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"Beyond Preeclampsia: Atypical HELLP Syndrome Masquerading as Extensive Deep Vein Thrombosis"

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ABSTRACT
Pregnancy and the postpartum are known to have risk factors for venous thromboembolism (VTE). This is due to pregnancy being a prothrombotic state, exhibiting all components of Virchow's triad: venous stasis, endothelial damage, and hypercoagulability. HELLP (Hemolysis, Elevated Liver enzymes, and Low Platelets) syndrome is a severe and uncommon condition, usually secondary to preeclampsia. However, some cases, known as atypical HELLP syndrome, may not present with hypertension or may develop it very late. This report describes the case of a 23-year-old primigravida at 34 weeks and 6 days of gestation who presented with an acute episode of bilateral lower limb deep vein thrombosis (DVT) with superimposed atypical HELLP syndrome. The patient was managed with an emergency lower segment cesarean section with the placement of an Inferior Vena Cava (IVC) filter. Despite a complex postoperative course, her condition improved, and she was discharged sound and healthy, on postoperative day 23. This case highlights the complexity of managing DVT and atypical HELLP syndrome concurrently in a pregnant patient.
Keywords: *Atypical HELLP Syndrome, venous thromboembolism (VTE), pregnancy*

INTRODUCTION

Pregnancy is a period of significant physiological changes that predispose women to a heightened risk of venous thromboembolism (VTE). The risk of developing a VTE is approximately 5 to 10 times higher in pregnant women compared to their non-pregnant counterparts, a risk that persists for at least six weeks postpartum. This prothrombotic state is attributed to the classic Virchow's triad: hormonal changes and the mechanical compression of the uterus lead to venous stasis, while changes in coagulation factors, such as increased fibrinogen and factors VII, VIII, and X, cause a state of hypercoagulability. Additionally, potential endothelial damage during delivery further contributes to this risk.

