**Objectives**

- Differentiate HELLP Syndrome from Preeclampsia and Eclampsia
- Discuss pathophysiology and epidemiology of HELLP
- Differentiate HELLP from other diseases
- Discuss laboratory and diagnostic testing to diagnose HELLP
- Discuss management of women with HELLP

**Preeclampsia**

“Preeclampsia is a pregnancy-specific hypertensive disease with multisystem involvement. It usually occurs after 20 weeks of gestation, most often near term, and can be superimposed on another hypertensive disorder.”

**Preeclampsia Redefined**

**Without severe features**
- Hypertension after 20 weeks
- B/P: $\geq 140$ mm Hg systolic OR $\geq 90$ mm Hg diastolic on two occasions 4 hours apart
- Urine protein/creatinine ratio $>0.3$ or 300mg in 24-hour urine collection
- With or without edema
- No severe features

**With severe features**
- Hypertension after 20 weeks
- B/P: $\geq 160$ mm Hg systolic OR $\geq 110$ mm Hg diastolic on two occasions taken 4 hours apart while on bed rest
- Headache
- Visual/auditory disturbances
- Hyperreflexia
- Significant edema/periorbital/wt gain
- Pulmonary edema
- Oliguria
- Epigastric or right upper quadrant pain
- Labs
  - Urine protein/creatinine ratio $>0.3$
  - Serum creatinine $>1.1$ mg/dl
  - Thrombocytopenia
  - $<100,000$/microliter
  - Elevated liver enzymes

**Treatment of Preeclampsia**

**Without severe features**
- Hypertension management
- Labetalol
- Hydrozidine
- Surveillance

**With severe features**
- Ensure IV access (large bore needle)
- Total IV fluids $\sim 125$ ml/hr
- Monitor blood pressure
- Oxygenation
- NPO
- Strict I&O
- Hypertension management
- Labetalol
- Hydrozidine
- Nifedipine
- Seizure prophylaxis
- Magnesium Sulfate
- Deliver of baby curative

**Eclampsia**

“The presence of new-onset grand mal seizures in a woman with preeclampsia. Eclampsia can occur before, during, or after labor.”

(ACOG, p. 17, 2013)
Treatment of Eclampsia

- Ensure IV access (large bore needle)
- Strict I&O
- Total IVF = 125 ml/hr
- NPO
- Monitor blood pressure
- Airway support
- Oxygenation
- Seizure precaution

- Hypertension management
- Magnesium sulfate
- Initiate additional bolus
- Increase
- Diazepam
- Lorazepam
- Midazolam

*Delivery of baby may be necessary if pregnant

HELLP Syndrome

Hemolysis on microangiopathic blood smear
Elevated Liver enzymes
Low Platelets

Is HELLP a variant of Preeclampsia or a separate disease?

**YES ☐**

**NO ☐**

Pathophysiology

- Pathogenesis is unknown
- Preeclampsia
- Fetal long chain 3-hydroxyacyl CoA dehydrogenase deficiency (LCHAT)
- Acute maternal immune rejection of maternal cells coming in contact with foreign fetal cells
- Placental instigation of inflammatory disease targeting liver

Pathophysiology cont.

- Hemolytic anemia
  - Formation of fibrin in small vessels = RBC destruction
- Elevated Liver enzymes
  - Activation of the coagulation cascade leading to consumption of platelets = Hepatic inflammation and multiorgan microvascular injury
- Multisystem involvement
  - Kidney failure
  - Hepatic necrosis or rupture
  - Neurologic problems
- Delivery is curative
- Improvement seen 48 hours after delivery

Epidemiology

- Cause is unknown
- No preventative therapies
- 0.1-0.8% of all pregnancies
- 3% develop between 17 and 20 wks
- 80% develop between 28-36 wks
- Possible to develop in 3rd trimester
- 30-48 hours to 7 days postpartum
- 70% develop before delivery
Epidemiology cont.

