Liver Infarction in a Patient with HELLP and Probable Anti-phospholipid Syndrome

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The aim of this report is to make an awareness about hepatic infarction in those patients with progressively elevating liver enzymes and anti-phospholipid syndrome. A 29-year-old G2P2 woman who has undergone cesarian section (C-section) due to fetal distress was referred to our department with progressively elevating liver enzymes and lowering platelets. Computed tomography revealed a hepatic infarction with a dimension of 14x10 cm. Further, laboratory parameters defined an elevated Cardiolipin IgM antibody (11.59 U/ml). She had a diagnosis of HELLP and probable anti-phospholipid syndrome due to these findings. She did well with the administered anticoagulant and steroid therapy.

Pregnant women with progressively elevating liver enzymes should be evaluated for hepatic infarction and underlying diseases such as anti-phospholipid syndrome.

Key Words: Anti-phospholipid syndrome, HELLP syndrome, Pregnancy

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Introduction

Criteria for anti-phospholipid syndrome (APS) were proposed at Sydney in 2006 including; i) clinical criterias such as vascular thromboses and pregnancy morbidity with three or more unexplained spontaneous abortions before 10 weeks of gestation, intrauterine fetal death, and one or more premature births of a morphologically normal neonate before 34 weeks of gestation. ii) laboratory criterias such as presence of Lupus anticoagulant (LA) in plasma, Anticardiolipin antibody (aCL) of IgG and/or IgM, Anti-β2-glycoprotein-I antibody isotype in serum or plasma, on two or more occasions, at least 12 weeks apart.[1] The diagnosis is made by presence of at least one of the clinical criteria and one of the laboratory criteria.

APS is associated with microangiopathic features resulting HELLP (hemolysis, elevated liver enzymes, and low platelet count) syndrome in these patients. Hepatic infarction, retinal vascular occlusions, and deep venous thrombosis (DVT) have been reported in patients with HELLP syndrome.[2] We describe a patient who developed hepatic infarction after C-section with HELLP syndrome and probable APS.

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Case Report

A 29-year-old G2P2 woman who has undergone C-section due to fetal distress, was referred to our department in her postoperative first day with laboratory findings reflecting HELLP syndrome. Physical examination was only notable for a blood pressure of 150/80 mmHg. Her laboratory parameters were; aspartate aminotransferase (AST): 820 U/L (normal 0-34 U/L), alanine aminotransferase (ALT): 739 U/L (normal 0-55 U/L), direct bilirubin: 0.65(0.1-0.5), alkaline phosphatase (ALP): 200IU/L (40-150), gamma-glutamyltransferase (GGT): 17 IU/L(9-64), lactate dehydrogenase (LDH): 3498 IU/L (125-243), platelet: 87 103 /mm^3 (normal 150-400 per mm3), D-dimer: 25000 µIU/L (80-500). Prothrombin time, partial thromboplastin time and fibrinogen was normal. AST (2473 U/L) and ALT (2101 U/L) parameters were elevated the day after her internation. A computed tomography scan revealed a hepatic infarction with dimension of 14x10 cm. (Figure 1). Colour Doppler ultrasound was correlated with a hepatic vein trombosis also. Cardiolipin IgG antibody was normal. Cardiolipin IgM antibody was elevated (11.59 U/ml) and Lupus anticoagulant was negative. C4 level was 43.2 mg/dl (10-40). Extensive evaluation for other causes of elevated liver enzymes were negative. With those findings, the patient was diagnosed with probable APS, associated with hepatic infarction and HELLP syndrome. She did well with anticoagulation and a course of steroid therapy. All of the laboratory parameters were within normal limits after her postoperative 8th day. Cardiolipin IgM and IgG antibodies were elevated after the patient's postoperative 3rd month control visit.