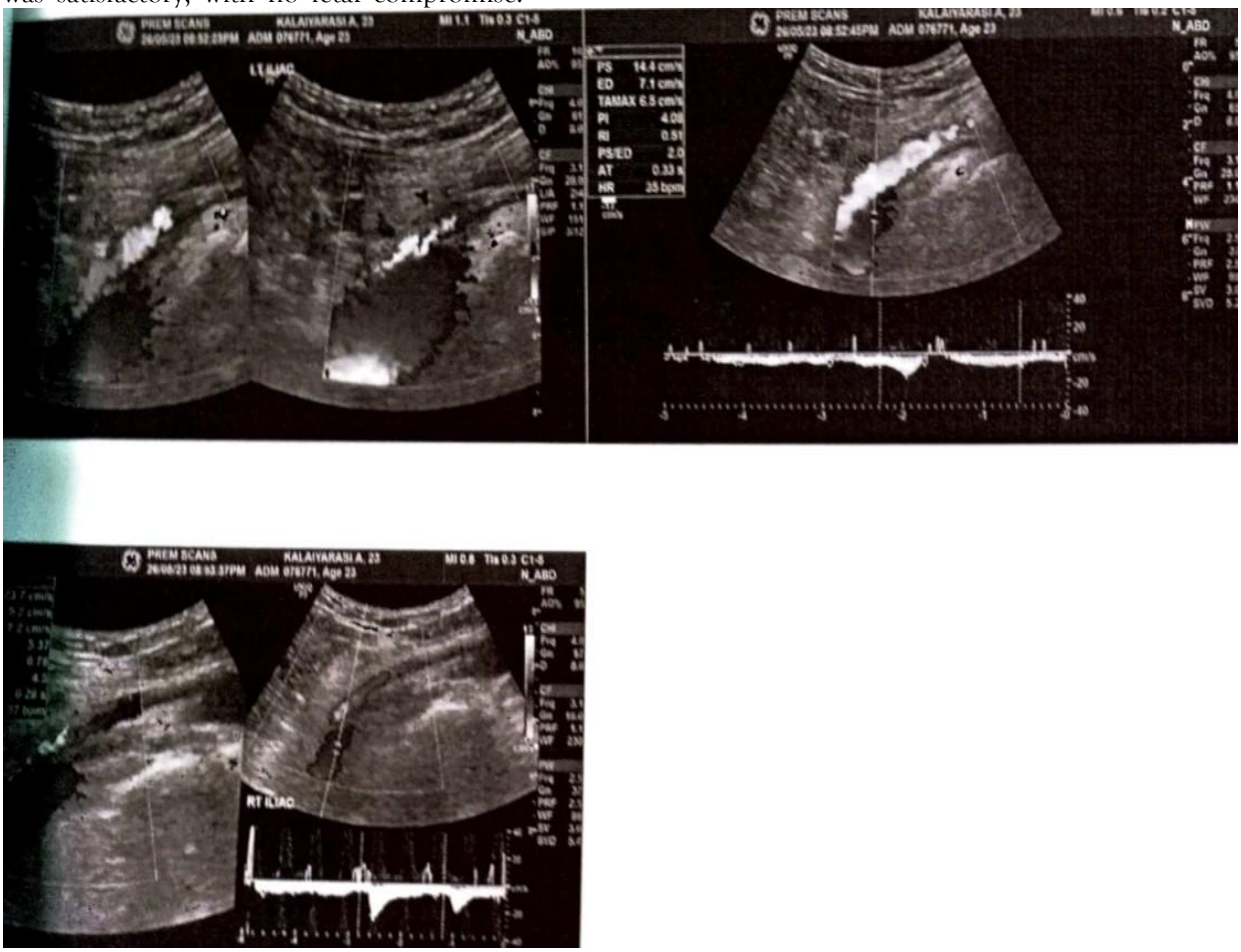
Deep vein thrombosis (DVT), a major component of VTE, can lead to a fatal pulmonary embolism if left untreated. Concurrently, Hemolysis, Elevated Liver enzymes, and Low Platelet count (HELLP) syndrome is a rare, life-threatening obstetric complication, typically considered a severe variant of preeclampsia. However, a less common but particularly challenging presentation, known as atypical HELLP syndrome, occurs when the patient lacks the classic symptoms of preeclampsia, such as hypertension or proteinuria. This absence of typical signs can lead to a significant delay in diagnosis, increasing the risk of maternal and fetal morbidity and mortality.

This case report details the rare co-presentation of extensive bilateral deep vein thrombosis and superimposed atypical HELLP syndrome in a young primigravida. It highlights the significant diagnostic and management challenges posed by the simultaneous occurrence of these two distinct pathologies. We aim to present the clinical course, multidisciplinary management, and successful outcome of this complex case, contributing to the limited literature on this unique clinical scenario.

CASE DETAILS

A 23-year-old primigravida, outside booked, at 34 weeks and 6 days of gestation, presented to OBGYN department, SRIHER, with bilateral lower limb swelling and pain for two days. On clinical examination, the patient was icteric, with bilateral grade 3 pedal edema and tenderness of both the lower limbs. Her blood pressure on receiving was 120/80 mmHg, with all the other vitals being stable. Laboratory investigations showed a hemoglobin of 12.G g/dl and a platelet count of 1.23 lakhs/cu.mm, lower than normal. The coagulation profile was grossly deranged with a serum

fibrinogen of 65.2mg/dl. Liver function tests showed elevated levels of liver enzymes and hyperbilirubinemia with total bilirubin 8.04 mg/dl. Serum creatinine was elevated to 2.1mg/dl, suggestive of acute kidney injury. A color doppler ultrasound of bilateral lower limbs confirmed Deep Vein Thrombosis involving the right and left common femoral veins, extending into the superficial and deep femoral veins, bilateral great saphenous veins, and bilateral posterior tibial veins. The patient was diagnosed with bilateral lower limb deep vein thrombosis with disseminated intravascular coagulation, acute kidney injury, and superimposed atypical HELLP syndrome. Growth scan with fetal doppler was satisfactory, with no fetal compromise.



