

The Diagnostic Challenge of Acute Fatty Liver of Pregnancy

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1. INTRODUCTION

- Acute Fatty Liver of Pregnancy (AFLP) is a rare and potentially fatal complication of pregnancy.
- It typically affects women in the third trimester or in the postpartum period.
- We present a case of AFLP in the postpartum period.

2. PURPOSE

- To highlight the diagnostic challenge of Acute Fatty Liver of Pregnancy

3. CASE PRESENTATION

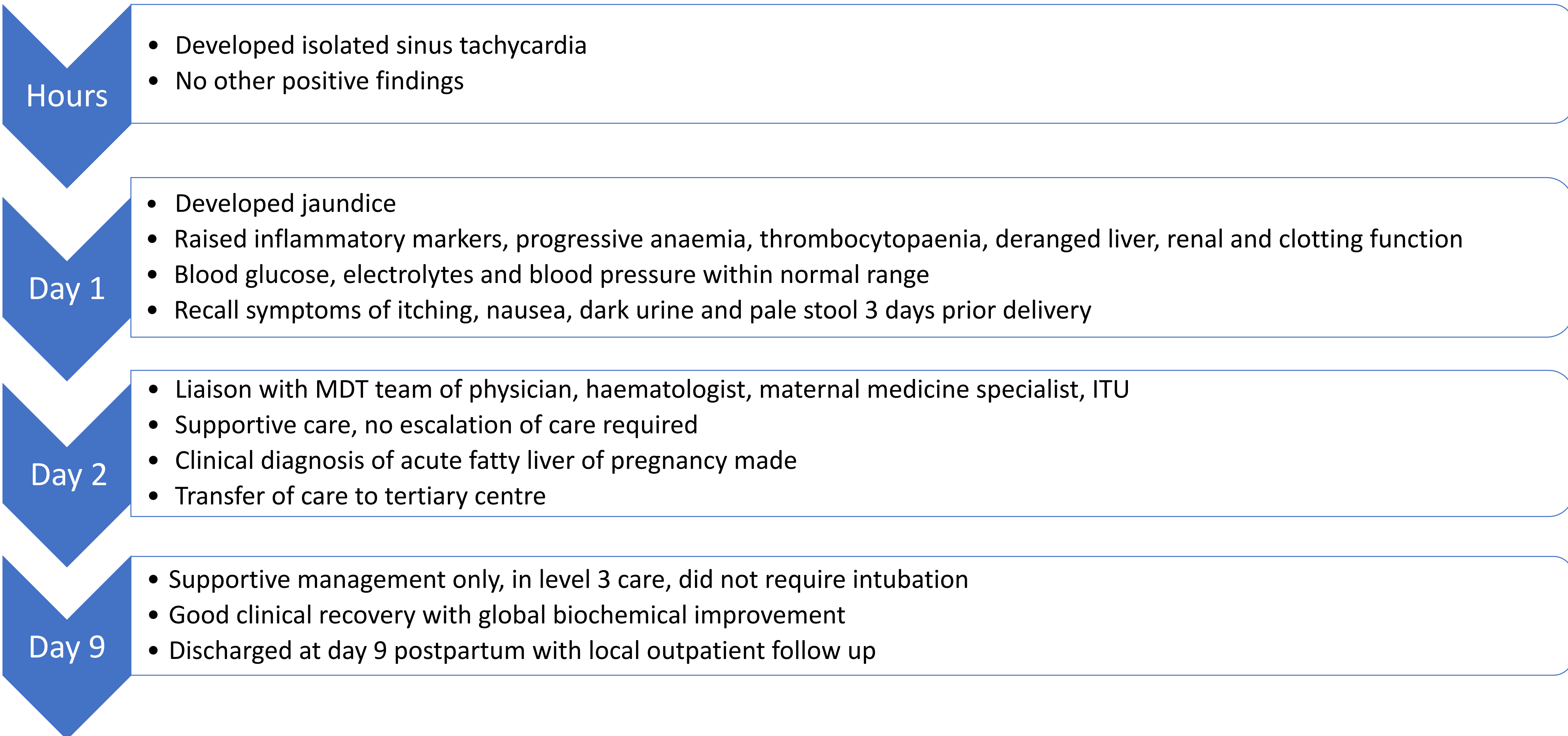
Background

- 31-year-old primigravida Caucasian female
- No significant past medical history
- Low-risk antenatally
- No history of hypertension or pre-eclampsia
- Biochemical markers were unremarkable at booking.

Labour

- Admitted to hospital with preterm premature rupture of membranes
- Progressed into preterm labour at 36/40, fully dilated within 3 hours of admission.
- Had an uncomplicated ventouse-assisted vaginal delivery.
- Delivered a single baby male in good condition.

