Phlegmasia caerulae dolens secondary to pelvic plasmacytoma and left femoral deep vein thrombosis

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ABSTRACT
INTRODUCTION: Phlegmasia caerulae dolens (PCD) is a clinical syndrome caused by venous obstruction leading to peripheral limb ischaemia. It can ultimately lead to venous gangrene, amputation or death in 25% of cases.

PRESENTATION OF CASE: A 52-year-old man with a background of myeloma developed PCD secondary to an obstructing plasmacytoma and left femoral vein deep vein thrombosis (DVT). These were treated with combined radiotherapy and anticoagulation, with resolution of the patient’s symptoms. His recovery was complicated by the development of heparin-induced thrombocytopenia (HIT) and cutaneous vasculitis.

DISCUSSION: Both plasmacytoma and DVT are recognised complications of myeloma. This is, to our knowledge, the first description of these phenomena in combination causing PCD. The combination of venous stasis from the obstructing plasmacytoma and hypercoagulability from the underlying myeloma may have contributed to clot formation. A multifaceted treatment approach was required which aimed at improving venous flow via radiotherapy to the plasmacytoma and dissolving the obstructing clot with anticoagulant therapy.

CONCLUSION: PCD has a high mortality and morbidity. Recognition is important to avoid an incorrect diagnosis of arterial occlusion and inappropriate surgical intervention. Treatment must be focused on removing the offending causes.

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1. Introduction
Phlegmasia caerulae dolens (PCD) is caused by an obstructing venous lesion leading to pain, swelling and ultimately signs of acute arterial ischaemia. This is due to increased compartmental pressures causing secondary arterial compromise. PCD has a high morbidity and mortality. In addition, the clinical presentation of PCD can be confused with acute limb ischaemia, which may lead to inappropriate surgical intervention and patient harm. Despite its severity, management of the condition has not improved greatly over the last few decades. Treatment focuses on anticoagulation of any deep vein thrombosis (DVT) and in select cases intra-arterial thrombolysis. Elevation and fluid resuscitation also have a role in patient management. Severe cases may require limb amputation.

2. Presentation of case
A 52-year-old male with a background of left pelvic plasmacytoma and myeloma presented with acute left leg pain and swelling. On examination, the left leg was cold, swollen with blue discolouration and loss of distal sensation. In addition, distal pulses were absent on Doppler examination. A junior vascular surgeon reviewed the patient and a diagnosis of acute limb ischaemia was made. Therefore, the patient underwent a computerised tomography (CT) angiogram, which demonstrated no arterial occlusion, although did demonstrate a large soft tissue mass encasing the left common iliac vessels. A subsequent ultrasound (US) of the leg demonstrated gross subcutaneous oedema with a non-compressible femoral vein, no colour flow on Doppler and no flow on calf compression consistent with a left femoral DVT. The patient showed no symptoms or signs of pulmonary embolism and his observations were stable.

Following review of a recent magnetic resonance image (MRI) scan (Fig. 1), the pelvic plasmacytoma was implicated in obstructing the venous outflow around the common iliac vein. In view of the signs on examination, US and CT angiogram findings, a diagnosis of phlegmasia caerulae dolens (PCD) was made secondary to pelvic plasmacytoma and left femoral DVT. The patient was commenced on subcutaneous low molecular weight heparin (LMWH), leg elevation and received radiotherapy for the plasmacytoma,
which improved the patient’s symptoms. However, the patient’s recovery was complicated by the development of heparin-induced thrombocytopenia (HIT). He was therefore changed from LMWH to Rivaroxaban with further clinical improvement.

In addition to the development of HIT, the patient began to develop a vasculitic rash on his lower limbs. A dermatologist reviewed the patient and a diagnosis of cutaneous vasculitis was made secondary to myeloma (Figs. 2 and 3). He continued to improve and was discharged home following a four-week period in hospital on long-term anticoagulation with Rivaroxaban.

3. Discussion

PCD literally translates as painful blue oedema. It may result from a large deep vein thrombosis or other occlusive process and causes pain, oedema and signs of peripheral ischaemia. It is differentiated from the less severe phlegmasia alba dolens where collateral vein patency maintains circulation and reduces ischaemia. PCD has a mortality of 25% with an amputation rate of 12–25%. It is more common in females and more commonly affects the left leg. In severe cases, it can lead to venous gangrene, hypovolaemic shock and the need for limb amputation secondary to arterial compromise. This is thought to be secondary to an increase in compartment pressures to 16–17 times normal.

PCD is commonly associated with malignancy, obstructing lesions, thrombophilia and less commonly the placement of inferior caval filters. The initial blockage of the venous system leads to the presenting symptoms of pain and swelling. This then progresses to tissue ischaemia and resulting gangrene. Ultimately, this may lead to massive pulmonary embolism or death in a significant proportion of patients. Our case may well have been multi-factorial, with the combination of plasmacytoma and deep vein thrombosis contributing to an obstructed venous system. Indeed, Virchow’s triad dictates that venous stasis, endothelial injury and hypercoagulability contribute to blood clot formation. Malignant conditions such as myeloma are well known to cause hypercoagulable states, which in combination with the venous stasis that occurred due to the encased iliac veins from the plasmacytoma, may have contributed to colt formation in our case.

Despite advances in technology, little progress has been made in the management of PCD over the last 30 years. Large-scale trials are difficult to conduct. Some success has been shown with the use of anticoagulation, elevation and fluid resuscitation in one small case series. A selected group of patients may also respond to thrombolysis, reducing the need for amputation. In extreme cases, surgical amputation is required. In our case, the plasmacytoma was treated with radiotherapy to reduce soft tissue bulk and improve blood flow in the encased vein. Furthermore, the likely femoral DVT was treated with anticoagulation. However, the onset of HIT required alternative anticoagulation with Rivaroxaban with good success. These newer anticoagulants may offer an alternative treatment to patients with HIT and have been shown to be effective in DVT treatment in a previous large-scale randomised controlled trial.

4. Conclusion

Phlegmasia caerulea dolens is a rare manifestation of an obstructed venous system with a high morbidity and mortality. Diagnosis may be difficult as loss of distal pulses may be confused with acute arterial ischaemia leading to inappropriate surgical intervention. In the case we present here, successful combination treatment was aimed at anticoagulation to reduce clot size and radiotherapy to reduce plasmacytoma bulk. Due to the onset of HIT, successful anticoagulation was achieved with Rivaroxaban.

Conflict of interest

No conflicts on interest or funding received in the production of this case report.

Fig. 1. Magnetic resonance imaging of the pelvis demonstrating plasmacytoma (red arrow) and encased left common iliac vein (blue arrow).

Fig. 2. Patient’s left limb demonstrating residual swelling and features of cutaneous vasculitis (lateral view).

Fig. 3. Patient’s left limb further showing vasculitic rash and residual swelling (medial view).
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None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Dr. Brett Doleman: Group 1 – Involved in management of patient and acquired images for case study. Group 2 – Produced the article. Group 3 – Final approval of the version to be published. Mr. Kumar Abayasekara: Group 1 – Managed patient and diagnosed condition. Group 2 – Revised the article for publication. Group 3 – Final approval of the version to be published. Dr. James Kirk: Group 1 – Guidance on radiological imaging during case study. Group 2 – Revised the article for publication. Group 3 – Final approval of the version to be published.

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