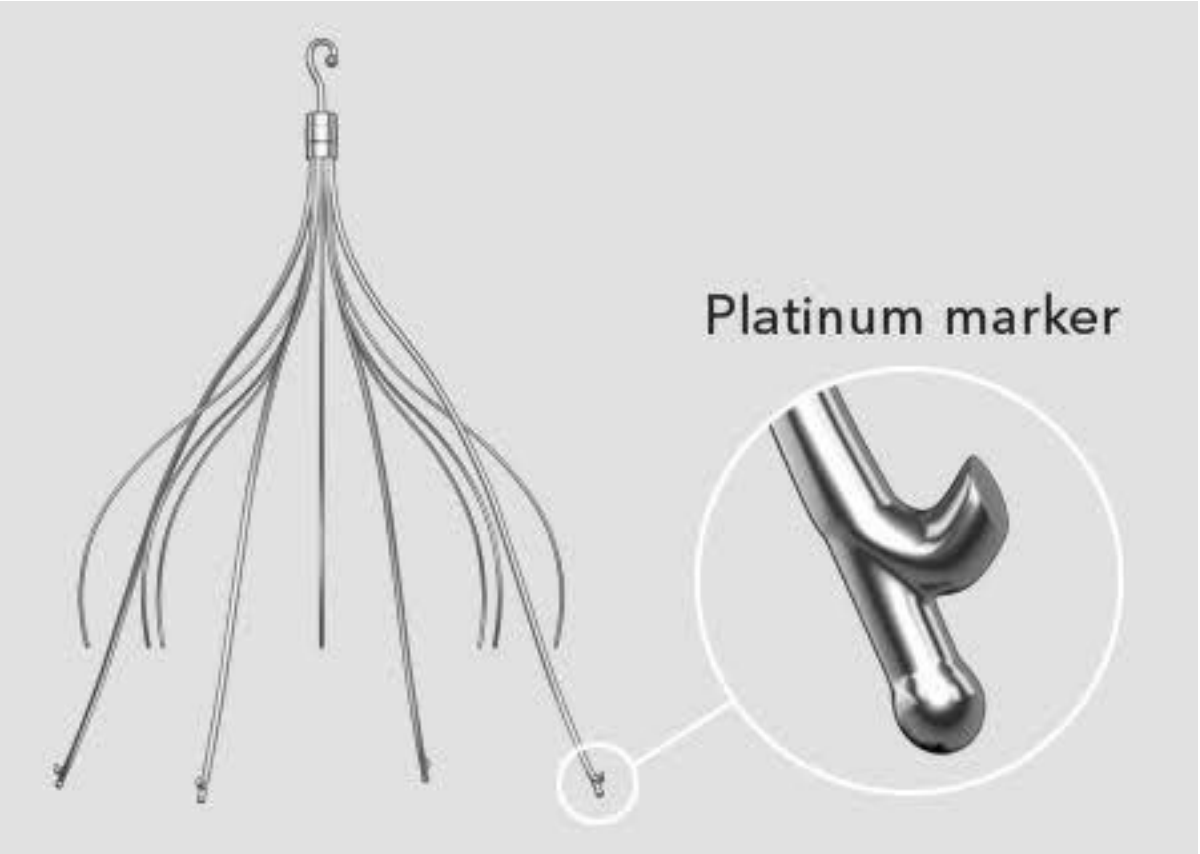
Color doppler ultrasound scan of left lower limb showing venous thrombosis

