RECURRENT ACUTE FATTY LIVER OF PREGNANCY IN A PATIENT WITH HEPATIC HAEMANGIOMA

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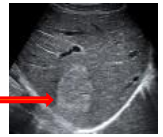
AIM

Acute fatty liver of Pregnancy (AFLP) is rare and serious maternal disorder, usually seen in third trimester of pregnancy or early postpartum with significant maternal and perinatal mortality. We present a rare case of recurrent AFLP with hepatic haemangioma.

CASE

31 years, G2P1, previous caesarean delivery of baby boy at 36 weeks for AFLP.

At Booking: Non-smoker, normal blood pressure, no proteinuria, BMI 22.3. Patient requested elective LSCS which was planned at 39 week.
Monthly full blood counts and liver function tests remained normal until 34 weeks. Serial growth scans remained >10th centile.
Abdominal ultrasound at 29^{+5}/40 showed a 7mm haemangioma in the right lobe of liver.



She had 2 episodes of reduced foetal movements during this pregnancy (27 and 32 week).

At 35^{+1}/40: Patient was dmitted to antenatal ward with raised biochemical markers (ALT 402 U/L, bile acids 67umol/L) and WBC (21.3x10^{9}/L); Hb dropped to 95 g/dl (from 113 g/dl at the start of pregnancy). Patient was asymptomatic at admission.
A multidisciplinary discussion with medical, haematology, neonatal and anaesthetic teams → delivery by LSCS was planned at 35^{+6}/40. An abdominal ultrasound was planned which could not happen as she deteriorated and required delivery before the scan.

At 35^{+4}/40: There was no improvement in her full blood counts and LFTs. She developed epigastric pain and polydipsia, and low fibrinogen levels of 1.6 g/l, prolonged PT (15.1 seconds) and aPTT (35.3 seconds), hence 2 grams of fibrinogen was administered. Her blood pressure, platelets, blood glucose, LDH and urine PCR were normal. She was now meeting 6 features of Swansea criteria and we diagnosed AFLP.

Delivery: A category 2 LSCS was performed due to clinical deterioration. A male baby delivered in good condition with birth weight of 2480 grams (12th centile). She developed atonic postpartum haemorrhage of 1011ml which settled with medical management. The ROTEM analysis remained normal.

Postnatal: Post-operative care in ITU as patient developed acute kidney injury (AKI), ALT continued to rise to 855U/L, platelets dropped to 100 x 10^{9}/L (from 202 pre operatively) fibrinogen remained low (1.4g/dl), PT and aPTT were prolonged. She improved gradually by day 5 postoperatively with normal platelets and coagulation profile. LFT were normalised after 2 weeks.

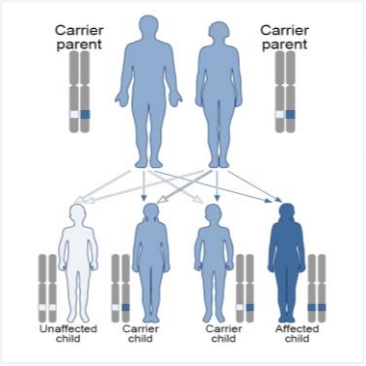
At Discharge: She was advised to avoid hormonal contraception, and a referral to clinical geneticist was recommended at postnatal follow up. She was reviewed and discharged by gastroenterologist post natal. Baby was followed up by neonatal team.

DIFFERENTIAL DIAGNOSIS

Severe pre-eclampsia, HELLP syndrome and Kasabach-Merritt syndrome (a rare possibility of rapidly growing hepatic haemangioma, thrombocytopenia and hypofibrinogenemia)

DISCUSSION

Recurrence of AFLP is very low. It can be linked to defects in maternal mitochondrial β oxidation of fatty acids, reported in mothers who are heterozygous carriers of gene mutation (G1528C) on chromosome 2 (2p23), resulting in deficiency of long-chain3-hydroxyacyl-CoA dehydrogenase (LCHAD) enzyme. Our patient is currently awaiting genetic counselling.



LCHAD Autosomal recessive - Chromosome 2

CONCLUSION

Genetic counselling, pre-implantation genetic diagnosis and neonatal follow up for assessment of LCHAD/other fatty acid oxidation deficiencies can be useful.

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