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An interesting case of an atraumatic painful swollen limb in a pregnant woman

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Abstract: Phlegmasia cerulea dolens is an uncommon complication of deep venous thrombosis. This is associated with high rates of morbidity if not treated effectively. We present a young lady 13 weeks pregnant with one-day history of left lower limb swelling with pain and discolouration. Bedside ultrasonography revealed thrombosis occluding the common femoral vein and collateral femoral vein. She had history of neonatal alloimmune thrombocytopaenia (NAIT), and had immunotherapy previously. The safest option was to give low molecular weight heparin (LMWH) on an inpatient basis. Anticoagulation with LMWH has been well established as thromboprophylaxis during pregnancy, however, the safety profile of systemic anticoagulation is matter of debate. As highlighted in this scenario the management needs to be tailored on an individual basis. The cause for the extensive deep vein thrombosis could be possibly due to the recent immunoglobulin therapy, undiagnosed prothrombotic state (outwith pregnancy) or the procoagulant state associated with pregnancy.

Keywords: Anticoagulants; Neonatal Alloimmune Thrombocytopaenia; Phlegmasia Cerulea Dolens; Pregnancy

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1. Introduction

Phlegmasia cerulea dolens (PCD) is an uncommon complication of deep venous thrombosis (DVT) in which there is marked swelling of the affected periphery with significant pain and discolouration. It is the clinical triad of acute limb swelling, ischaemic pain, and discolouration (cyanosis). The complications are due to arterial occlusion leading to gangrene formation needing amputation (1). Systemic embolization can lead to pulmonary embolism. Due to its rarity the incidence rate in the context of DVT can only be assessed from clinical studies. The main treatment in PCD is to improve venous patency, decrease the incidence of recurrence as well avoid the post-thrombotic syndrome (2,3). This can be achieved through systemic or local thrombolysis, surgical thrombectomy and mechanical thrombolysis. This case is unique and a clinical challenge as anticoagulation required careful consideration with our patient's obstetric history of neonatal alloimmune thrombocytopaenia (NAIT). Our case highlighted the need for a multidisciplinary discussion for our patient. We highlighted the importance of bedside ultrasonography in clinical practice in the emergency department. Our patient very kindly gave her consent for this article on her clinical condition including her images.

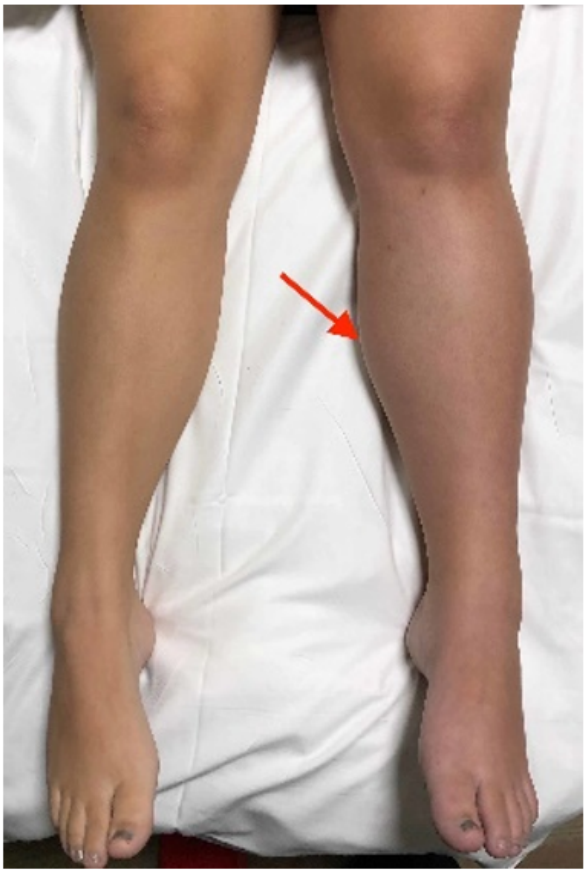

Figure 1 Left lower limb discolouration

2. Case presentation

A young lady in her mid-30s being 13 weeks pregnant self-presented with sudden onset left limb pain over the past 24 hours. The symptoms of pain, swelling, and discolouration was noted to be worsening over this period of time. She had