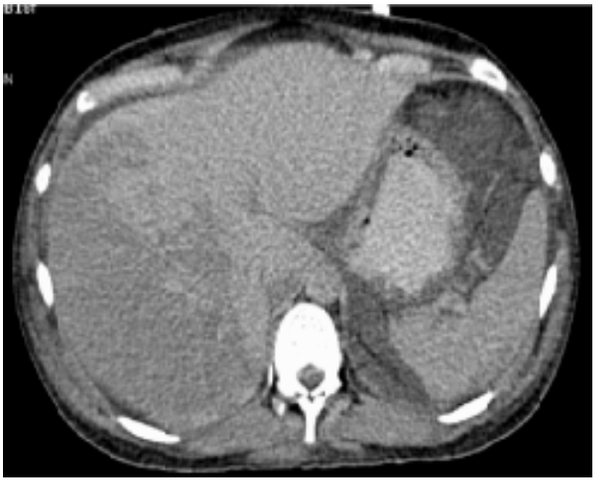

Figure 1: Computed tomography. White arrow showing the liver infarction

Discussion

APS does not have a known etiology. There are several hypotheses suggesting that APS is triggered mainly by infections, trauma, surgery, anticoagulation withdrawal, malignancies, lupus, pregnancy and puerperium.[3] The relationship between liver infarction, HELLP syndrome and APS is that, hepatic infarctions during pregnancy are almost always associated with the antiphospholipid syndrome and in addition with HELLP syndrome. Laboratory parameters such as positive anti-nuclear antibodies (ANA), anti-doublestranded (anti-ds) DNA, immunoglobulin G (IgG) anti-Cardiolipin antibodies (ACA) and lupus anticoagulant (LAC) are used in diagnosis of APS. Recently, determination of anti- β2-glycoprotein I antibodies has been recommended in the diagnosis of APS.[4] In our patient Cardiolipin IgM antibody was elevated suggesting APS After her postoperative 12 weeks IgM was positive again and we confirmed our diagnosis in conjunction with APS criteria. The imaging studies such as Doppler ultrasound and computed tomography may be helpful in diagnosis of hepatic vessel occlusion resulting in hepatic infarction. In our case both of these imaging studies facilitated the diagnosis of APS also.

In the treatment, women with anti-phospholipid antibodies and a history of 2 or more early pregnancy losses or 1 or more late pregnancy losses who have no prior history of thrombosis should receive a combination of aspirin and heparin (unfractionated or low molecular-weight) during pregnancy.[5] Aspirin and low-dose heparin or low-molecular weight heparin combined therapy is effective in 70-80% of patients with APS.[6] Our present case was treated with low molecular-weight heparins and steroids and her vital and laboratory findings did well after her postoperative 8th day.

Pregnant women with progressively elevating liver enzymes should be evaluated for hepatic infarction and underlying diseases such as APS. Preconceptional counseling also gives the physician the opportunity of managing a safe pregnancy in patient with history of APS.

HELLP ve Muhtemel Anti-Fosfolipid Sendromu Olan Bir Hastada Karaciğer Enfarktı

Anti-fosfolipid sendromu olan ve progresif olarak karaciğer enzimleri artan hastalarda karaciğer enfarktı olabileceği hakkında farkındalık yaratmayı amaçladık.

Yirmidokuz yaşında G2P2 olan fetal distress nedeniyle sezaryen olmuş hasta obstetri kliniğimize progresif olarak artan karaciğer enzimleri ve düşük platelet sayısı nedeniyle yönlendirildi. Bilgisayarlı tomografide 14x10 cm boyutunda karaciğer enfarktı saptandı. Sonraki laboratuvar incelemelerinde yüksek Kardiolipin IgM antikoru (11.59 U/ml) saptandı. Bu sonuçlara göre HELLP sendromu ve muhtemel anti-fosfolipid sendromu tanıları kondu. Antikoagülan ve steroid tedavileri sonrasında hasta sorunsuz iyileşti.

Progresif olarak artan karaciğer enzimleri olan gebe kadınlar karaciğer enfarktı ve altta yatan anti-fosfolipid sendromu gibi hastalıklar açısından değerlendirilmelidirler.

Anahtar Kelimeler: Anti-fosfolipid sendromu, HELLP sendromu, Gebelik

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