MANAGEMENT

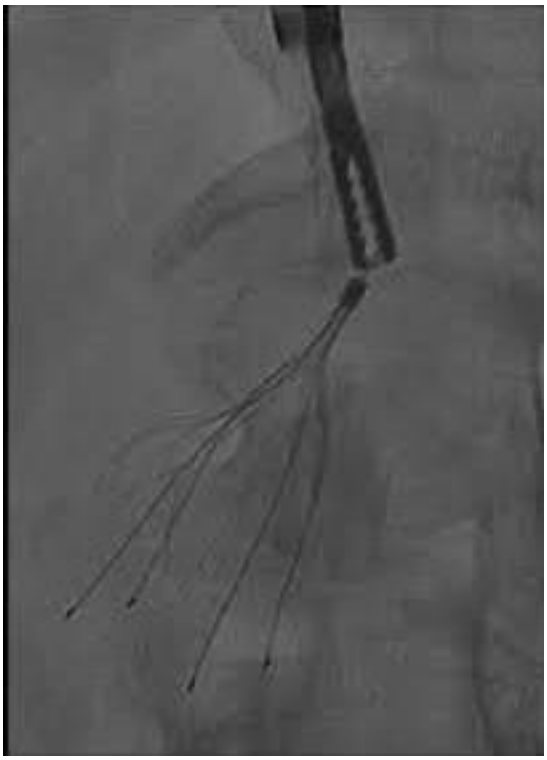
After thorough evaluation, patient was diagnosed to be a case of bilateral lower limb deep vein thrombosis with superimposed atypical HELLP syndrome, with an increased risk of venous thromboembolism. A team of obstetrics, hematology, intervention radiology, cardio vascular surgery, anesthesia and pediatrics were involved for further management. She was taken up for emergency lower segment cesarean section with Inferior vena cava filter placement, under general anesthesia. A Cook's Celect IVC filter was placed at the level of T12 vertebra through internal jugular vein, followed by lower segment cesarean section. Baby cried on birth and had no signs of distress. Postoperatively patient was intubated and shifted to intensive care unit for further management. On postoperative day zero, undue bleeding through vagina was seen, with a further drop in serum hemoglobin, platelets, and fibrinogen. She was transfused with 4 units of cryoprecipitate, 4 units of fresh frozen plasma, and 1 unit of packed red blood cells.

Metabolic acidosis was corrected with bicarbonate and KCL infusions. Patient was extubated on postoperative day one, as liver function tests improved. By the third postoperative day, serum fibrinogen and platelets were improved, and patient was started on subcutaneous unfractionated heparin injections. Postoperatively, serial coagulation profile monitoring was done, and patient was changed from unfractionated heparin injections to oral anticoagulants and low molecular weight heparin injections. As patient's condition improved, with serial measurements of serum INR, anticoagulants dosage was adjusted. She was discharged, sound and healthy, on postoperative day

23. Patient was advised to follow up with weekly serum INR values, for 1 month, followed by, once in 2 weeks for 2 months. A follow-up attempt to remove the IVC filter, 3 months after placement, was unsuccessful and was left in situ.



Cook Celect IVC filter



Placement of IVC filter at level of T12 vertebra

DISCUSSION

Our case report describes the rare co-occurrence of extensive bilateral deep vein thrombosis (DVT) and superimposed atypical HELLP syndrome in a pregnant patient. This dual pathology presented

a unique and complex clinical challenge, highlighting the need for a rapid, multidisciplinary approach to management. The patient's presentation with bilateral lower limb swelling, pain, and jaundice, without significant hypertension, initially masked the severity of her condition. The diagnosis of atypical HELLP syndrome was made based on deranged laboratory values (elevated liver enzymes, low platelet count, and hemolysis) in the absence of the classic hypertensive symptoms. This underscores a critical diagnostic pitfall, as these nonspecific symptoms could easily be misattributed to other conditions, delaying definitive treatment.

