HELLP Syndrome, Acute Fatty Liver of Pregnancy, and Fatty Acids Oxidation Defects.

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ABSTRACT

Introduction: HELLP syndrome is one of very common and important complications, and is prevalent during 3rd trimester of pregnancies. Clinical characteristics are; acute micro-angiopathic hemolysis, Elevated liver enzymes, and thrombocytopenia. Its etiology is unknown but the concordance of disease with the common complications of pregnancy such as pre-eclampsy and eclampsy have been reported. Its incidence is about 1 percent of all pregnancies. HELLP syndrome is a severe and progressive disease with high morbidity and mortality rate. Acute fatty liver of pregnancy (AFLP) also is a severe but less common complications. Morbidity and mortality of this disease is also high and cardinal findings are; abdominal pain, nausea, vomiting, and rapidly progress to liver failure, bleeding and hepatic encephalopathy. Fatty acids oxidation defects (FAODs) are a group of genetic-metabolic disorders that affected patients are not able metabolize fatty acids. They transmitted by autosomal recessive inheritance. Carrier mothers for these defects are at higher risk for HELLP syndrome and AFLP.

Cases Presentation: A 33-year-old pregnant mother was admitted to hospital at 22nd gestational week. The symptoms were abdominal pain, headache, visual impairment, vomiting and a seizure attack. Clinical findings were; agitation, disorientation, jaundice, high blood pressure and proteinuria. She was suspected to HELLP syndrome biochemical investigations showed; high LDH, high SGOT, and low platelet count. After 24 hour of treatment the fetus was aborted. Metabolic screening of the mother was suggestive for fatty acid oxidation defect.

Conclusion: Heterozygote pregnant women with FAODs as autosomal recessive diseases, might lead to the important & relatively common complications like; severe pre-eclampsia, AFLP & HELLP syndrome. So, it should be considered in at risk pregnancies.

Keywords: HELLP syndrome, acute fatty liver of pregnancy, fatty acid oxidation defects.

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