4. PROGRESS



5. INVESTIGATIONS

US Abdomen	no abnormality, no post-hepatic obstruction
CT Abdomen Pelvis	normal liver and no structural causes for symptoms
Blood culture	Negative
Urine culture	Negative WCC 1144 RBC 22483, nil casts
Viral screen	Hepatitis A/B/C, CMV, EBV, adenovirus, HIV – all negative
Autoantibody screen	IgA, IgG, IgM all negative
DAT screen	Negative (25/9/21) Positive (28/9/21)
Alpha-1-antitrypsin	1.7 (1.1-2.1)
Beta-2 macroglobulin	3.35 (<2.16)
Complement C3 C4	C3 0.50 (0.90-1.80), C4 <0.10 (0.14-0.54)
VBG	Within normal range
Anticardiolipin	IgG within normal range IgM equivocal 14.0 (0.8-9.9)
ANA, ANCA, anti dsDNA, Anti GBM	All negative

Blood film	RBC: Crenated cells, WCC: occasional metamyelocytes, myelocytes present. Slight neutrophilia, neutrophils show toxic changes, smeared cells present Large platelet clumps
Ammonia	79
Ferritin	58 (15-300)
Transferrin	1.4 (2.0-3.6)
Troponin	54 (<14)
CK	166
Haptoglobin	<0.3 (0.3-2.0)
Reticulocyte count	83.2 (50-100)

	Booking	Day 0 AM	Day 0 PM	Day 1 AM	Day 1 PM	Tertiary discharge	2 weeks	3 weeks	5 weeks
Hb (115-165)	136	124	108	91	83	93	104	112	112
WCC (4.0-11.0)	10.2	25.1	23.5	21.5	20.0	11.8	10.9	5.9	5.7
Plt (150-400)	248	110	138	94	90	125	455	506	250
CRP (<5)		14	18	28	33			2	
ALT (<35)			378	266	213		98	64	38
AST (<35)				241	182	95			41
ALP (30-130)			317	281	267	386	383	205	147
Bilirubin (0-21)			118	113	107	64	53	33	18
Conjugated					69				6
Unconjugated					38				12
GGT (<38)				65	59	446			51
Albumin (35-50)			25	21	20	25	38	44	46
Creatinine (45-95)			187	195	189	181	83	76	79
Bile acid (0-14)				39.2					
Uric acid (140-360)				480	494				388
PT (10.3-13.3)				28.3	26.4	10.5	10.6	11.2	10.9
APTT (25.7-35.3)				80.0	74.9	39.9	36.5	35.7	33.1
INR (0.8-1.2)				2.2	2.1	1.0	0.9	0.9	0.9
Lactate (0.6-2.5)		1.76	3.2	1.58	1.14				
Glucose			5.1				5.0		
LDH (24—480)				751					335

6. DISCUSSION

- This case fulfilled the Swansea criteria and was diagnosed as AFLP. However, differentiating this against an atypical presentation of HELLP, sepsis and haemolytic uraemic syndrome remained challenging.

Swansea Criteria for AFLP (6 or +) ¹	
Vomiting	Abdominal Pain
Polydipsia	Encephalopathy
Elevated bilirubin	Hypoglycaemia
Elevated urate	Leucocytosis
Elevated transaminase	Ascites or bright liver on USS
Elevated ammonia	Renal impairment
Coagulopathy	Microvesicular steatosis on liver biopsy

HELLP syndrome ¹	
Complete	Partial
Elevated AST >70 iu/l	Elevated AST >40 iu/l
Low platelet <100 x10 ⁹ /l	Low platelet <150 x10 ⁹ /l
Haemolysis (LDH >600U/l)	Absence or presence of haemolysis

Pregnancy associated atypical HUS (3 or +) ²
Mechanical haemolytic anaemia (Hb <10g/dl, raised LDH, undetectable haptoglobin, presence of schistocytes)
Low platelet <150 x10 ⁹ /l
AKI (Creatinine >1.1mg/dl, >25% increase from baseline)
Typical features of thrombotic microangiopathy in kidney biopsy (fibrin/platelet thrombi, endothelial cell swelling, detachment from basement membrane, double contours)

- Our patient developed tachycardia only in the first instance and appeared clinically well. Detailed history and examination elicited signs and symptoms of hepatic dysfunction, together prompting further investigation.
- Careful history-taking is important in order to investigate for reasons for PPROM in a woman presenting in pre-term labour.

7. CONCLUSION

- Diagnosing AFLP remains challenging in the acute phase. Signs and symptoms of AFLP can be subtle and unsuspecting.
- A high index of suspicion is required for early recognition of deterioration and urgent involvement of the multidisciplinary team.
- Management of AFLP should be reinforced in the teaching curriculum in an Obstetrics Department.

1. Ch'ng C, Morgan M, Hainsworth I, Kingham J, 2002. Prospective study of liver dysfunction in pregnancy in Southwest Wales. *Gut*, 51(6), pp.876-880.
2. Bruel A, Kavanagh D, Noris M, Delmas Y, Wong E, Bresin E et al., 2017. Hemolytic Uremic Syndrome in Pregnancy and Postpartum. *Clinical Journal of the American Society of Nephrology*, 12(8), pp.1237-1247.