Management of Acute DVT in Pregnancy

The management of acute DVT during pregnancy requires a delicate balance between preventing maternal morbidity and ensuring fetal well-being. According to guidelines, low-molecular-weight heparin (LMWH) is the gold standard for anticoagulation due to its inability to cross the placental barrier, which prevents fetal exposure and teratogenic effects. The patient's co-presentation with atypical HELLP syndrome complicated this standard approach, as the associated thrombocytopenia and hypofibrinogenemia increased her risk of hemorrhage. This heightened bleeding risk, combined with the extensive nature of the DVT, justified the decision to perform an emergency cesarean section and simultaneously place an inferior vena cava (IVC) filter.

The Role of Inferior Vena Cava (IVC) Filters

The use of IVC filters in pregnancy is typically reserved for cases where anticoagulation is contraindicated or has failed to prevent recurrent pulmonary embolism (PE). In our patient's case, the extensive bilateral DVT posed a significant and immediate risk of a fatal PE. The IVC filter served as a critical mechanical intervention, acting as a barrier to prevent the thrombi from embolizing to the lungs, thereby providing immediate protection while the underlying coagulopathy was managed. The decision to place a retrievable filter was appropriate, given that the hypercoagulable state of pregnancy is temporary. However, the unsuccessful attempt at filter retrieval three months postpartum highlights a known complication and emphasizes the importance of patient counseling regarding long-term risks associated with these devices.

Atypical HELLP Syndrome and Its Clinical Implications

Atypical HELLP syndrome is a rare variant that can occur without the hypertensive and proteinuria components of preeclampsia. Patients may present with nonspecific symptoms such as malaise, epigastric pain, or, as in our case, jaundice and hematological abnormalities. The underlying pathophysiology involves widespread endothelial dysfunction, microangiopathic hemolytic anemia, and hepatic injury. The definitive treatment is delivery, as the condition often progresses rapidly and can lead to severe complications, including disseminated intravascular coagulation (DIC), acute kidney injury, and liver hematoma or rupture. Our patient's clinical course, including the initial drop in hemoglobin and platelets and deranged coagulation profile post-delivery, is consistent with the progression of the disease and a positive response to delivery as the curative treatment. The need for transfusions of cryoprecipitate, FFPs, and PRBCs underscores the severity of the DIC, which further complicated the management of her DVT.

This case serves as a valuable learning tool, demonstrating that a multi-disciplinary approach involving perinatologists, hematologists, and interventional radiologists is essential for the timely diagnosis and effective management of such complex obstetric pathologies. It highlights the importance of considering atypical presentations of common pregnancy complications to ensure optimal maternal and fetal outcomes.

CONCLUSION

This case report highlights the critical importance of maintaining a high index of suspicion for rare and atypical presentations of obstetric complications. The patient's co-presentation of extensive bilateral deep vein thrombosis and atypical HELLP syndrome is an extremely rare and complex clinical scenario that required a prompt, multidisciplinary, and aggressive management strategy.

The successful maternal and neonatal outcome in this case was a result of several key factors:

1. Early Recognition of Atypical Presentation: The team's ability to diagnose atypical HELLPsyndrome based on laboratory markers, despite the absence of classic hypertensive symptoms, was paramount. This prevented a potentially catastrophic delay in treatment.

2. Strategic Multi-Disciplinary Management: The coordinated care involving obstetrics, hematology, and interventional radiology allowed for a decisive and timely intervention, balancing the competing risks of bleeding (from HELLP syndrome) and thromboembolism (from DVT).
3. Timely Intervention: The emergency cesarean section served as both the definitive treatment for HELLP syndrome and a window of opportunity for the intraoperative placement of an IVC filter. This dual-purpose intervention was critical for managing the patient's immediate risks. This case serves as a valuable reminder that while pregnancy is a state of physiological hypercoagulability, the presentation of DVT and HELLP syndrome can be atypical and severe. It underscores the need for vigilant clinical and laboratory monitoring in all pregnant patients, regardless of blood pressure, to ensure timely intervention and improve maternal and fetal outcomes.

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