CASE REPORT

Lower extremity compartment syndrome in the setting of iliofemoral deep vein thrombosis, phlegmasia cerulea dolens and factor VII deficiency

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SUMMARY

Acute compartment syndrome requires urgent fasciotomies to prevent irreversible muscle damage. We present a case of massive iliofemoral deep vein thrombosis (DVT) presenting as acute compartment syndrome. A healthy 21-year-old man presented with a 2-day history of worsening left leg pain with swelling and bluish discolouration. Clinical diagnosis of compartment syndrome secondary to phlegmasia cerulea dolens (PCD) was made and he underwent emergency fasciotomies. Postoperative venous duplex confirmed a massive iliofemoral DVT and intravenous heparin was started. Following skin grafting, the patient made a good recovery. Massive iliofemoral DVT is an uncommon cause of compartment syndrome and has been reported in lower limbs, secondary to PCD. Failure to treat early carries a high degree of morbidity, with amputation rates up to 50% and mortality rates between 25% and 40%. It is important to recognise compartment syndrome as an acute presentation of PCD. Urgent fasciotomies can prevent limb amputation and mortality.

BACKGROUND

Acute compartment syndrome can occur following trauma to a limb. Increased pressure within the enclosed fascial space causes compromise to micro-circulation leading to myoneural necrosis.¹ Patients present with excruciating pain out of proportion to the injury, and require emergency fasciotomies to decompress the muscle compartments and to reduce tissue pressure to enable greater arterial inflow.² Failure to recognise and treat this orthopaedic emergency results in irreversible muscle damage and subsequent limb amputation.

We present a case of massive iliofemoral deep vein thrombosis (DVT), presenting as acute compartment syndrome in a previously healthy young man.

CASE PRESENTATION

A 21-year-old fit and healthy man presented with a 2-day history of left leg pain that had initially started gradually but had exacerbated on the day of admission and had spread proximally up the thigh. There was no history of trauma and no risk factors for venous thromboembolism (VTE).

On examination, the left lower limb was swollen with a bluish discolouration, but neither erythema nor induration. The patient was reporting excruciating pain on light touch, and the area was exquisitely tender on passive stretching of the toes and

ankle. The left thigh and gluteal regions were tender and soft on palpation (figure 1).

An initial clinical diagnosis of acute compartment syndrome was made and the patient underwent emergency fasciotomy within an hour. Intraoperatively, the muscles were bulging but contractile. There was neither sign of myonecrosis nor of infection. The following morning, he had a venous duplex to confirm a massive iliofemoral DVT as the cause for the acute compartment syndrome.

He was subsequently reviewed by the haematology team and a diagnosis of factor VII deficiency was made. Owing to the bleeding risk with treatment dose anticoagulation therapy associated with open fasciotomy wounds, an inferior vena cava (IVC) filter was inserted to prevent clot fragmentation and subcutaneous enoxaparin dose limited to 40 mg once daily (figure 2).

Within 48 h, the patient was taken to theatre for a second look at the fasciotomy wounds; the muscles were found to be soft but with significant swelling, therefore the wounds were not closeable. A further venous duplex scan demonstrated no venous return and no collateral flow in the left lower limb, and therefore intravenous heparin was started. The patient yet again returned to theatre, for left popliteal vein cannulation and thrombolysis. Subsequently, a tissue plasminogen activator (TPA) infusion was started and he was transferred to the high dependency unit.

Following extubation, the patient had persistent bleeding from the surgical wounds despite tranexamic acid, and the TPA infusion was stopped. He was transfused 3 units of red blood cells and 5 units of fresh frozen plasma, and returned to theatre the following day for further clot removal and left iliac vein stenting. A CT of the abdomen and pelvis excluded any intrinsic/extrinsic mass causing DVT.

However, the patient subsequently developed right lower limb pain and a venous duplex demonstrated a right lower limb DVT, an extension of iliac DVT to the right femoral vein. The right lower limb compartment syndrome required emergency fasciotomies, which demonstrated healthy muscle groups (figure 3).

A repeat duplex ultrasound scan of the right leg demonstrated improved venous flow throughout the limb. Once the wounds were clean, the patient was transferred to the plastic surgeons for split skin grafting. Following this, he was able to mobilise with crutches and discharged home.

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Figure 1 Patient presenting to the emergency department, with a swollen, painful leg with bluish discoloration.

OUTCOME AND FOLLOW-UP

At 1-year follow-up, the patient was fully weight bearing with a single walking stick due to ankle plantarflexion and is currently awaiting left Achilles tendon lengthening (figure 4).

DISCUSSION

Massive iliofemoral DVT is an uncommon cause of compartment syndrome. This has been reported in the lower limbs, secondary to phlegmasia cerulea dolens (PCD)—an ischaemic variety of DVT due to total or near total thrombotic occlusion of extremity venous outflow.^{3–5}

Iliofemoral vein thrombosis causes complete or partial occlusion of venous outflow from the superficial and deep venous system, resulting in a rapid increase in venous pressure.^{6–7} This results in increased capillary hydrostatic pressure and interstitial pressure resulting in marked interstitial oedema.^{8–10} Within 6 hours, the intravascular pressure can increase up to 16–17 times the normal limits and predispose to compartment

Figure 2 Patient in high dependency unit following left leg fasciotomies and initiation of an inferior vena cava filter.

