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Case Report

PREGNANCY WITH AUTOIMMUNE HEPATITIS – A THREAT TO MOTHER AND FETUS

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INTRODUCTION

Autoimmune hepatitis (AIH) is a disorder of unknown etiology in which there is progressive destruction of hepatic parenchyma, frequently leading to cirrhosis. This liver disease predominantly affects women of all ages and can manifest at any time during gestation and postpartum period. Diagnosis and the treatment of autoimmune hepatitis remains challenging. Diagnostic criteria are based on elevation of IgG, demonstration of characteristic autoantibodies and histological features of hepatitis in the absence of viral disease. Autoandibodies are the hallmark of autoimmune hepatitis. Adequate dose of steroids are the mainstay of remission induction treatment , while remission maintenance is best achieved by azathioprine. The disease activity of AIH is usually attenuated during pregnancy because of the immune tolerance induced by the pregnancy. There are increased risk of obstetrics complications , including toxaemia of pregnancy, preterm delivery, low birth weight infant and fetal loss. Both prednisone and azathioprine are considered safe during pregnancy and lactation. We report this case because of the challenging efforts taken to manage this case of Autoimmune hepatitis type- 1 with cirrhosis and portal hypertension.

CASES

We describe here a 28 year old female , second gravida with previous history of one abortion, presented to out patient department of SGPGIMS , Lucknow with 28 week pregnancy. She was a diagnosed case of autoimmune hepatitis (type – I) with cirrhosis, portal hypertension and hyperspleenism and she was on prednisolone . On clinical examination she was corresponding with the period of gestation with mild pallor and dependent edema. Blood investigation showed deranged liver function with thrombocytopenia and hypoproteinemia. Ultrasonography of abdomen revealed normal fetal growth with maternal hepatospleenomegaly. The upper gastrointestinal endoscopy showed Grade I esophageal varices (Figure - 1). All viral markers were negative . Her serology were positive for anti smooth muscle antibody (ASMA) and anti double stranded DNA (ds DNA). Anti nuclear antibody and antiphospholipid antibody were negative. With the help of gastroenterologist and immunologist , she was managed till 36 week of gestation without any further complication with close vigilance on her liver function , platelet count and fetal growth. Initially she was on high dose of prednisolone (60mg/day) which was later reduced to 20 mg/day. She developed sever preeclapmtic toxaemia at 36 week of pregnancy. Termination of pregnancy by emergency lower section cesarean section was done as her bishop score was poor. She delivered a 2.5 kg female baby with good apgar. Peroperative patient had primary post partum hemorrhage which was managed medically. Her blood pressure remained on higher side which was treated with nitroglycerine infusion. In postoperative period, 10 hours after delivery, she developed postpartum eclampsia which responded to magnesium sulfate regimen. Patient required eight units of platelet transfusion. She was discharged on 9th postoperative day on steroid and antihypertensive drugs and was counseled for contraception possibility of and liver transplant in future.

DISCUSSION

Autoimune hepatitis (AIH) carries all features of an autoimmune disease: genetic predisposition, association with other autoimmune disease,spontaneous disease fluctuations, autoantibodies,and auto – reative T cells, inflammatory infiltrate, and a good response to imunosuppression (Lohse and Vergani