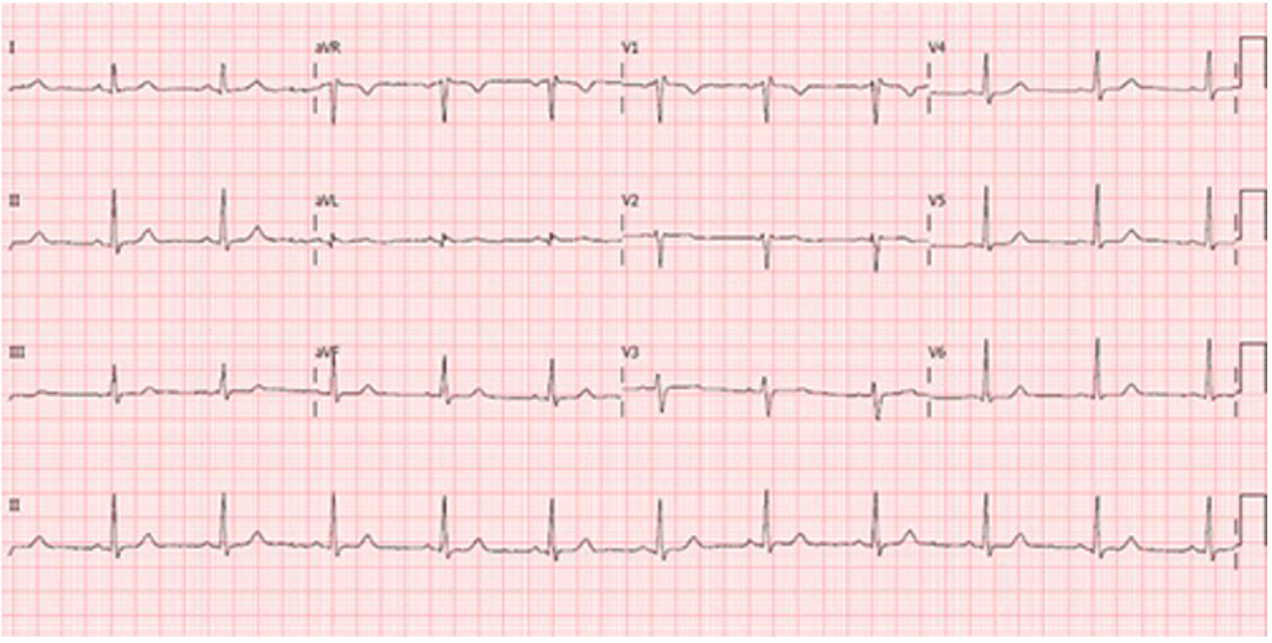

Figure 2 Electrocardiogram of the patient on admission

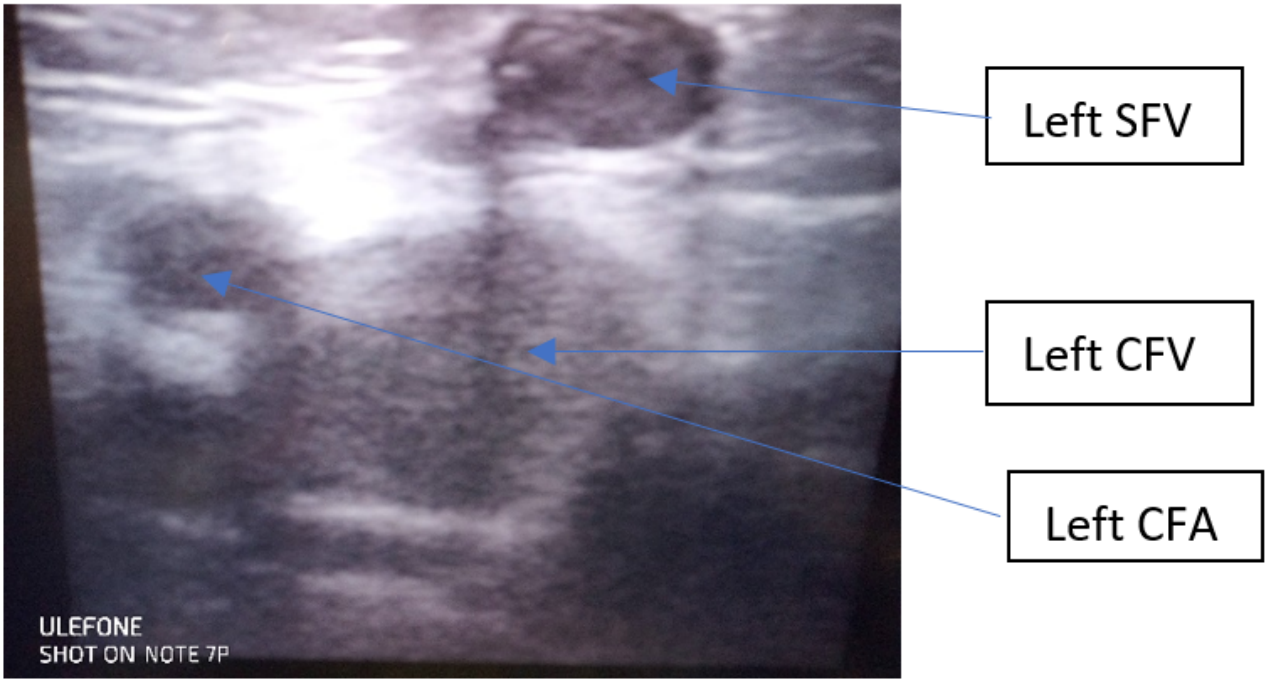

Figure 3 Ultrasound scan of the left groin using linear probe (SFV: Superficial femoral vein; CFV: Common femoral vein; CFA: Common femoral artery)

undergone immunoglobulin therapy the previous day due to obstetric history of neonatal alloimmune thrombocytopaenia (NAIT). There was no additional past medical history or family history apart from the complex obstetric history of NAIT leading to an intracranial haemorrhage with termination of pregnancy previously.

