# Title of Case

HELPP Syndrome: An Atypical Presentation

# Authors of Case

Please indicate corresponding author by *

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# Summary

Up to 150 words summarising the case presentation and outcome

Patient is a G4P3, 30 year old, Caucasian, female at 37 weeks gestation with twins. She initially presented to the office at 36 weeks and 6 days gestation with chief complaint of itching on the abdomen, hands, and feet. Patient's blood pressure was 122/76, and she had no other complaints at that time.

Patient was sent to Labor and Delivery for a CBC and CMS, which showed elevated AST and ALT, marginally low platelets, and elevated uric acid. A fetal non-stress test was reactive; indicating that neither twin was in distress. Patient was admitted to L&D for possible severe pre-eclampsia and early HELLP syndrome.

CBC and CMS were repeated in the morning showing increased AST and ALT, lowered platelet count, and increased uric acid from the previous day. Patient also started complaining of RUQ pain and had a BP of 132/90. The patient was scheduled for a c-section later that day. Healthy twin boys were delivered successfully with no complications. Repeat CBC and CMS on POD #1, showed slowly decreasing AST/ALT, increasing platelet count, and decreasing uric acid. The patient was discharged on POD#3 with no further complications.

# Background

Why you think this case is important – why you decided to write it up

HELLP syndrome and pre-eclampsia can be very serious complications of pregnancy. Most of the time early diagnosis of pre-eclampsia can be made based on high blood pressure, proteinuria, and edema. However, atypical presentations do exist and noticing the signs and symptoms are critical. In this case, the patient did not show any symptoms other than pruritus; which in this case was a sign of impending liver problems. Had the complaint of pruritus not been investigated further, the health of the mother and the twins she was carrying could have been put in jeopardy. I feel this case is a good example of the need to fully listen to the patient and not to discount any sign or symptom.

# Case Presentation

Presenting features, medical/social/family history

HPI:

- L.H. is a 30 y/o, G4P3, female, presenting at 36 and 6/7 weeks gestation with twins following a failed bilateral tubal ligation. She has a chief complaint of pruritus of the abdomen, hands, and feet.
- Began receiving prenatal care at 8 weeks gestation. Gestation has been uneventful. Blood pressure has remained stable in the 120's/80's.
- Fetal ultrasounds have shown normal growth of Baby A and Baby B. Last ultrasound at 36 weeks showed Baby A lying transverse, and Baby B lying breech. Patient was informed of the possible need for Cesarian section due to fetal lie.
- Weekly fetal non-stress tests beginning at 34 weeks have all been reactive.

PMH/PSH:

- Genital HSV
- Laparoscopic bilateral tubal ligation 2006
Meds: Prenatal vitamin, Acyclovir 400mg twice a day since 34 weeks gestation
Allergies: NKDA
Family Hx: non contributory
Social Hx:
- Denies ETOH, tobacco, or elicit drug use
- Lives with boyfriend, his four children from a previous relationship, and her three children from a previous relationship
- No history of domestic violence

ROS:
- Skin: Pruritus of the abdomen, hands, and feet. No rash. Stretch marks over abdomen.
- Back: Increasing back pain over course of gestation. No history of injury.
- Genitourinary: Genital HSV with no active lesions. Increased frequency of urination over course of gestation.
- All other systems unremarkable.

PE:
- Vitals in office: Ht. 5’ 10”, Wt. 230#, T 97.2, P 119, R 22, BP 122/76
- Gen: AA0x3, NAD
- Skin: Erythematous stretch marks over entire abdomen. Excoriations on abdomen. Patient noted to be scratching vigorously at abdomen, hands, and feet. No evidence of rash.
- HEENT: Neck supple, trachea midline, no scleral icterus
- Heart: RRR, no MRG
- Lungs: CTA B/L
- Abd: Gravid uterus consistent with 36 weeks gestation of twins, soft, non-tender
- Genital: Cervix dilated to fingertip, thick, and high. No presenting parts of either fetus in pelvis. No external genital lesions.
- Back: No CVA tenderness
- Extremities: 1+ edema LLE/RLE. No clubbing or cyanosis.

INVESTIGATIONS

Antenatal Screening Tests:
- Rubella: Immune
- Blood Type/Rh: O+
- RPR: -
- GBS 36 weeks: -
- Genital HSV: +
- GC/Chlamydia: -
- HBSag: non-reactive
- OGT 28 weeks: WNL

Laboratory Studies 3/6/09:
- Sodium: 135
- Potassium: 3.6
- Chloride: 104
- HCO3: 22.3 (L)
- BUN: 8
- Creatinine: 0.6
- Glucose: 103
- WBC: 6.3
- H/H: 13.3/38.4
- Platelets: 175
- AP: 222 (H)
- AST: 42 (H)
- ALT: 63 (H)
- Uric Acid: 6.6 (H)
- Acute Hepatitis Profile: -
- UA: 1+ leukocytes
Laboratory Studies 3/7/09 (pre-op):
- AST: 54 (↑)
- ALT: 73 (↑)
- Platelets: 147 (↓)
- Uric Acid: 7.2 (↑)
- BP: 132/90

Ultrasound: Baby A Transverse, Baby B Breech

**DIFFERENTIAL DIAGNOSIS** *If relevant*
- Acute Fatty Liver of Pregnancy
- Thrombotic Thrombocytopenic Purpura
- Hemolytic Uremic Syndrome
- Cholecystitis
- Esophagitis
- Gastritis
- Hepatitis

**TREATMENT** *If relevant*
- Cefazolin 1 gram pre-op
- Immediate operative delivery
- Primary LTCS (low transverse Cesarian section) with bilateral tubal ligation

**OUTCOME AND FOLLOW-UP**
- LTCS was performed without complication with delivery of healthy twin boys.
- Laboratory Studies performed POD #1:
  - WBC: 8.5
  - H/H: 11.4/32.8
  - Platelets: 160 (↑)
  - AST: 31 (↓)
  - ALT: 41 (↓)
  - Uric Acid: 6.2 (↓)
  - BP: 110/74
- Patient was discharged on POD #3 without complication. CBC and CMS were monitored while in-patient and returned to normal levels upon discharge. Two week post-op visit was scheduled.

