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Acute Fatty Liver of Pregnancy: Case series report

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Abstract

Acute fatty liver of pregnancy, a rare clinical entity demands early diagnosis, immediate delivery of fetus, systematic approach for correction of coagulopathy, renal injury & sepsis along with hemodynamic and ventilatory support till complete recovery. We report a series of 3 AFLP cases, who underwent emergency LSCS.

Case description: The first case was admitted with abdominal pain, vomiting, icterus and pedal edema. Clinical examination was unremarkable with normal blood pressures and no bleeding manifestations externally. After further investigations, LSCS was performed under GA with controlled ventilation and multiple blood products. Postoperatively patient developed acute kidney injury, for which Hemodialysis was instituted and continued for 10 days. Patient discharged following recovery of renal and liver functions after 45 days.

The second case presented with similar clinical profile, had persistent severe lactic acidosis, required higher inotropic support, progressive worsening of liver and renal function tests not amenable to corrective measures leading to mortality.

The third patient presented without coagulopathy in spite of elevated bilirubin and liver enzymes. Prophylactic vitamin K therapy was given and LSCS was performed under spinal anesthesia. Patient had uneventful postoperative course with normal LFT.

Conclusion: Patients with AFLP have significant perioperative mortality and morbidity secondary to renal dysfunction, coagulopathy and massive transfusion related complications. Clinical outcome can be improved by early diagnosis, urgent delivery of fetus and supportive care from multidisciplinary team.

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Introduction

Severe liver function disorders in late pregnancy are extremely dangerous while relatively rare, due to the high risk for the mother and the fetus. Acute fatty liver of pregnancy is a potentially fatal metabolic disorder unique to the third trimester of pregnancy. Incidence of AFLP is 1 in 7,000 to 16,000 pregnancies and is associated with micro vesicular fatty infiltration of the liver, hepatic failure, and encephalopathy.(1) We present case series of Acute Fatty Liver of Pregnancy reported in our institute.

Case 1 was a 22-year-old primagravida, 39 weeks of gestation presented with nausea, vomiting, high colored urine, pedal edema, polyuria and jaundice. Her blood pressure was 110/70 mm Hg and pulse rate was 86/min. Taken for emergency LSCS.

Initial investigations revealed elevated levels of bilirubin, creatinine, ammonia, liver enzymes, and PT/PTT with metabolic acidosis. 4 units of fresh frozen plasmas were transfused prior to induction of anaesthesia. GA was given and prophylactic uterine artery embolization was performed before caesarean section. In view of coagulopathy and metabolic acidosis, patient was shifted to ICU for elective ventilation.

In ICU, AKI was diagnosed and was hemodialysed. Due to coagulopathy, massive non-resolving abdominal hematoma was formed, which was evacuated by laparotomy under GA. Acidosis and persistent coagulopathy resulted in transfusions of multiple blood products and ventilator support for 12 days. Patient was discharged on 45th day.

Case 2 was a 30-year-old woman at 38 weeks of gestation, admitted with nausea, vomiting, yellowish discoloration of skin and eyes, high colored urine and itching. Her blood pressure was 100/70 mm Hg and pulse rate was 96/min.

The pre-operative investigations revealed elevated levels of bilirubin, creatinine and ammonia with severe metabolic acidosis. Initially no clot was formed in PT/PTT. After transfusing 8 units of FFP & 10 units of Cryoprecipitate, patient's INR was 1.7. Caesarean section was performed under general anaesthesia. Patient was shifted to ICU for elective ventilation due to persistent coagulopathy and severe metabolic acidosis.

Patient continued to deteriorate in spite of inotropic support and haemodialysis. Due to persistent metabolic acidosis and coagulopathy refractory secondary to fulminant hepatic failure, patient expired on the 5th POD.

Case 3 was a 33-year-old female at 32 weeks was admitted with complaints of epigastric pain, vomiting and high colored urine. Her blood pressure was 120/70 mm Hg and pulse rate was 98/min. Immediate termination of pregnancy was advised by gastroenterologist along with vitamin K prophylaxis.

The pre-operative investigations revealed elevated levels of bilirubin, liver enzymes with 1.25 INR. Patient underwent emergency caesarean section under spinal anaesthesia and was shifted to ICU for monitoring of coagulation, liver and renal function.

Postoperatively patient was monitored in ICU for 3 days. Inj. Vitamin K was given intramuscularly for 3 days. No Packed Red Blood Cells, Fresh Frozen Plasma, Cryoprecipitate and single donor platelets were

transfused. Patient was shifted to ward on 4th POD and was discharged on 7th POD.

Discussion

Acute fatty liver of pregnancy (AFLP) is a potentially fatal metabolic disorder unique to the third trimester of pregnancy. Diagnosis of AFLP is made by using the Swansea criteria (Table 2). All the three cases reported satisfied Swansea criteria.[2]

The most frequent initial symptoms are nausea or vomiting (approximately 75 percent of patients), abdominal pain (particularly epigastric, 50 percent), malaise, anorexia, and jaundice. Extrahepatic complications can occur. Transient polyuria and polydipsia due to central diabetes insipidus also can be seen; this is thought to be caused by decreased levels of arginine vasopressin secondary to reduced clearance of vasopressinase by the impaired liver. Rare patients develop pancreatitis, which can be severe. Pancreatitis generally becomes apparent only after development of hepatic and renal dysfunction.