Her vital signs were respiratory rate= 18 breaths/min, oxygen saturation= 95% on room air, heart rate= 95 beats/min, blood pressure= 125/65mmHg and temperature= 36 degrees Celsius. On examination, she was noted to be in discomfort due to the swelling in her left lower limb. She was noted to have normal heart sounds and was noted to have vesicular breath sounds on auscultation. On examination of her peripheries, there was discoloration of the skin as represented in figure 1. All pulses were present on both sides. She did complain of chest discomfort on mobilising with intermit-

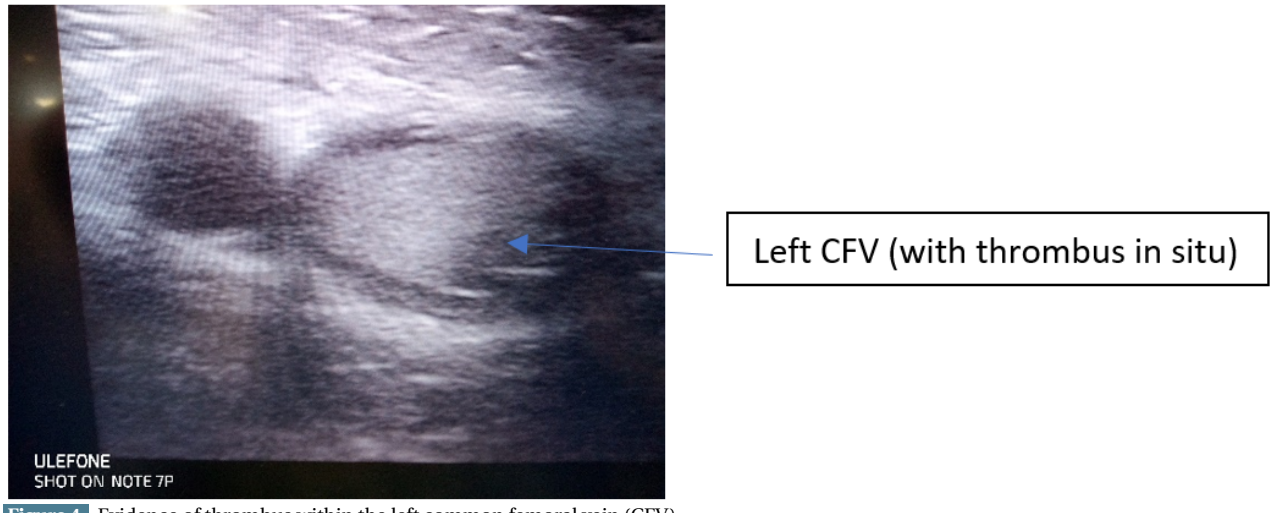

Figure 4 Evidence of thrombus within the left common femoral vein (CFV)

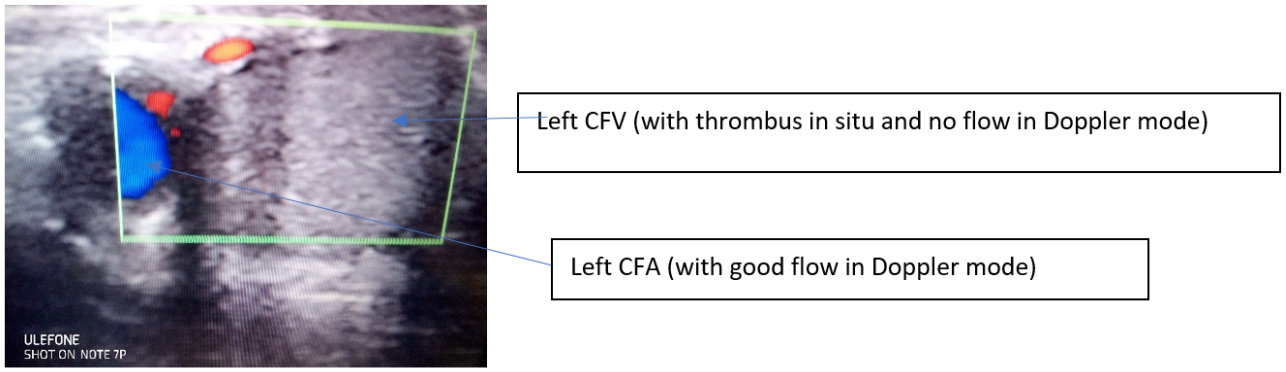

Figure 5 Ultrasound Doppler scan of the common femoral artery (CFA) and common femoral vein (CFV) highlighting the good flow in the left CFA (blue) and complete obstruction in flow in the left CFV (right)

tent self-resolving dyspnoea. She underwent electrocardiogram which indicated incomplete RBBB (Figure 2). Laboratory results were as follows:

International normalised ratio= 1.0 (I.0-1.3); haemoglobin= 13.6 (13.5-17.5 g/dl); white blood cell count= 12.7 (3.5-10.5 x 10^9/L); platelet count= 219 (150-450 x 10^9/L); urea and electrolytes all reported within normal range; D-dimer was noted to be raised as 14000 (upper limit =500).

Due to the clinical findings and concerns, bedside ultrasound scan was carried out to exclude proximal venous thrombosis with possibility of arterial occlusion due to the extensive nature of the limb swelling (Figures 3-5). It confirmed the presence of left sided common iliac vein, femoral vein and popliteal vein thrombosis. There was no deterioration in clinical state following treatment with tinzaparin as an inpatient with antenatal care. She had weekly coagulation screen and full blood count tests to assess the trend.

3. Discussion

The Royal College of Obstetrician and Gynaecology (RCOG) advises that a raised D-dimer is not useful in the diagnosis of venous thromboembolism (VTE) in the pregnant patient (4). There is no diagnostically useful threshold for diagnosis or the ruling out of VTE. In pregnancy and in the post-natal phase, conventional biomarkers have no utility in the diagnosis of VTE (5). The discussion of the treatment options are as follows:

• Endovascular treatment with ultrasound (US) guided percutaneous aspiration thrombectomy: was noted to be a potential treatment option. Whilst this treatment strategy has been proven to be effective in patients with acute and subacute iliofemoral DVT with massive swelling and pain (2) further larger prospective studies are needed to evaluate its true efficacy. The main risks are that of radiation doses with risk of major bleeding.

• Systemic anticoagulation with intravenous unfractionated heparin: this can be considered as an alternative to when low molecular weight heparin (LMWH) cannot be used (6). It is useful in women with acute VTE, massive pulmonary embolism or with severe renal impairment. Close monitoring of the activated partial thromboplastin time (aPTT) levels is needed to ensure that these are within the therapeutic range.

• Low molecular weight heparin (LMWH): use of tinzaparin has been noted to have the better safety profile with regards

to crossing the blood brain barrier, and from maternal to foetal circulation.

• Use of direct oral anticoagulation (DOAC) or vitamin K antagonist (VKA): the evidence from clinical trials of DOACs in pregnancy is not available (7) with current concerns that DOAC can cross the placenta leading to reproductive toxicity. With this current understanding, it would seem unethical to currently investigate the incidence of DOAC-induced teratogenesis.

Having reviewed the above treatment options, in joint discussion with our patient, haematology and obstetrics teams, the informed decision to start LMWH in the form of tinzaparin was started.

Pregnancy alters the haemostatic system leading to a hypercoagulable state which increases as the pregnancy approaches term. The hypercoagulable state contributes to the development of gestational vascular complications and can lead to placental loss. Anticoagulation with LMWHs has been well established as thromboprophylaxis during pregnancy that potentially leads to reduced gestational vascular complications (8). Whilst there are some concerns about heparin induced thrombocytopaenia (HIT), as was in our clinical case, the incidence of HIT is low in approximately 1 in 4000 pregnancies (9). Hence daily monitoring of platelet is not recommended, checking platelet levels every couple of days is sufficient.

NAIT is caused by alloantigens carried on foetal platelets with a significant cause of morbidity and mortality in newborns (10). This is treated using intravenous immunoglobin and prednisolone. The relationship between increased thromboembolic events and intravenous immunoglobin (IVIG) is a topic of debate. Several studies have in fact shown a protective effect of IVIG in the context of atherosclerotic diseases with a reduction of the progression of the atherosclerosis.

4. Conclusion

As highlighted in this scenario, the management of PCD needs to be tailored to the individual patient with investigations to ascertain the underlying cause. The cause for the extensive DVT could be possibly due to the recent immunoglobulin therapy, undiagnosed prothrombotic state (outwith pregnancy) or the procoagulant state associated with pregnancy.

5. Declarations

5.1. Acknowledgment

We would like to thank our patient for kindly allowing us to undertake this article based on her clinical management.

5.2. Authors' contribution

D. Chowdhury was the sole author in the production of this article from conceptualisation to the writing, editing and revising.

5.3. Conflict of interest

The author has not identified any conflicts of interest in undertaking this article.

5.4. Funding

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