HELLP Syndrome

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Introduction

- The acronym HELLP was coined by Weinstein in 1982 to describe a syndrome consisting of
- Hemolysis,
- Elevated Liver enzymes and
- Low Platelet count.
- It is a variant of severe pre-eclampsia or a complication of it.

Definition

- It is a syndrome that is characterised by
- preeclampsia,
- hepatic endothelial disruption,
- platelet activation, aggregation and consumption,
- resulting in microangiopathic hemolysis, ischemia and hepatocyte death.

Incidence

- 0.5 to 0.9% of all pregnancies
- 10 to 20% of cases with severe preeclampsia.

Pathogenesis

- Pathogenesis of preeclampsia-
  - Endothelial dysruption
  - Abnormal vascular tone
  - Vasospasm
  - Coagulation defects
- Involves smaller terminal arterioles
- This vasculopathy if involves single segment or entire liver leads to HELLP syndrome

Classical histolgical lesion in Liver

- Periportal or focal parenchymal necrosis with deposits of hyalin like material

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Intra hepatic haemorrhage

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Subcapsular haematoma

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Eventual rupture of Glisson's capsule

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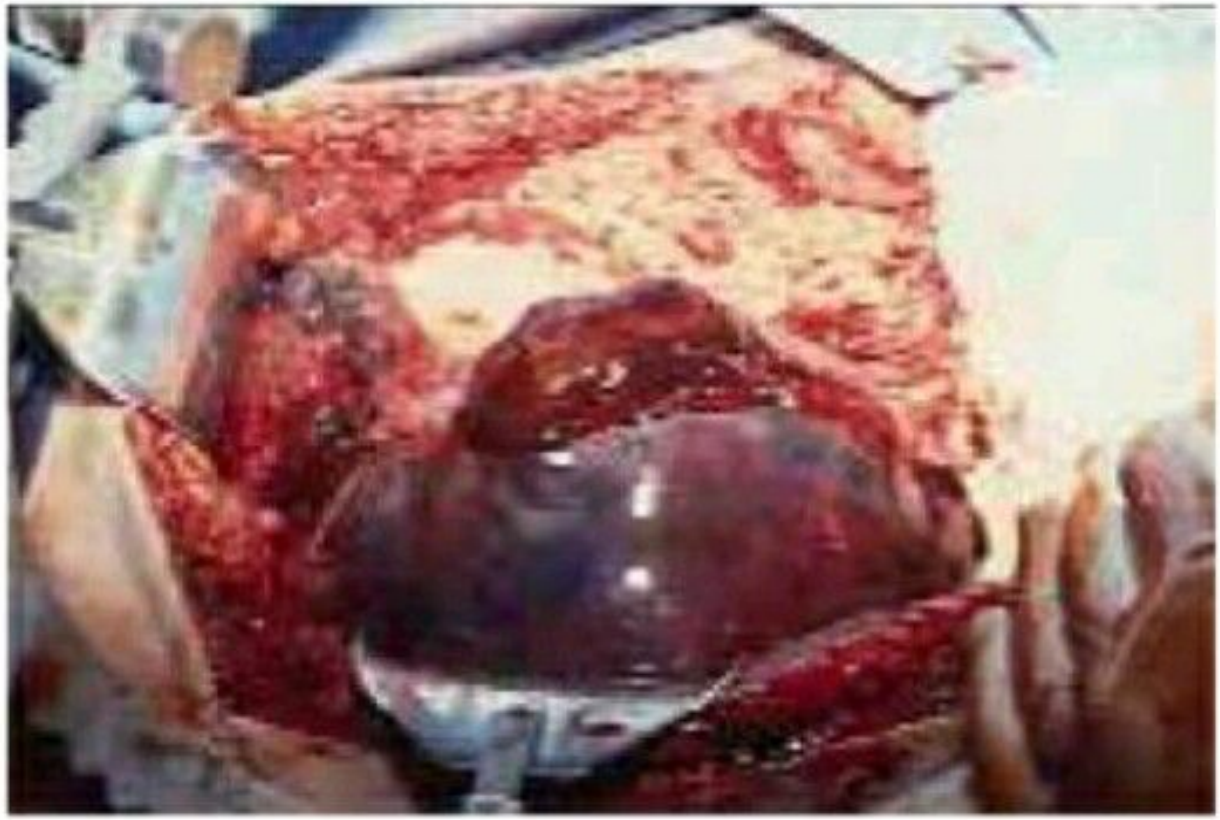

Hemolysis

- It is due to thrombotic microangiopathy

Endothelial dysfunction

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Intimal damage, foam cell, hyaline & fibrin deposition