In our case series of operative delivery for AFLP, all 3 patients presented with nausea, vomiting jaundice, normal BP and platelet counts at the time of admission. All of the patients fulfilled the Swansea criteria for diagnosis of ALFP. Recovery was faster and uneventful in the patient without coagulopathy compared to patients with coagulopathy, who required multiple blood and component transfusions, renal replacement therapy, mechanical ventilation and inotropic support for prolonged period. Early diagnosis and delivery resulted in good outcome in 2 patients. Supportive therapy for coagulopathy and renal failure facilitated recovery. Persistent coagulopathy, renal failure and worsening of liver function resulted in MODS and mortality in our case series.

Previous case reports[3–6] analyzed showed similar kind of illness with coagulopathy ,renal insufficiency and metabolic acidosis were noted. Fibrinogen, platelets counts and bicarbonate serves as morbidity markers as they directly indicate the liver function. Resolution of the disease is usually spontaneous following prompt delivery, although the need for liver transplantation has been reported in a few cases in the studies we compared.

In our case series, out of 3, one patient who presented later in pregnancy with severe coagulopathy and severe metabolic acidosis died due to development of fulminant hepatic failure. Prompt delivery of fetus warranted a good clinical outcome in a patient whose pregnancy was terminated in earlier gestational age.

Liver and coagulation functions should be carefully evaluated in women in the third trimester of pregnancy who become acutely ill, particularly with otherwise unexplained nausea and vomiting.

In the perioperative period, avoidance of hepatotoxic drugs and strategies to preserve hepatic blood flow is recommended. Correction of coagulopathy, hypoglycemia, metabolic acidosis is warranted. Invasive hemodynamic monitoring and mandatory monitoring of blood sugar and coagulation tests is required.

Table 1

Case no.	Age (yrs.)	Clinical findings	Anesthesia technique	Investigations														outcome
					Hb gm/dl	TC cells/cu.mm	Glucose mg/dl	platelets	Bilirubin mg/dl	BUN mg/dl	Creatinine mg/dl	INR/PTT	Ammonia µmol/l	SGOT/SGPT IU/l	Albumin gms/dl	Alk.phos U/l	Hco3 mmol/L	Fibrinogen mg/dl	
1	22	Nausea, vomiting jaundice, polyuria, and pedal edema	General anesthesia	Pre op	11.6	19500	65	156000	8.8	14	1.8	No clot	144	144/137	2.4	283	15	150	Discharged wit normal biochemica and clinica parameters
				ICU	7.0	19000	75	120000	9.7	27	2.4	2.4/54.8s	146	200/215	2.6	345	26	187	
				Discharge	93	9100	105	343000	4.1	11	0.5	1.34/35.3s	30	33/27	2.7	335	24	233	
2	30	Nausea, vomiting jaundice, and itching	General anesthesia	Pre op	15.7	20800	65	150000	17.93	11	1.8	No clot	46	73/25	3.3	58	7	357	Expired due t fulminant hepati failure
				ICU	9.6	21600	150	60000	16.14	28	2.5	3.08/46.5s	56	150/125	2.8	295	13	176	
				Discharge	11.4	2900	98	90000	14.5	30	2.2	4.86/42.1s	94	234/215	2.7	773	11	51	
3	33	Epigastric pain, vomiting and jaundice.	Spinal anesthesia	Pre Op	10.4	17900	10.4	204000	77	8	0.8	1.25/36	34	56/59	2.7	171	16	576	Discharged
				Discharge	9.9	12000	9.9	250000	100	9	0.7	1.16	18	34/40	3.1	149	20	600	

Conclusion

HELP syndrome is the commonly suspected diagnosis during third trimester, when liver enzymes are elevated. Though uncommon, acute fatty liver of pregnancy should be suspected in pregnant cases presenting with complaints of nonspecific symptoms like nausea, vomiting, abdominal pain along with elevated liver enzymes. While the natural history of the disease is improvement within 24–48 hours of delivery, it is recommended that patients who are critically ill at the time of presentation, who develop complications, or who continue to deteriorate despite emergency delivery, should be managed in the intensive care unit.

Table 2: Swansea criteria

Symptoms	Case 1	Case 2	Case 3
Vomiting	✓	✓	✓
Abdominal pain	✓		✓
Polydipsia/polyuria	✓		
Encephalopathy		✓	
Elevated bilirubin	✓	✓	✓
Hypoglycaemia	✓	✓	✓
Elevated urate		✓	
Leucocytosis			
Ascites or bright liver on ultrasound scan (USS)	✓		✓
Elevated transaminases		✓	
Elevated ammonia	✓	✓	✓
Renal impairment	✓	✓	
Coagulopathy	✓	✓	✓
Micro vesicular steatosis on liver biopsy	✓		

Six or more of the following features in the absence of another explanation

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