Radiology Case Reports 16 (2021) 631–637

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journal homepage: www.elsevier.com/locate/radcr

Case Report

Successful thrombectomy and thrombolysis of deep vein thrombosis in a patient with acute phlegmasia cerulea dolens with May-Thurner anatomy ☆

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ARTICLE INFO

Article history:
Received 3 November 2020
Revised 10 December 2020
Accepted 11 December 2020

Keywords:
Phlegmasia cerulea dolens
Thrombectomy
Thrombolysis
May-Thurner
Interventional

ABSTRACT

Phlegmasia cerulea dolens (PCD) is a rare and important diagnosis to consider in the setting of a painful, swollen, and cyanotic lower extremity. We report a 59-year-old female diagnosed with PCD 3 days status post extended head and neck surgery with additional imaging findings concerning for May-Thurner anatomy. This case presentation is pathognomonic for PCD. Risk factors, pathogenesis, and management are described.

Introduction

Phlegmasia cerulea dolens (PCD) is a rare, life-threatening, massive thrombosis of the deep venous system. Recognizing PCD as early as possible is crucial in the management of the disease, as the prognosis rapidly deteriorates with time. Complications include pulmonary embolism (PE), amputation, and death [1]. PCD presents with a triad of swelling, ischemic pain, and cyanosis of the affected extremity. PCD generally involves the lower extremity, more common on the left, although bilateral PCD has been reported. Our patient presented with pathognomonic PCD with superimposed compression of the left common iliac vein by the right common iliac artery (May-Thurner anatomy) as revealed on venography.

Case report

Here we present a patient who underwent a left facial nerve exploration, left sterno-omohyoid free flap transfer with

☆ Competing interest: The authors have declared that no competing interests exist.

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https://doi.org/10.1016/j.radcr.2020.12.035

Fig. 1 – Photograph of the patient's lower extremities demonstrating swelling and color change of her left leg.

masseteric nerve graft performed for left hemifacial palsy. During this 15-hour surgery the patient's left leg was not placed in the sequential compression boot in anticipation for a sural nerve graft. On postoperative day 3, the patient presented to the emergency department with approximately 8-12 hours of increased pain, swelling, and color change of her left lower extremity. She described the pain as progressive and beginning in the lower left leg and moving up to involve her thigh and groin. Physical examination showed an edematous and cyanotic left lower leg with weak but palpable pulses (Fig. 1). Bedside ultrasound at presentation showed thrombus within the popliteal venous system (Fig. 2). The patient was started on unfractionated heparin and multiphase computed tomography (CT) of the abdomen and pelvis with lower extremity runoff was performed.

Multiphase CT of the lower extremities showed extensive thrombus extending from the inferior vena cava (IVC) to the left calf veins (Fig. 3). In addition, there was delayed contrast enhancement of the left lower extremity arterial system when compared to the right and extensive circumferential edema of the soft tissues of the left lower extremity (Fig. 4). Compression of the left common iliac vein by the right common iliac artery (May-Thurner anatomy) was suggested at the time of CT.

Following CT, the patient was consented for IVC filter placement and follow-on left lower extremity venous thrombectomy and lysis. Under moderate sedation the patient was initially positioned supine and a Cook Celect (Cook, Bloomington, IN) IVC filter was placed via a right internal jugular vein approach (Fig. 5). Once the IVC filter was placed, the patient was repositioned prone, and the left lower extremity was prepped. Access to the left small saphenous vein was achieved using a 5Fr micropuncture system under ultrasound guidance, which readily traversed the saphenopopliteal junction into the popliteal vein. A Glidewire Advantage wire (Terumo, Somerset, NJ) was utilized and the micropuncture sheath was exchanged for an 8 Fr 11cm

