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Acute fatty liver of pregnancy: An unusual post-partum presentation

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Abstract
Introduction: In obstetrics, any low risk women can suddenly develop multiorgan dysfunction and pose diagnostic challenge. Here is reported a case with similar presentation and major clinical implications.
Material and Methods: A case of a booked low risk primigravida who presented with preterm pains, underwent an uneventful caesarean for fetal bradycardia, developed postpartum sudden catastrophic multiorgan failure and was rescued with supportive measures is reported.
Results: The case highlights the approach to diagnosis of acute fatty liver of pregnancy (AFLP) as a diagnosis of exclusion on retrospective analysis. The details of clinical course, management, differential diagnosis of AFLP, a rare and life-threatening complication of pregnancy is discussed.
Conclusion: The third trimester nausea, vomiting, abdominal pain, or preterm labor can be a potential presentation of AFLP and should be treated cautiously. With watchful alertness to earliest derangements of vitals, a timely and anticipatory involvement of multidisciplinary and tertiary level intensive care team can be done which is lifesaving in such cases with catastrophic multi organ dysfunction.

Keywords: acute fatty liver of pregnancy, multi organ dysfunction, coagulopathy, HELLP syndrome, hepatorenal failure, disseminated intravascular coagulation

Introduction
Maternal multiorgan dysfunction (MOD) is a rare but life-threatening event with a wide-ranging etiology. The purpose of this case report is to discuss the diagnostic approach for a woman presenting as postpartum sudden MOD with diagnosis made by exclusion on retrospective analysis.

Case report
A nineteen years primigravida who had regular antenatal care presented with preterm pains at 33 weeks and was hospitalized for supportive care. On admission, her baseline investigations and ultrasound were normal, with hemoglobin of 11.5gm%, leucocytes of 7800, SGPT 65miu/ml, creatinine 1.0 gm%. Despite the treatment, her contractions persisted and after 24 hours, she had leaking followed by fetal bradycardia for which cesarean section was done with an uneventful intra-operative period and a male 2kg baby with APGAR of 8 was delivered.
Postoperatively, she had good recovery with return of bowel functions and urine output of 100ml per hour till 18th postoperative hour. Then, she complained of pain in whole abdomen for which she was reassessed to have normal wound site and bowel sounds in soft abdomen. Her vitals, abdomen ultrasound and blood counts at this time were normal, then she suddenly showed anuria, refractory to fluid challenge, over next 2 hours. Immediate transfer to intensive care unit in a tertiary center was done with involvement of intensivist. She showed a rapid and catastrophic worsening to cardio-respiratory collapse, DIC, encephalopathy, hepatorenal failure immediately after shifting to ICU. The critical care of the multi organ dysfunction was managed by multi- disciplinary team of nephrologists, intensivist, gastroenterologists, surgeon, hematologist and chest physician with institution of prompt resuscitation with ventilator, ionotrops, dialysis, blood components, broad spectrum antibiotics and total parentral nutrition. She stayed afebrile throughout her course of illness and had repeated severe hypoglycemic episodes.

She had a gradual and steady hemodynamic recovery with total 74 units blood components transfusion over 7 days. She had moderate ascites with rising hyperbilirubinemia reaching to a steady peak of 21mg/ml lasting for 7 days. She required hemodialysis for 21 days then showed gradual recovery of renal functions followed by hepatic functions. She was finally discharged on 40th day of surgery.

The baby was under observation at nursery, stayed well and on 3rd day was discharged.

Her blood picture at the time of shock (Table 2), showed severe coagulopathy, uremia, elevated creatinine, modest elevation of hepatic enzymes, severe hypoglycemia, acidosis. Meanwhile, all cultures from blood, wound, vagina and urine were negative with normal leukocyte counts and body temperature.

On logical and retrospective analysis of complete clinical course, the multidisciplinary team ruled out all the differential diagnosis and AFLP was diagnosed as a diagnosis of exclusion.

Table 1: Investigation profile of the case reported

Lscs day	On Admission (1 day before Lscs)	Day 1	Day 2 18-hour post lscs	Day 2 22-hour post lscs (sudden collapse)	DAY 3	DAY 4	DAY 5
Hb GMS%	10.5	10.8	9.9	7.7	8.3	7.5	8.6
Wbc PER Cu.MM	7800	9400	6500	5800	7.2K	31.2	17.8
Platelets/cu.mm	2.34	2.10	1.70	80,700	38000	37000	33000
SGPT IU/ DL	75			250		102	
Creatinine mg/DL	1.1			3.37	4.8	5.4	
RBS MG/ dl	98			35	103	145	92
Bilirubin mg/DL				3.1	8.4	11.5	
PT sec				23.8	26	17.3	15.5
INR				2.04	2.29	1.35	1.17
APTT sec				37.3	37	36.8	34
Fibrinogen mg/dl				143		434	
SGOT Mg/dl				557			
Urea mg/dl				75			
Total proteins				4.33			
A/G				0.88			
LDH				2250			
HIV/HBSAG/HCV				Non-Reactive			

Table 2: The Swansea Criterion [5] - The reported case had 9/15 criteria present namely abdominal pain, elevated urea, creatinine, ALT, ammonia, with Coagulopathy, encephalopathy, ascites and hypoglycemia

Vomiting
Abdominal pain
Polydipsia/polyuria
Encephalopathy
Bilirubin>0.8 mg/dl
Hypoglycemia<72 mg/dl
Elevated urea>950 mg/dl
White blood cell count >11×10⁹/l
Ascites
ALT >42 U/l
Ammonia >66_µmol
AKI or Cr>1.7 mg/dl
Coagulopathy or PT >14 s
"Bright liver" on ultrasound
Microvesicular steatosis on liver biopsy

Legend: A patient positive for at least 6 of the 15 criteria should be considered for a diagnosis of AFLP. However, the Swansea criteria are meant to be applied in cases where no other liver disease of pregnancy—such as HELLP or preeclampsia—has been diagnosed.

