothane,barbiturates,clonidine,hydralazine,methyl dopa,lidocaine,metoclopramide,metronidazole,oral contraceptive pills,pancuronium,phenytoin,ranitidine,sulphonamides and ergot derivatives can precipitate an acute attack. These drugs are commonly used in obstetric practice. Hence it is important that the treating obstetrician be aware of these contraindications.

CONCLUSION

Awareness of this condition can minimize misinterpretation, misdiagnosis and unnecessary operative procedures. It is important to prevent the onset of an acute attack by avoiding the precipitating factors, especially drugs known to precipitate an attack.

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