**DISCUSSION** *including very brief review of similar published cases (how many similar cases have been published?)*

1.) What is the failure rate of tubal sterilization?
   Failure rate of tubal ligation sterilization is dependent upon many different variables including patient characteristics, operator experience, sterilization technique, and method of sterilization chosen. The US CREST (Collaborative Review of Sterilization) study included 10,685 women undergoing sterilization from 1978 and 1987 and followed the cohort for 14 years. The overall 10-year probability of pregnancy was 18.5 per 1000 procedures or 1.85%. A Canadian retrospective study of 311,960 female sterilizations from 1980 to 1999 determined a 10-year probability of pregnancy of 8.4 per 1000 procedures or 0.84%.
   Current probabilities of pregnancy following tubal sterilization are more difficult to determine as newer methods of tubal sterilization have developed in the past decade. Procedures such as laparoscopic tubal ligation and Essure tubal ligation have come into popularity over the past three to five years and are just recently becoming integrated into the common practices of OB/GYN’s across the United States. Further studies with longterm follow-up will need to be performed to determine failure rates of newer methods of female sterilization. *(RCOG 2004)*

2.) What are the most common presenting symptoms of HELLP syndrome? Is pruritus a symptom commonly associated with HELLP syndrome?
   The symptoms of HELLP syndrome can be vague and variable; which can lead to delayed diagnosis or misdiagnosis with another disorder. The most common symptoms of HELLP syndrome are general malaise (90%), epigastric pain (65%), headache (31%), and nausea/vomiting (30%). Physical exam is often normal in patients with HELLP syndrome. Right upper quadrant tenderness is the most common physical exam finding, occurring in up
to 90% of women with HELLP syndrome. Extremity edema is generally considered not to be useful in diagnosis of HELLP syndrome, as it is present in up to 30% of normal pregnancies. Hypertension and proteinuria, which are diagnostic for preeclampsia, may be absent or mild in HELLP syndrome. (Haram 2009)

After a thorough literature review, pruritus was not found to be a common symptom of HELLP syndrome. Pruritus during pregnancy is typically associated with intrahepatic cholestasis of pregnancy. However, diffuse pruritus occurring during pregnancy is a symptom which warrants further laboratory investigation. (Roy 2008)

Through the literature review, I concluded that pruritus is not a symptom that should prompt the diagnosis of HELLP syndrome, but that it can be a symptom of other complications of pregnancy and should be investigated. Also, HELLP syndrome can be difficult to diagnosis, so any symptom out of the norm for pregnancy should be followed with adequate work-up. I would consider the patient in the above case to have an atypical presentation of HELLP syndrome. If further investigations had not been performed, HELLP syndrome may have not been diagnosed until it was severe.

3.) Which laboratory tests are useful in the diagnosis of HELLP syndrome?

The three main abnormalities found in HELLP syndrome are hemolysis, elevated liver enzymes, and low platelet count. Hematocrit may be decreased or normal and is usually the last of the three abnormalities to appear. Serum haptoglobin levels may decrease before the hematocrit and can be an early indicator of hemolysis. Increased serum transaminase levels (AST and ALT) signify liver injury and can be as high as 4,000 U per liter. However, lower levels are more common and an AST or ALT greater than 40 is considered significant in the diagnosis of HELLP syndrome. Platelet counts can be as low as 6,000, but a platelet count less than 150,000 warrants further investigation.

Proteinuria and increased uric acid have also been used to monitor HELLP syndrome; however recent guidelines have determined that proteinuria and uric acid are only useful in the diagnosis of preeclampsia and not HELLP syndrome. Platelet count is considered to be the best diagnostic indicator of HELLP syndrome. A positive d-dimer with pre-existing preeclampsia has recently been reported to be predictive of patients who will develop HELLP syndrome. (O’Hara 1999, Adukauskiene 2006)

In the current case, liver transaminases on initial laboratory investigations were AST 42 and ALT 63, platelet count was 175, hemoglobin was 13.3. At this point in time, with only minimally elevated liver enzymes and normal platelet count, the diagnosis of HELLP syndrome was questionable. However, further evaluation of fetal/maternal health and monitoring of labs was needed. With an increase in liver enzymes and decrease in platelets in repeat labs the following day, the diagnosis of HELLP syndrome was warranted. We also used increasing uric acid levels as a marker for HELLP syndrome. However, current literature states that uric acid is not a reliable marker in HELLP syndrome and is only useful in the diagnosis of preeclampsia.

LEARNING POINTS/TAKE HOME MESSAGES 3to 5 bullet points

• ALWAYS listen to your patient
• NEVER discount a diagnosis due to atypical presentation
• ALWAYS have a low threshold for further investigation

REFERENCES

• References:
  • Manda, P, E Dorman, F Olagbaiye, and O Akinfenwa. A case report of spontaneous splenic capsular rupture associated with atypical presentation of haemolysis, elevated