Discussion

AFLP is an uncommon but potentially fatal disease unique to pregnancy that typically occurs in the third trimester[1] with incidence of 1:7000–15,000 pregnancies[2].

AFLP typically presents in late third trimester, although it is not always diagnosed prior to delivery. The initial symptoms of AFLP mainly nausea, vomiting, abdominal pain, malaise, and anorexia are nonspecific, atypical and can be overlooked. However, patients may rapidly develop manifestations of acute hepatorenal failure, encephalopathy, coagulopathy or hypoglycemia[3]. This case presented with preterm labor and had an uneventful postpartum phase of eighteen hours followed by a rapid and sudden onset of anuria followed by multiorgan failure.

In AFLP, due to autosomal recessive genetic defects in fetal fatty acid metabolism, unmetabolized long-chain fatty acids can accumulate in maternal blood as toxins with deleterious effects on maternal hepatocytes, with strong predilection with primiparous with a male fetus[4].

In this case, the corticosteroid beclomethasone given for fetal lung maturity and the prompt delivery indicated by fetal bradycardia might have delayed the systemic inflammatory response induced by toxemia.

A presumptive diagnosis of AFLP is usually made clinically based upon the presence of significant hepatic dysfunction, after other potential causes have been excluded. The features disfavoring septic shock were afebrile state, no leukocytosis/leucopenia and negative cultures. HELLP and Hemolytic uremic syndrome was disfavored by absence of hemolysis. Viral /autoimmune or drug induced hepatitis and Dengue hemorrhagic fever was ruled out with absence of fever and specific antigens and only mild rise of SGOP/SGPT.

It is a clinical diagnosis and made by exclusion on retrospective analysis[4]. The Swansea criteria[5] may be used for diagnosis when clinical suspicion is present (Table 2). Rapidity of worsening is clue to diagnosis. Findings on imaging may support the diagnosis. Liver biopsy revealing microvesicular steatosis, the gold standard diagnostic test is invasive and impractical hence the diagnosis is made on clinical grounds.

With early diagnosis, prompt delivery and intensive supportive

care, maternal mortality has decreased from as high as 85% to the current range of 12.5%-18% [6]. Treatment is largely supportive aiming for maternal stabilization and recovery of liver dysfunction[7]. Importantly the reason for the MOD may not initially be obvious, therefore resuscitation augmented later by specific treatments as the diagnosis becomes apparent by a multidisciplinary team can result in good perinatal outcomes. Affected women should be counseled about the possibility of effect on neonate and the recurrence in future pregnancy.

Conclusion

This report highlights that nausea, vomiting, abdominal pain in third trimester or preterm labor can be an early manifestation of AFLP hence watchful alertness is essential. Since any low risk obstetric women can potentially develop sudden multi- organ dysfunction, earliest deviation in vitals should be monitored and managed. In case, vitals or urine output show persistent derangement, a low threshold for tertiary level ICU & multidisciplinary care is recommendable because timely action in anticipation saves life.

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Consent for publication.

Written informed consent was obtained from the patient for publication of this case report.

References

1. Nelson DB, Yost NP, Cunningham FG. Acute fatty liver of pregnancy: clinical outcomes and expected duration of recovery. Am J Obstet Gynecol. 2013; 209:456e1-456e7.
2. Allen AM, Kim WR, Larson JJ *et al.* The epidemiology of liver diseases unique to pregnancy in a us community: a population-based study. Clin Gastroenterol Hepatol. 2016; 14:287-94.
3. Minakami H, Morikawa M, Yamada T *et al*. Differentiation of AFLP from syndrome of hemolysis, elevated liver enzymes and low platelet counts. J Obstet Gynaecol Res. 2014; 40:641-9. [PubMed]
4. Ko H, Yoshida EM. Acute fatty liver of pregnancy. Can J Gastroenterol. 2006; 20(1):25-30. doi: 10.1155/2006/638131. [PMC free article] [PubMed] [CrossRef]
5. Joshi D, James A, Quaglia A, Westbrook RH, Heneghan MA. Liver disease in pregnancy. Lancet. 2010; 375:594-605. [PubMed]
6. Rajasri AG, Srestha R, Mitchell J. AFLP (AFLP)--an overview. J Obstet Gynaecol. 2007; 27:237-40. [PubMed]
7. Al-Husban N, Al-Kuran O, Al Helou A. Postpartum acute fatty liver of pregnancy: a case report. J Med Case Rep. 2018; 12(1):67. Published 2018 Jun 1. doi:10.1186/s13256-018-1593-3