Figure 3 Bilateral leg fasciotomy wounds following compartment syndrome.

syndrome.¹¹ Compartment syndrome is a clinical diagnosis with pain out of proportion to the injury and exacerbation of pain on passive stretching of the compartments. As our patient was cooperative with the clinical examination, compartment pressure measurement was not required, but can be diagnostic in the case of the unconscious patient and in diagnostic uncertainty.

PCD usually clinically presents with a triad of features such as acute ischaemic pain, oedema and bluish discoloration and is slightly more common among men compared with among women: 1.5:1.¹² In PCD, the left lower limb is three times more likely to be affected possibly due to compression of the left iliac vein by the right iliac artery.¹³ Risk factors include malignancy and hypercoagulable states such as antithrombin III, protein C and protein S deficiency.⁴ Failure to treat early with anticoagulation therapy carries a high degree of morbidity—including compartment syndrome—with amputation rates of up to 50% and mortality rates between 25% and 40%.^{14–16} Chinsakchai *et al* carried out a systematic review of all published

Figure 4 Patient at 12-month follow-up, displaying healthy wounds and regained functional activity.

cases of PCD, which included 62 patients. Approximately 18% (n=11) of these patients underwent emergency fasciotomies for acute compartment syndrome, with over half (n=6) requiring subsequent limb amputation. Thirty-six per cent of patients (n=4) improved following fasciotomies but one patient died due to multiorgan failure.¹⁷

Owing to the rarity of PCD, there is currently no consensus on its management. Options include systemic anticoagulation, thrombolytic therapies, catheter-directed thrombectomy, pharmacomechanical thrombectomy and surgical thrombectomy.¹⁷ However, early identification and treatment of PCD can prevent further propagation of thrombus, which may prevent compartment syndrome.^{14 15} For example, Bhatt *et al* reported a case of PCD in a 71-year-old African man with no history of VTE and no underlying coagulation disorder. The patient presented with right leg pain, swelling and blue discolouration to the foot. Sonographic examination revealed a complete deep venous thrombosis extending from the right common femoral vein to the popliteal vein. Subsequently, he responded well to intravenous heparin and oral coumadin and was discharged.¹⁶

Semba and Dake¹⁸ reported a 40% risk of thrombus propagation despite the patients being on anticoagulation therapy. Another study reported a 5.5% overall rate of VTE recurrence during the initial 3 months of anticoagulation therapy.¹⁹

Junk and Shah recently reported a case of PCD and factor V Leiden. A 16-year-old boy with a strong family history of DVT presented with a week history of progressive right leg pain, swelling and discolouration, a week after a minor fall. Ultrasound followed by contrast CT revealed a large occlusive DVT extending from the calf proximally up to the IVC. Heparin followed by catheter-directed thrombolysis with TPA and balloon angioplasty was performed, but clot reaccumulation occurred, so a right common iliac and femoral vein stent was placed. Genetic testing later confirmed factor V Leiden. The patient made a full symptomatic recovery.²⁰

Hawkins *et al* reported a case of a 54-year-old man who had recently undergone a right hemicolectomy for Dukes C colorectal adenocarcinoma. Preoperatively, he was started on warfarin therapy and an IVC filter for a left subclavian venous thrombosis. He later presented with a 4-day history of rapidly worsening left leg pain with an oedematous tender leg. Urgent fasciotomies were performed for impending compartment syndrome. Postoperative ultrasound demonstrated extensive thrombosis in the popliteal vein, extending into iliac veins, and the patient was diagnosed as having PCD with impending compartment syndrome. This case highlights that underlying malignancy and thrombosis are major risk factors for PCD, and that anticoagulated patients can still possess a risk of thrombosis.²¹

Vanfleet *et al* report a case of compartment syndrome secondary to a popliteal vein thrombosis in a healthy 36-year-old woman who presented with acute-onset calf swelling of 12 h history followed by tingling and numbness. She had a history of neither VTE nor bleeding disorders, no recent immobilisation and no trauma. Clinical examination revealed compartment syndrome of the right lower limb and the patient underwent urgent fasciotomies. A postsurgery venogram revealed a thrombus in the right popliteal vein; heparin was started and no long-term functional impairment of the leg occurred.²² Similarly, Veltchev *et al* described a case of PCD in a 71-year-old woman. The patient developed sudden onset right leg pain, swelling and cyanosis during her hospital admission for diabetes mellitus. A diagnosis of PCD was suspected and intravenous heparin started. The patient required fasciotomy and necrectomy of superficial areas and was switched to enoxaparin due to

heparin-induced thrombocytopenia. Despite anticoagulation therapy, the leg became more gangrenous requiring a lower limb amputation.²³

Learning points

- It is important to recognise compartment syndrome as an acute presentation of phlegmasia cerulea dolens.
- Patients with compartment syndrome present with pain out of proportion to the injury and on passive stretching of the compartment.
- Failure to recognise and treat compartment syndrome early with emergency fasciotomies can lead to limb amputation or mortality.

Competing interests None declared.

Patient consent Obtained.

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