- 10-20% diagnosed with preeclampsia/eclampsia
- 15-20% do not present with preeclamptic/eclamptic symptoms
- Maternal age > 25 years
- Caucasian/European
- History of preeclampsia or HELLP
- Family history of preeclampsia or HELLP
- > 50% are multiparas

Presentation

- Common
  - Right upper quadrant/epigastric/abdominal pain
  - Nausea/vomiting
  - Malaise/fatigue/not feeling well
  - Proteinuria
  - Hypertension
- Less common
  - Headache
  - Visual disturbances
  - Fluid retention and weight gain
  - Nose bleeds or other bleeding that will not stop
  - Jaundice
  - Seizures (rare)

Often Misdiagnosed

- Flu/viral illness
- Appendicitis
- Gallbladder
- Hepatitis
- Fatty liver of pregnancy
- Hemolytic-uremic syndrome (HUS)
- Idiopathic Thrombocytopenic Purpura (ITP)
- Thrombocytopenic Purpura (TTP)
- Lupus

Complications of HELLP

- Disseminated intravascular coagulation (DIC) – 21%
- Pulmonary edema – 6%
- Acute renal failure – 8%
- Liver failure/subcapsular hepatic hematoma – 1%
- Retinal detachment – 1%
- Placental abruption (if still pregnant) – 16%
- Maternal death
- Neonatal morbidity and mortality

*If still pregnant, delivery of the baby is the road to recovery.

Management

Identification

- Comprehensive assessment
- Health/Ob history
- Labs/Imaging

Surveillance

Emergent Stabilization

Admit to monitored unit
Comprehensive Assessment

- Accurate health/obstetrical history
- Comprehensive physical assessment
- Assess for abdominal tenderness/epigastric pain 65% - 90%
- New or increased bleeding
- Vital signs
  - Hypertension
  - Tachycardia
  - Tachypnea
- Jaundice 5%
- Edema 50% - 67%
  - Periorbital
  - Nondependant

Labs and Imaging

- Type and Screen
- CBC
  - Hemoglobin/Hematocrit ↓
  - Platelets ↓
- CMP
  - LDH ↑
  - AST/ALT ↑
  - Total Bilirubin ↑
  - BUN↑
  - PT normal
  - PTT ↑
  - D-dimer ↑
  - Fibrinogen ↓
  - Haptoglobin ↓
  - CT of liver

Multidisciplinary Care

- Nurses
- Obstetrician (if still pregnant)
- Intensivist/PCP
- Surgeon
- Hematologist
- Renal Specialist
- Lab/Blood bank
- Radiology

Emergent Stabilization

- Deliver baby (if still pregnant)
- Bed rest/seizure precautions
- Manage hypertension
  (Target: 160/105 or 110)
- IVF management
- Monitor bleeding
- Blood products
- Laboratory surveillance
- Imaging (CT, U/S)
- Antihypertensives
- Magnesium Sulfate
- Corticosteroid therapy
  (controversial)
- Ongoing Assessment
  - Repeat labs every 4-6 hours
  - Consults
    - Surgery
    - Hematology
    - Renal

Hepatic Complications

- No signs of rupture/contained
  - Manage conservatively
    - Serial CT/MRI of liver or careful ultrasound
    - Avoid increased intra-abdominal pressure
    - May need transfusion and volume replacement

- Rupture is most commonly anterior superior right upper lobe
  - Shoulder pain, N/V
  - Abdominal swelling
  - Marked increase in AST
  - Shock
  - Transfuse
    - RBC
    - FFP
    - Platelets
    - Surgery
    - Transplant

*50% morbidity for mother and child

Prognosis

- Generally good outcomes
- Maternal mortality 1% - 3%
- Perinatal mortality 35% in Grade I or complete HELLP
- Most stabilize within 24-48 hours after delivery
- Recurrent rate 2% - 27%
  - HELLP 7%
  - Preeclampsia 18%
  - Gestational hypertension 18%
- No therapy to prevent recurrence
Special Consideration

- Separation from newborn
- Breastfeeding barriers
- Emotional Support
- Postpartum depression
- PTSD
- Financial impact

Patient Education

- Signs and symptoms
- When to seek help
- Speak slowly
- Use plain nonmedical language
- Teach back
- Reinforcement material
- Include support person

Conclusion

Positive clinical outcomes

Awareness
Identification
Emergent stabilization
Collaborative care
Admit to specialized unit

Q & A

References


References