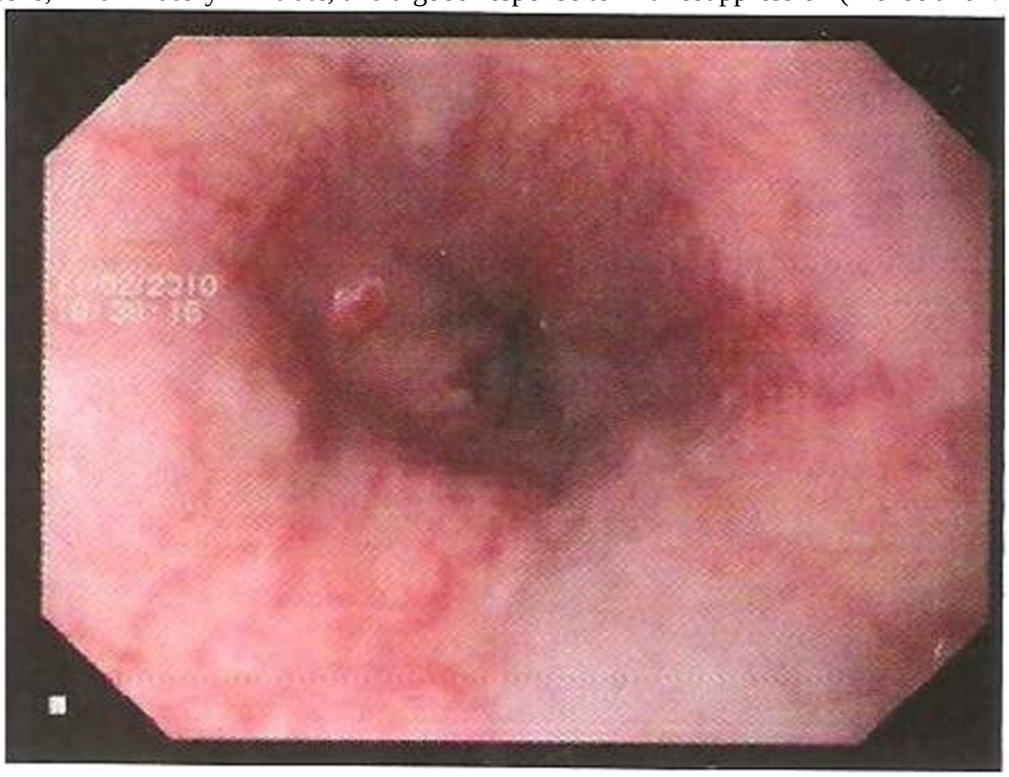

Figure 1: Upper GI endoscopy showing Grade I esophageal varices

2011). Liver is an organ of immunological tolerance due to several regulatory mechanism, breaking of self tolerance in the liver may therefore be more difficult than any other organs.This acconts for the rarity of the disease.

A female predilection has been confirmed in almost all studies with a female to male ratio of around 3:1 (Lohse and Vergani 2011). Reduced fertility has been associated with AIH .However the risk of an unsuccessful pregnancy has been reported to be high , a fetal death rate of 50% in one series (Heneghan et al. 2001). In one of yhe largest study fetal loss was high (23%) due to spontaneous abortion (Heneghan et al. 2001). Our case has one spontaneous abortion prior to the pregnancy. The activity usually attenuated during pregnancy and doses of medication can be decreased because of the state of immune tolerance .nonetheless flares have occurred in 11% during pregnancy and upto 25% in postpartum period (Fleming and Zein 2005).

Disease flares are associated with poor disease control prior to pregnancy , associated portal hypertension and absence of therapy (Westbrook et al. 2012). Patients with Cirrhosis and portal hypertension with pregnancy are at increased risk for obstetrical complications like preterm deliveries, intrauterine growth retardation, fetal death , toxaemiam of pregnancy. Hepatic decompensation with jaundice, bleeding from esophageal varices, ascitis and fulminant hepatic failure can occure. Our patient developed eminent eclampsia at 36 week of pregnancy. Occasional flares up of disease may occur during pregnancy, but potpartum flares of the disease are frequent and therefore peripartum increased dose of steroid is recommended. We had increased the dose of steroid to 1mg/kg body weight in peripartum period.

Hyperspleenism was an additional complication in our patient which reflected as low platelet counts and lead to coagulopathy in postoperative period.

In one series adverse pregnancy outcome were highly associated with the presence of antibodies to SLA / LP (odds ratio 51; $p < 0.003$) and Ro/SSA (odd ratio 27; $p < 0.02$).. The presence of autoantibodies may be a risk factor for adverse pregnancy outcome in AIH patients (Schramm et al. 2006).

Optimal management during pregnancy of patient with autoimmune hepatitis (AIH) remains undefined. In general diuretics and spironolactone are not advisable, banding of bleeding esophageal varices are safe during pregnancy. Both prednisolone and azathioprine are considered safe during pregnancy and lactation. Aazathioprine is used for its "steroidal sparing" effect in AIH. If discontinuation of azathioprine therapy is contemplated, a compensatory increase in steroid dosage may be required to sustain remission (Heneghan et al. 2001). We followed same policy for our patient and gave prednisolone accordingly during pregnancy and lactation.

Conclusion

Successful completion of pregnancy is a realistic expectation for patients with well controlled AIH. There are no clinical markers that predict the course of pregnancy and the pathophysiologic mechanisms are not always understood,but the knowledge and management of the preconception liver disease and efficacious prepregnancy and prenatal care are essential. Treatment options are mainly steroids and azathioprine which appears to be safe during pregnancy and lactation.

vigilance in recognizing liver disorders in pregnancy and early coordinated management among the primary care physician,obstetrician, gastroenterologist, and transplant surgeon are essential for promoting good maternal and fetal outcome .However patients need to be monitored carefully during pregnancy and for several months postpartum.

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