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CASE REPORT

Intrahepatic Cholestasis of Pregnancy: A Serious but Underestimated Problem

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ABSTRACT

Intrahepatic cholestasis of pregnancy (IHCP) is the most common liver disorder unique to pregnancy. In spite of the substantial risk, IHCP remains widely disregarded as a serious clinical problem. We report a case of IHCP with deranged coagulation which resulted in maternal morbidity in the form of severe postpartum hemorrhage and fetal mortality. This case emphasizes the fact that IHCP should be considered a high-risk condition, and careful fetal assessment and appropriate medical intervention might—improve maternal and perinatal outcome.

Keywords: Intrahepatic cholestasis, Pregnancy, Coagulopathy derangement.

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INTRODUCTION

Intrahepatic cholestasis of pregnancy (IHCP) is an under-recognized entity in routine clinical practice. The incidence of IHCP varies throughout the world. The highest incidence is considered to be in Chile and Bolivia (5-15%), while in other countries of Europe and North America the incidence is less than 1%.[1] No reliable statistics for Indian scenario is available. The usual manifestation of the condition is associated with increased perinatal morbidity and mortality and although coagulation derangement in the mother is recognized as an entity but its actual incidence is extremely rare.

We report a case of IHCP, which presented primarily with coagulation derangement and resulted in increased morbidity in the mother and mortality in fetus.

CASE REPORT

A 26-year-old unbooked G4P1L1A2 presented in emergency with 38 weeks gestation, extreme yellow discoloration of eyes and body since last 3 days, generalized itching all over body for last 1 week. She received irregular antenatal care at a private clinic from where she was referred to our hospital with above complaints.

On examination patient was found to be severely icteric, with no hepatosplenomegaly. Her obstetrics examination revealed a term size relaxed uterus with estimated fetal weight of 3,200 gm. Patient was not in active labor. Investigations done at that time were as follows: Hemoglobin—9.5 gm%, total bilirubin—11.7 mg% with mixed components, blood group—AB positive, HBSAG, HCV, HEV and HAV were negative. Coagulation studies showed deranged prothrombin time (PT-38 seconds, PT control—10.8 second, INR—3.5), platelet count was 1.5 lacs, bleeding time—1 minute 5 seconds, clotting time—5 minutes, ALT—72 IU/l, AST—231IU/l and serum alkaline phosphatase—654 IU/l. Total bile acids were sent for assessment from external agency as the hospital laboratory does not offer the test.

In view of deranged PT 4 units of fresh frozen plasma were transfused with supportive treatment and injection vitamin K 10 mg given. Later she went into labor and 9 hours after admission she was found to be fully dilated with absent fetal heart rate. Efforts to deliver her spontaneously failed and she was delivered by ventouse application without episiotomy. Baby was 3,500 gm, freshly still born with uniform meconium staining of fetal skin, cord and placenta.

Patient had severe postpartum hemorrhage (PPH) in the form of continuous trickling of blood in presence of well-contracted uterus. She was transfused 2 units of packed RBCs and 4 more units of fresh frozen plasma. Serum bile acids report was collected on 3rd postnatal day which were 109.90 micromol/l (normal value 0.0-10.0 micromol/l). A marked increase in total bile acids confirmed the diagnosis of IHCP.

DISCUSSION

Intrahepatic cholestasis of pregnancy is characterized by pruritus, jaundice and biochemical disturbances in liver tests. IHCP predominantly occurs in the third-trimester of pregnancy, resolve after delivery and recur in subsequent pregnancies.

The most sensitive laboratory abnormality in IHCP is an increase in serum total bile acid concentrations.[2] During a 3 years period in a prospective study of 84 women with IHCP, elevation of aminotransferase activities from 2 to 15-fold were noticed in 85% of patients, bilirubin concentration from 2 to 4-fold in 14%, fasting serum bile acids from 1.5 to 20-fold in 78% and alkaline phosphatase upto 2 to 3-fold in 60% of patients.[3] The PT is usually normal. When present, prolonged prothrombin time denotes vitamin K deficiency. In our patient serum aminotransferase, bilirubin concentration, alkaline phosphatase, serum bile acids and PT levels were significantly high.

In IHCP maternal prognosis is favorable.[2] Pruritus usually disappears in the first few days following delivery, accompanied by normalization of serum bile acid concentrations and other liver tests. PPH is reported in 8 to 22% of cases following delivery. Shukla Chhavi et al[4] reported an unusual case of IHCP with severe jaundice and prolonged PT which resulted in severe PPH necessitating hysterectomy.

In contrast to favorable prognosis for mothers, IHCP poses significant risk for fetus. The major complications are premature deliveries, still births, fetal distress and meconium staining. Zimmermann P et al[5] have reported that fetuses of women with

IHCP have adequate birth weights for gestational age and normal Doppler umbilical artery velocimetry suggesting that chronic placental insufficiency is not the primary cause of fetal death. In a study by Glantz A et al[6] a correlation between fetal complications and serum bile acid levels (≥40 micromol/l) was found. The authors reported no increase in fetal risk detected in IHCP patients with total bile acids levels <40 micromol/l, and proposed that these women can be managed expectantly. However, further validation of these results is needed to determine the accuracy of this cutoff value. Sentilhes L et al[7] have reported a case of fetal death at 39 weeks and 3 days in a patient with IHCP, who had low serum bile acid concentrations at the time of diagnosis.

In our case the baby was well grown (birth weight, 3500 gm) with meconium staining of skin, cord and placenta. Our patient came for follow-up 6 weeks after delivery. She had no jaundice and pruritus and her liver function tests were found to be normal.

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