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How to Differentiate Acute Fatty Liver in Pregnancy (AFLP) with Hemolysis, Elevated Liver Enzymes, and Low Platelets (HELLP) Syndrome in Resource-Limited Settings? – The Importance of Swansea Criteria

Dear Editor,

We present a rare case of acute fatty liver in pregnancy (AFLP) which sometimes can be clinically overlapped with hemolysis, elevated liver enzymes, and low platelets (HELLP) syndrome in pregnancy. Both of them are accounted as liver diseases in pregnancy that may be accompanied by severe consequences and associated with high maternal and fetal mortality [1,2]. They should be differentiated to make the correct and proper diagnosis, especially in resource-limited settings such as in this case.

A 36-year-old multiparous woman came to Obstetrics-Gynecology Emergency Ward as referred by Primary Health Care due to shortness of breath. She previously had a vaginal delivery assisted by a midwife in 36–37 gestational weeks. She complained of abdominal pain since after delivery. The patient had no previous history of high blood pressure. The patient was somnolent with a blood pressure of 130/90 mmHg, pulse rate 80 times/min, respiration rate 45 times/minute, and temperature of 36 °C. Her proteinuria test was + 3. Her total leukocyte level was 68 $\times$ 10^9/L and thrombocytopenia (61 $\times$ 10^9/L); total bilirubin was 205.2 μmol/L; blood glucose was 3 mmol/L; serum creatinine level was 176.84 μmol/L; AST and ALT serum levels were 4700 IU/L and 3129 IU/L; prothrombin time (PT) was 16 s and activated partial thromboplastin time (APTT) was 40 s.

AFLP and HELLP syndrome have similar clinical, laboratory, histological and genetic features, making a differential diagnosis between them often difficult. The main difference between the two diseases can be seen in the clotting and coagulation ability as well as platelet levels. AFLP usually causes more abnormal coagulation and clotting problems. Patients with HELLP syndrome tend to have a more progressive decrease in platelet count, which can be measured over time [2,3]. The differences may be due to differences in etiopathogenesis underlying two syndromes – AFLP develops due to a deficiency of fatty acid oxidizing enzymes while HELLP syndrome is caused by endothelial injury leading to periportal hepatic edema, hemorrhage, and necrosis [4].

The diagnosis of AFLP can be made based on clinical manifestations and laboratory results, whereas a liver biopsy is not required. This is very helpful, especially in the resource-limited area. The Swansea criteria proposed by Ch'ng et al. came up as a screening tool and predictor of AFLP severity and showed an appropriate sensitivity [4,5]. Based on the pathophysiological aspect, AFLP is exclusively associated with pregnancy-induced antithrombin deficiency (PIATD) [2]. HELLP Syndrome is diagnosed with Mississippi Classifications which divides HELLP syndrome into 3 classes based on platelet count, AST or ALT levels, and LDH levels [1].

In our case, the patient was diagnosed with postpartum AFLP since the Swansea score was 7 (Table 1**).** She could be overlapped with moderate HELLP syndrome but since AFLP usually causes more abnormal coagulation and clotting problems and no progressive decrease in platelet count did not take place, she was diagnosed promptly with AFLP. She delivered a female baby girl by vaginal delivery with a birth weight of 3200 g and an APGAR 6 and 8 in 1 min and 5 min, respectively. Formula milk was delivered by orogastric tube. The mother was given supportive treatments such as packs of blood transfusion, cryoprecipitate, and fresh frozen plasma based on her conditions with intensive and regular monitoring for two consecutive days. Follow-up results showed that her vital signs were getting worse over time and no progressive decrease in platelet count was found. The mother had a cardiac arrest that sent her to death after resuscitation on the second day of hospitalization. Her baby was monitored regularly by pediatricians. No complications were found in her baby and she was discharged. No specific treatment was given to her baby.

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Table 1
Swansea criteria vs Mississippi criteria of HELLP syndrome in this patient.

Swansea criteria (≥ 6)	Mississippi criteria		
Vomiting	**Class 1**	**Class 2**	**Class 3**
Abdominal Pain	**(Severe)**	**(Moderate)**	**(Mild)**
Polydipsia/polyuria	≤ 50,000/μL	50,000–100,000/μL	100,000–150,000/μL
Encephalopathy	≥ 70 IU/L	≥ 70 IU/L	≥ 40 IU/L
Elevated bilirubin (> 14 μmol/L/> 8 mg/dL)	≥ 600 IU/L	≥ 600 IU/L	≥ 600 IU/L
Hypoglycemia (< 4 mmol/L/< 72 mg/dL)			
Elevated urate level (> 340 μmol/L/> 950 mg/dL)			
Leukocytosis (> 11 × 10^9/L)			
Ascites or bright liver on ultrasound scan			
Elevated transaminase (AST or ALT > 42 IU/L)	13 %	8 %	No increased risk
Elevated ammonia (> 47 μmol/L)			
Renal impairment (Creatinine > 150 μmol/L/1.7 mg/dL)			
Coagulopathy (*prothrombin time* > 14 s or *activated partial thromboplastin time* >34 s)			
Microvesicular steatosis on liver biopsy			

Notes: The patient was diagnosed with postpartum AFLP since the Swansea score was 7 based on the available results. *LDH was not included in this patient due to resource-limited settings. She could also be diagnosed with moderate HELLP syndrome but since AFLP usually causes more abnormal coagulation and clotting problems and no progressive decrease in platelet count did not take place, she was diagnosed promptly with AFLP.

Ethical approval

This study does not require an ethical approval as determined by the institutional and departmental review board.

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Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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