

OBJECTIVE

To discuss a rare case occurring postnatally with an initial differential diagnosis of HELLP (Haemolysis, Elevated Liver enzymes and Low Platelets) syndrome or Acute Fatty Liver in Pregnancy (AFLP).

BACKGROUND

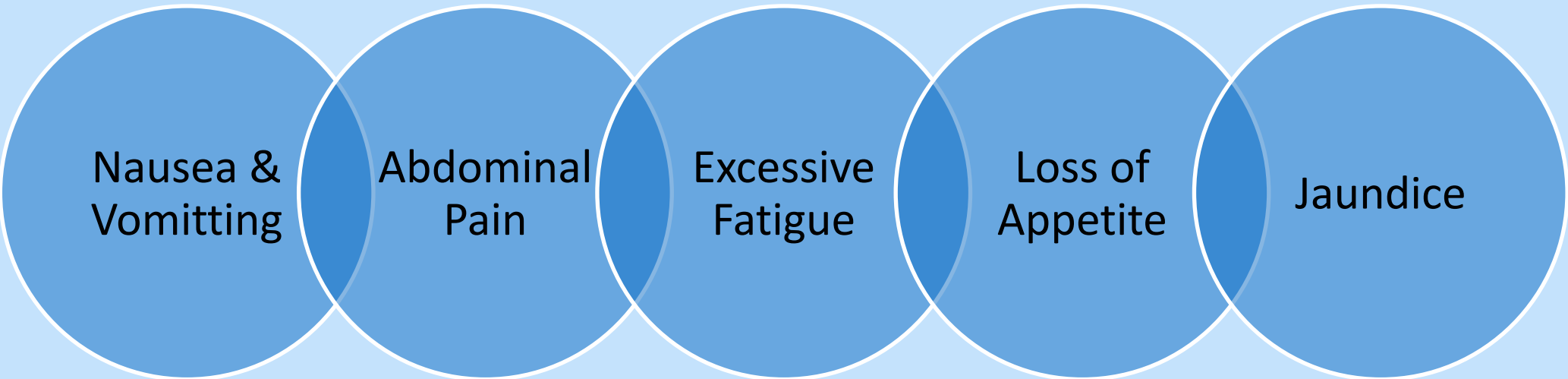
AFLP is a rare but potentially fatal disorder of late pregnancy which develops in patients who have a genetic disorder (ie. G1528C genetic mutation) that leads to mitochondrial fatty acids which are defective. This then causes slow & subtle fat accumulation in hepatocytes which can disrupt normal liver function such as detoxifying chemicals and metabolising drugs.

The normal fat content of the liver is approximately 5%. In women with AFLP, this percentage can range from 13% to 19%.

It occurs in about 1 in 20,000 pregnancies and is more common in first pregnancies, male babies and twins.

AFLP shares clinical and biochemical features with other disorders in pregnancy such as HELLP, pre-eclampsia, obstetric cholestasis or viral hepatitis.

Signs & Symptoms Associated with AFLP:

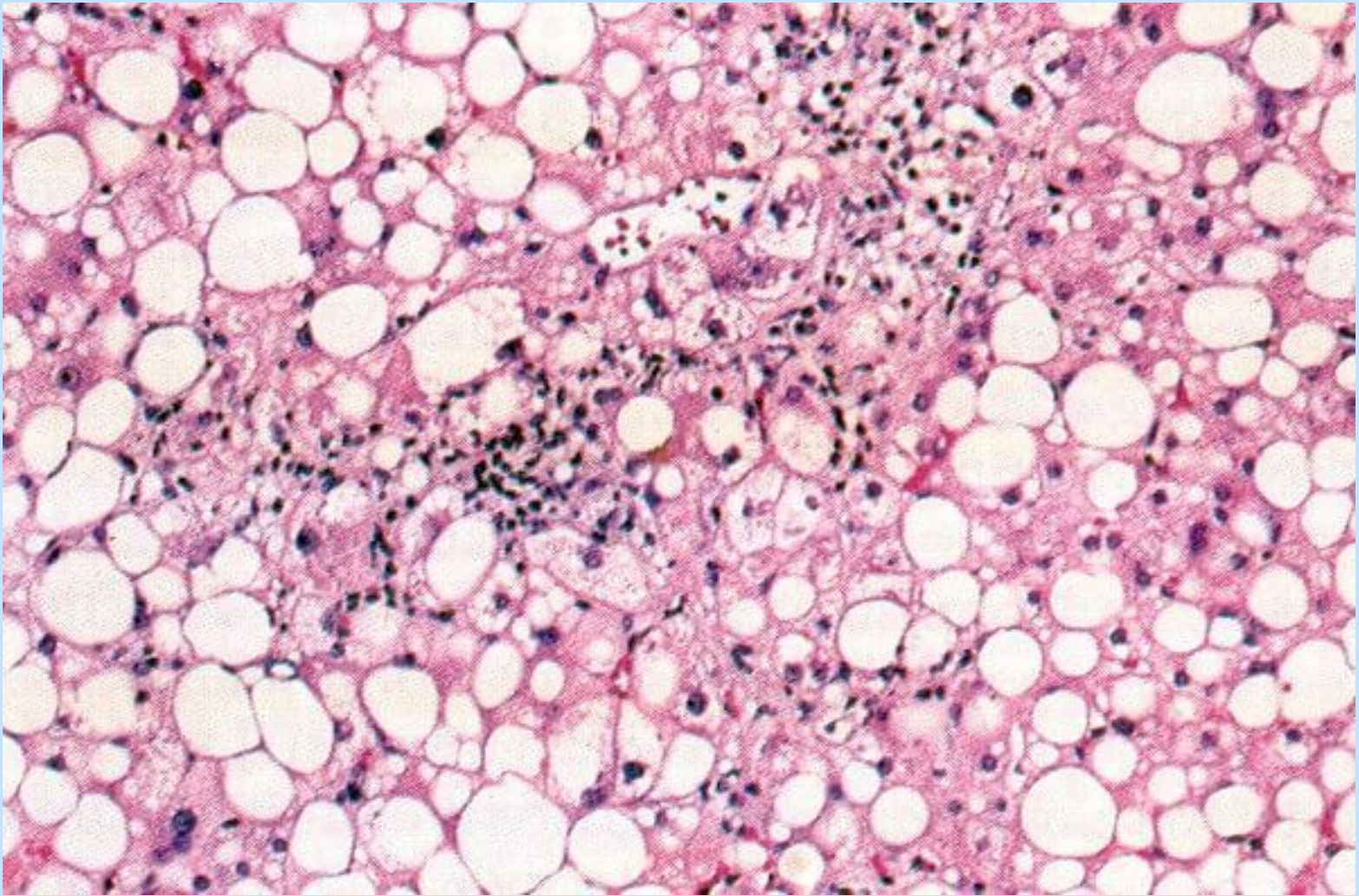


One of the complications of AFLP is maternal metabolic acidosis secondary to impaired clearance of serum lactate by damaged hepatocytes which can that affect fetal acid-base status. Therefore, immediate correction of maternal metabolic acidosis is essential to ensure fetal well-being.

CASE REPORT

36 year old female with a BMI 31 had an elective lower segment caesarean section for dichorionic diamniotic (DCDA) twins at 37/40. Patient felt unwell 24 hours prior to delivery. Patient had no history of hypertension or Diabetes. Following the Caesarean Section, patient sustained a postpartum haemorrhage of 1.2 litres due to large placental bed sinus and an atonic uterus. Patient had a vacant episode during delivery of the second twin and recovered quickly. Patient scored a MEWS of 4 for a BP of 159/99, HR 62, RR 19 in the PN ward. Capillary blood glucose (CBG) at the time was 3.3. Patient was given glucotabs, biscuits and orange juice. A repeat CBG was 3.6 an hour later and treated with glucogel. She had 2+ proteinuria, with moderate hypertension, hence treated as pre-eclampsia . Urine PCR & PET bloods were requested. Her CBG was then 2.7mmols/L, treated with IV glucose 20% and the CBG normalized.She developed chest tightness with a normal ECG. AP CXR showed slight enlargement of the heart. She was treated with a stat dose of Labetolol 200 mg and commenced on Magnesium sulfate infusion after transfer to HDU along with thromboprophylaxis and supportive measures.

Initial Blood Test Results		
ALT	992	Chest X-Ray (AP) •Enlarged Heart
Bilirubin	45	Bedside ECHO •Normal
ALP	145	MRI Stress Perfusion •Normal
GGT	116	Liver Ultrasound •Normal
Fibrinogen	2.08 - > went down to 1.99	
Clotting	Normal	
Calcium	Normal	
Urate	453	
LDH	596	
Haptoglobin	0.56	
Ammonia	47	
Hb	145	
Platelets	193	
WCC	13.6 -> went up to 25.11	
Troponin	762 -> went down to 213	
Pro-BNP	497 -> went down to 68	
Urine PCR	729	
Na+	133	
K+	3.9	
Creatinine	79	
Estimate GFR	83 (AKI Stage 1)	



Histology of AFLP. Source: <https://www.preeclampsia.org/public/frontend/assets/img/gallery/ourstories/AFLP.jpg>

DISCUSSION

Acute Fatty Liver of Pregnancy can be confused with pre-eclampsia & HELLP Syndrome. It can be distinguished from HELLP syndrome by differences in how the two diseases develop.

AFLP	VS	HELLP/Pre-Eclampsia
Less likely	Hypertension	Common
Present	Abnormal Glucose	Absent
More Severe	Abnormal LFT	Less Severe
Common	Abnormal Renal Function	Less Common
Increased	Bilirubin	Normal
Present	Proteinuria	Present
High	Ammonia	Normal
Prolonged	APTT/PT	Normal
+	Encephalopathy	-

Swansea criteria is used to assess the likelihood of AFLP while avoiding liver biopsy in patients who are frequently coagulopathic and thrombocytopenic.

6 or more of the following are required in the absence of another cause to diagnosis AFLP:

Vomitting ✓	Abdominal Pain ✓	Polydypsia/Polyuria	Encephalopathy	Bilirubin>14 µmol/L ✓	Hypoglycemia <4mmol/L ✓	Urea >340 µmol/L
Leucocytosis >11x10 ⁹ /L ✓	Ascites/Bright liver on USS	ALT>42 IU/L ✓	Ammonia>47µmol/L ✓	Creatinine >150µmol/L	PT>14 secs or APTT >34 secs	Microvascular steatosis on liver biopsy

In this case, It was difficult to differentiate between AFLP and HELLP syndrome. Hypertension, elevated PCR, (raised/normal?) LDH, Urates and normal clotting was favouring HELLP syndrome. However, the final diagnosis of AFLP was made on day 2. Increasing WCC and low fibrinogen levels was also diagnostic of AFLP. As the patient had normal Clotting , this was also an important feature to differentiate AFLP from HELLP

Conclusion

In this case, the patient met the 7 Swansea criteria out of 14 for the diagnosis of AFLP. The final diagnosis of AFLP was made on day 2 when the patient developed