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Vessel wall narrowing

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Fragmentation of red cells

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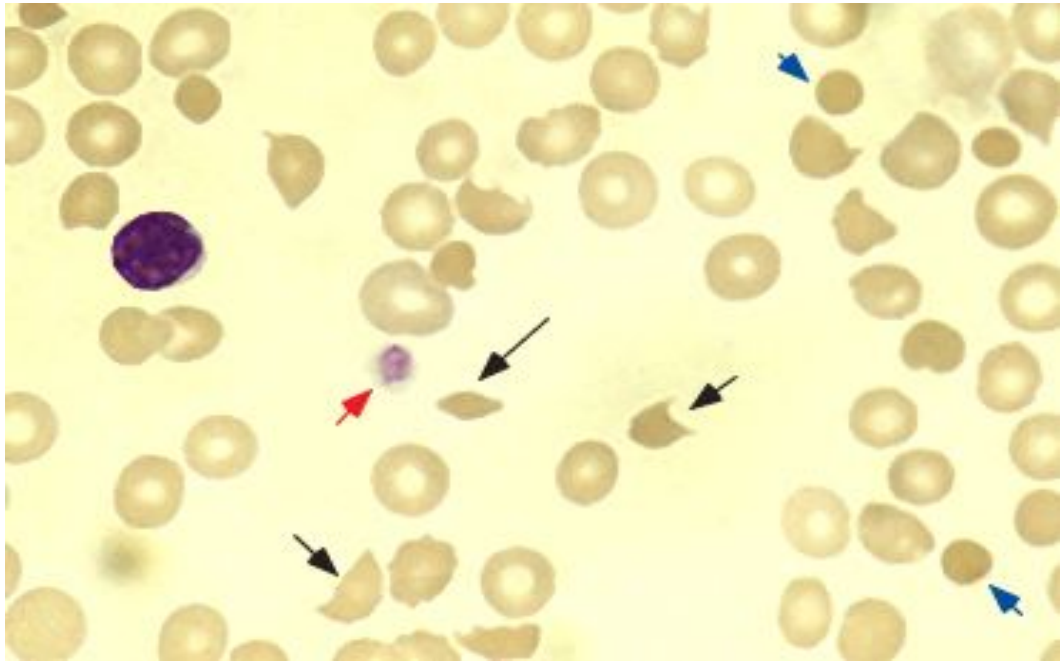
Peripheral smear shows

- Schizocytes, burr cells, hemet cells, etc



Haemolysis cont…

- Increase in serum LDH & decrease in Hb concentrations
- Haemoglobinemia & haemoglobinuria
- Unconjugated bilirubinaemia
- Haptoglobin levels – low or undetectable(more specific indicator)

Thrombocytopenia

- Platelet count < 150,000/cmm
- Due to increased consumption
- DIC is the primary process in HELLP syndrome

Immune system disorder theory

- Abnormal humoral as well as cell mediated immune dysfunction is also observed in patients with HELLP syndrome

Risk factors

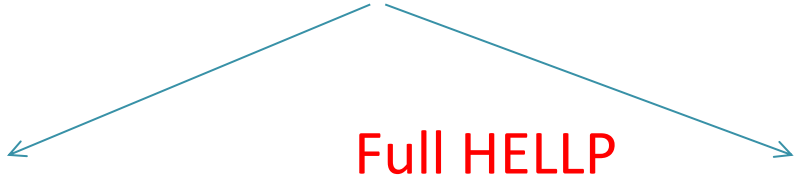
- Multiparity
- Age >25 yrs
- White race

CLASSIFICATION

- Tennessee Classification System

Based on laboratory criteria

1. Platelet count < 100,000/cmm
2. AST >70 IU/L & LDH > 600 IU/L
3. Hemolysis on peripheral smear



Partial HELLP

Any 2 of 3 criteria

Full HELLP

All of 3 criteria

Mississippi classification

	Class1	Class 2	Class3
Platelet count(cmm)	<50,000	50,000 - 100,000	>100,000
AST	> 70 IU/L	> 70 IU/L	>40 IU/L
LDH	>600 IU/L	>600 IU/L	>600 IU/L
Hemolysis on smear	present	present	present

Diagnosis

- Clinical features
  - 70% of the cases develop between the 27th and 37th gestational weeks
  - 20% beyond the 37th gestational week
  - 10% occur before the 27th week
  - With postpartum presentation, onset is typically within first 48 hrs of delivery

Symptoms

- Right sided upper abdominal or epigastric pain (86-90%)
- Nausea (45-85%)
- Headache (50%)
- Malaise (80-90%)

Signs

- Protenuria (85-90%)
- Right upper quadrant tenderness (86%)
- Increased blood pressure (67%)
- Edema (55-65%)

Laboratory findings

- Low platelets <100,000/cmm
- Elevated liver enzymes – AST >70 IU/L
- Hemolysis – abnormal peripheral smear
- Total bilirubin >1.2 mg%
- PT, aPTT, S. Fibrinogen - if abnormal, DIC is suspected
- S. uric acid is raised

Differential diagnosis

- Diseases related to pregnancy
  - Benign thrombocytopenia of pregnancy
  - Acute fatty liver of pregnancy
- Infectious and inflammatory diseases, not specifically related to pregnancy
  - Viral hepatitis
  - Cholangitis
  - Cholecystitis
  - Gastritis,gastric ulcer
  - Acute pancreatitis

Complications

- Maternal
  - Subcapsular liver hematoma & liver rupture
    - DIC
    - Acute renal failure
    - Cerebral edema
    - Pulmonary edema
    - Wound hematoma/infections
    - Retinal detachment
    - Cerebral infarction & haemorrhage
    - Maternal death

Fetal/neonatal complications

- Perinatal death
- IUGR
- Preterm delivery
- Neonatal thrombcytopenia
- RDS

Management

- Admission to hospital
- Stabilization
- Evaluation
- Secure IV line
- Transfusion of Blood and blood products
- Catheterization
- Respiratory assessment
- Fetal assessment(NST, BPP, colour doppler)

- Immediate delivery:
  - > 34 weeks' gestation or later
  - Nonreassuring fetal status
  - Presence of severe maternal disease: multiorgan dysfunction, DIC, liver infarction or hemorrhage, renal failure.

- 27 to 34 weeks of gestation
  - Deliver within 48 hrs after stabilization and evaluation
  - Steroid treatment for fetal lung maturity

- Before 27 weeks
  - Termination of pregnancy should be strongly considered.

Summary

- HELLP syndrome is unique to pregnancy
- 0.5 to 0.9% of all pregnancies
- Delivery and supportive management is cure
- Multidisciplinary approach
- Tertiary care
- Outcome is generally good if intervened early

Thank you