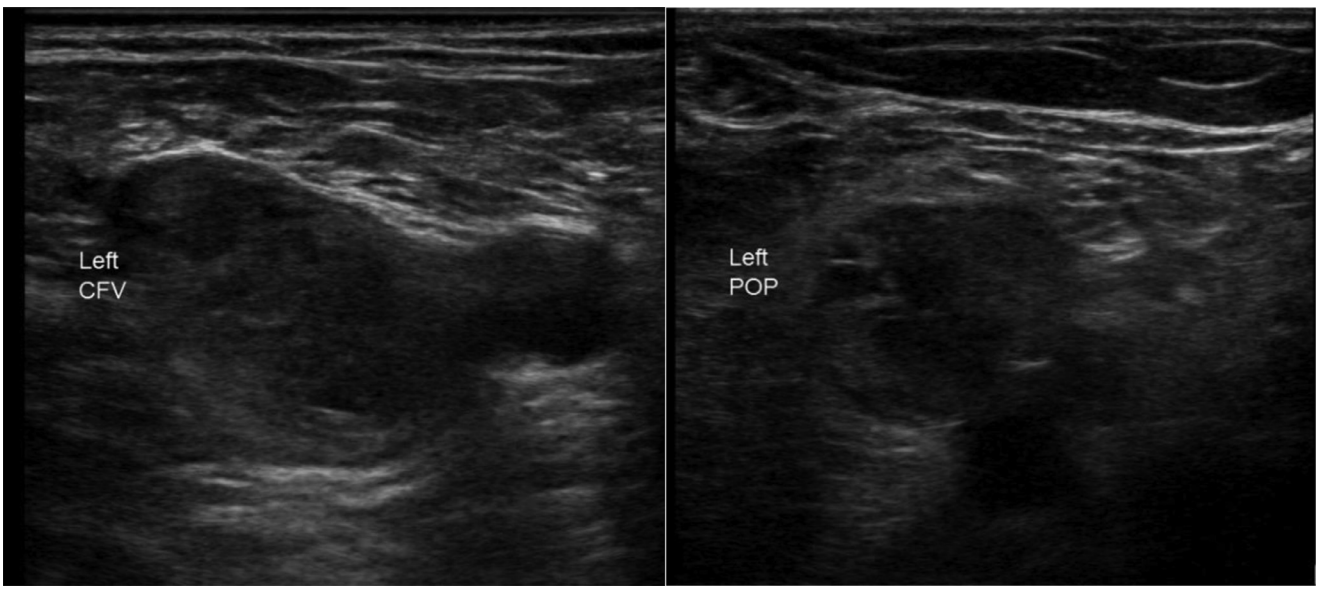

Fig. 2 – Point of care ultrasound of the left common femoral vein (CFV) and popliteal (POP) vein showing noncompressible echogenic thrombus filling the vessel lumen.

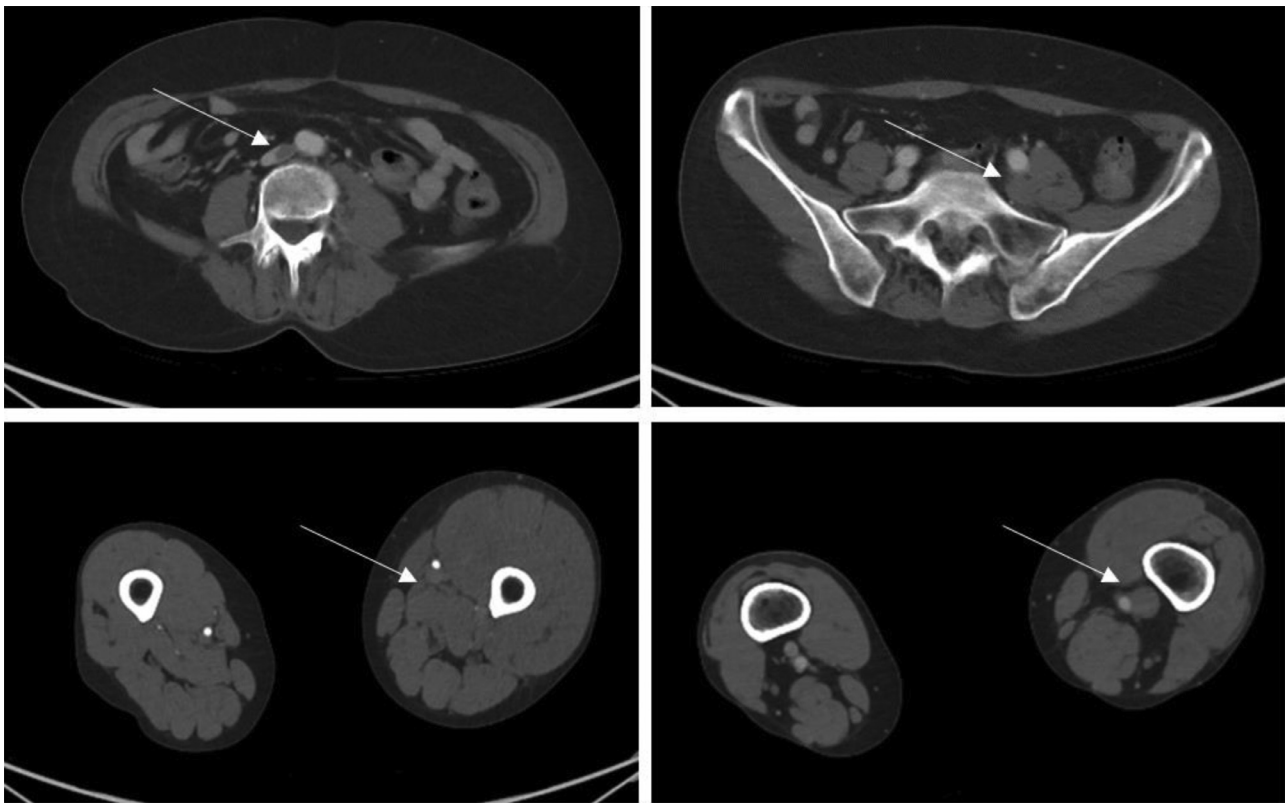

Fig. 3 – Axial contrast enhanced CT in the venous phase demonstrates eccentric nonocclusive thrombus in the IVC and occlusive thrombus extending from the left common iliac vein to the left popliteal vein.

