

Case Report

Pregnancy with Wilson's disease complicated with thrombocytopenia: a case report

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ABSTRACT

Wilson's disease in pregnancy is rare. It manifests in the form of liver and neurological disorder due to abnormal copper accumulation. Pregnancy becomes high risk due to involvement of liver and high incidence of abortion, preeclampsia, HELLP syndrome. High risk pregnancy management and treatment with zinc sulphate have shown successful pregnancy outcomes. Reporting here a case of pregnancy with Wilson's complicated with thrombocytopenia managed successfully.

Keywords: Pregnancy, Wilson's disease, Thrombocytopenia

INTRODUCTION

Wilson's disease is an inherited disorder of copper homeostasis first described in 1912. The Wilson's disease gene was discovered in 1993, with the identification of ATP7B. This P-type ATPase is involved in copper transport and is necessary for the export of copper from the hepatocyte. Thus, in patients with mutations in ATP7B, copper is retained in the liver, leading to increased copper storage.¹ This abnormal accumulation leads to liver cirrhosis and neurological defects like movement disorders and ataxia.

Though it is associated with subfertility, with improvement in therapeutic options, many patients are conceiving and having successful pregnancy outcome. The reported prevalence is 1:50000-1:100000 live births.²

Due to involvement of liver and brain, pregnancy becomes high risk and is associated preeclampsia, thrombocytopenia and deranged coagulation. Reporting

here a case of pregnancy with Wilson's disease complicated with thrombocytopenia without preeclampsia managed successfully.

CASE REPORT

A 28 year old second gravida with previous caesarean section presented in antenatal OPD in second trimester for routine antenatal examination. She was a diagnosed case of Wilson's disease two years back when she had developed neurological symptoms in the form of ataxia. She was taking tablet zinc sulphate 30 mg thrice a day as advised by physician.

Her previous caesarean section was done 4 years back for contracted pelvis with no other maternal or neonatal complication. During her initial presentation, the patient complained of slurring of speech and tremors, for which the neurologist was consulted. The dose of zinc sulphate was increased to 50 mg thrice a day after which the neurological symptoms subsided gradually.

Ophthalmologic examination revealed classical Kayser-Fleischer rings as shown in Figure 1. Serum copper and ceruloplasmin levels were then obtained which were 46.2 µg/dl (118-302 µg/dl) and 0.04 g/L (0.25-0.63 g/L), respectively.

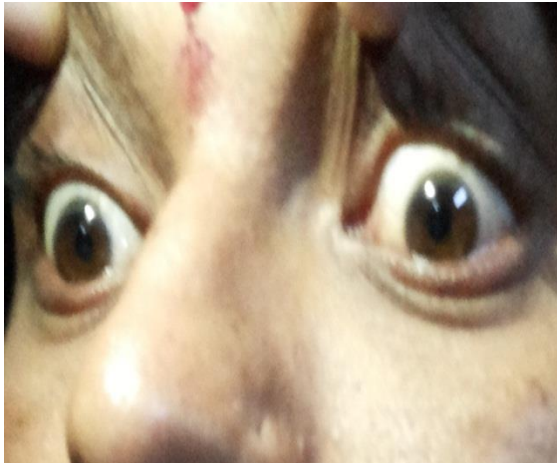


Figure 1: Kayser-Fleischer ring.

Ultrasound of abdomen along with obstetric sonogram was normal.

Renal and liver function tests remained within normal limits throughout the pregnancy. Serum copper level was again obtained at 37 weeks of gestation was 40.12 µg/dl and serum ceruloplasmin level was 0.04 g/L.

Antenatally all investigations which were done at monthly interval were within normal limits. Though patient was posted for elective caesarean section in view of previous caesarean section and contracted pelvis, at 37 weeks patient went into labour. She was taken for emergency caesarean section. Liver function tests and haematocrit and coagulation profile were repeated which revealed normal reports except thrombocytopenia (platelet count 40000/cu.mm). Patient was transfused with 6 units of platelets pre and intraoperatively. Caesarean section with bilateral tubal ligation was done under regional anaesthesia. Intraoperative blood loss was average and there was no post-partum haemorrhage. A 2.8 kg male baby was born with normal APGAR score and had normal neonatal period.

Post-operative period was uneventful. Due to movement disorder patient needed assistance for breast feeding the baby. Specific treatment with zinc sulphate was continued. Patient was discharged on eighth post-operative day after suture removal.

DISCUSSION

Wilson's disease affects mainly liver and nervous system, leading to various neurological symptoms gradually or rapidly leading to disability. It affects liver function with symptoms similar to acute hepatitis.

In our case it was diagnosed prepregnancy. There was presence of classical Kayser-Fleischer rings and a low serum ceruloplasmin. Antenatal period in the first and second trimester was uneventful. As found in one of the case series by Ayesha et al, in our case too there was worsening of neurological symptoms in third trimester for which zinc sulphate dose was increased.^{3,4} Thrombocytopenia as a component of HELLP syndrome has been reported by Członkowska A et al.⁵ Thrombocytopenia as a single complication of Wilson's disease without preeclampsia is not reported in literature which was unique in our case.

Pregnancy with Wilson's disease becomes high risk as it is associated with high incidence of abortions, preeclampsia, jaundice, deranged coagulation. Zinc sulphate is a treatment of choice in pregnancy for maintaining the copper and ceruloplasmin level. Zinc performs its function by induction of intestinal cells metallothionein which has a high affinity for copper and prevents serosal transfer of copper into blood. Copper status should be optimized prior to pregnancy. Reducing the zinc sulphate to a minimal dose, i.e. 30 to 50 mg/day in the last trimester avoids insufficient copper supply to the foetus or insufficient wound healing after caesarean section or episiotomy.

Brewer et al. report use of zinc sulphate in 26 pregnancies out of which 24 had a normal infant, one infant had a congenital heart defect and one had microcephaly.⁶ In our case patient was treated with zinc sulphate there was no evidence of any congenital anomaly in the neonate.

Postnatally patient required assistance in baby handling and breast feeding due to ataxia.

Estrogens may interfere with biliary copper excretion and intrauterine devices contain copper. Thus, mechanical or chemical barrier contraceptives and progesterone-only preparations can be safely prescribed for contraception. In our case tubal ligation was during caesarean section.

As it is a genetic disorder genetic screening and counseling should be offered and screening for disease in partner is justified.

CONCLUSION

With improved treatment options patients with Wilson's disease are able to conceive. Antenatally the woman is at high risk of abortions, preeclampsia, and complication caused by liver dysfunction. Zinc sulphate controls the copper levels in pregnancy and safe in first trimester also.

As found in our patient WD can be associated with thrombocytopenia or in undiagnosed case differential diagnosis of Wilson's disease should be kept in mind while managing HELLP like syndrome. With improved treatment options and high risk pregnancy management,

Wilson's disease patients can have successful pregnancy outcome.

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