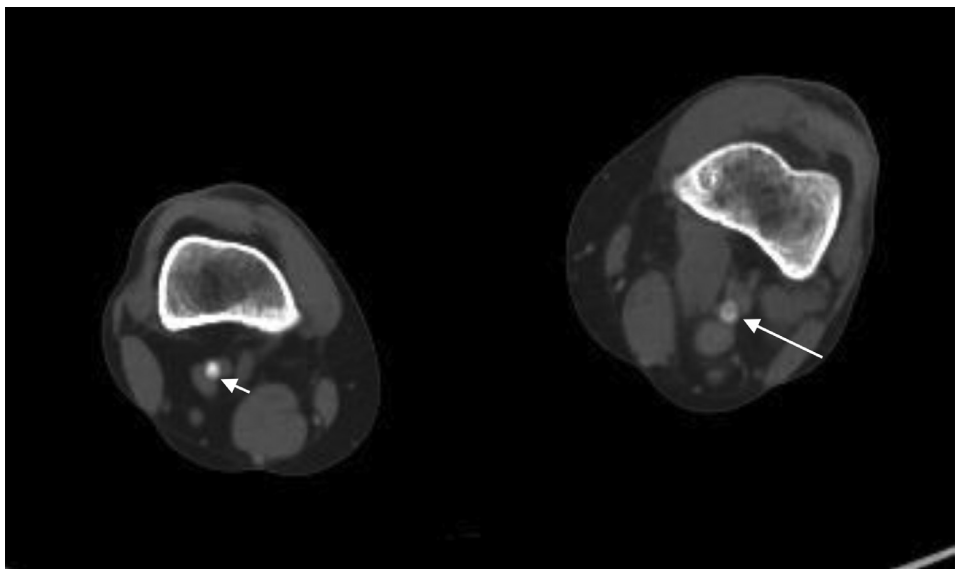

Fig. 4 – Axial contrast enhanced CT in the arterial phase demonstrates delayed opacification of the left popliteal artery (long arrow) in comparison to the right (short arrow) as well as asymmetrically enlarged left lower extremity circumference.

vascular sheath (Terumo, Somerset, NJ). Suction was applied to the sheath after insertion, and significant thrombus was extracted. Initial venography revealed thrombotic occlusion of the popliteal and femoral vein just above the sheath tip, with no contrast transit above that level (Fig. 6). The Glidewire Advantage wire was then advanced into the IVC. As the patient was already heparinized, a Penumbra CAT D suction thrombectomy catheter (Penumbra Inc., Alameda, CA) was selected and introduced. Beginning at the popliteal vein, suction thrombectomy was performed in an ascending manner to the level of the left common femoral vein. A significant amount of fresh thrombus was extracted, and venography revealed

Fig. 5 – Cavogram (Left) demonstrating eccentric filling defect in the low IVC (arrow) and occlusive filling defect in the left common iliac vein (arrowhead) consistent with thrombus. Appropriately positioned infra-renal IVC filter (Right).

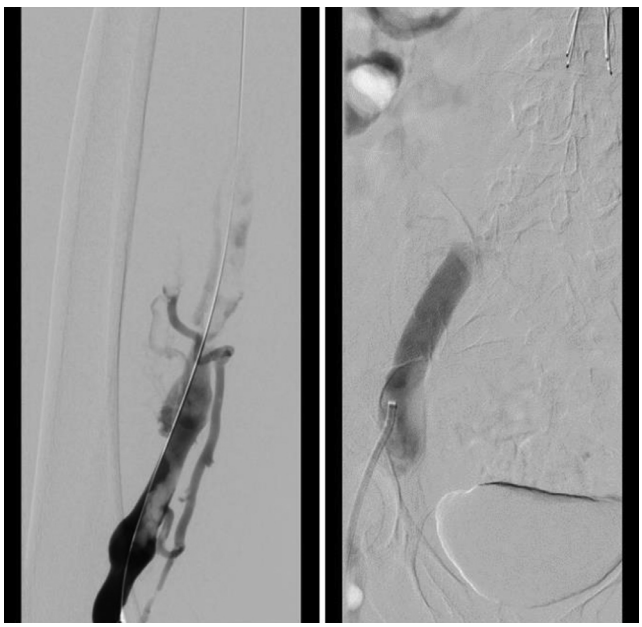

Fig. 6 – (Left) Digital subtraction angiogram (DSA) demonstrates extensive filling defects within the popliteal vein extending into the common femoral vein. (Right) DSA of the left external iliac and common iliac veins following initial thrombectomy demonstrates persistent occlusive thrombus without opacification of the IVC.

significant clearance of thrombus within the femoral veins, but no contrast transit through the iliocaval veins was seen (Fig. 6). An 85 cm penumbra CAT 8 catheter was then selected for additional thrombectomy of the left iliac veins. This was successful to the level of the most proximal left common iliac vein and IVC, including successful thrombectomy of the clot previously noted within the IVC. A total thrombectomy volume of 400 mL were extracted (Fig. 7), however, thrombus burden remained at the left iliocaval junction causing significant stasis. Next, an 8 French AngioJet Zelante (Boston Scientific, Marlborough, MA) DVT catheter was inserted and additional pharmacomechanical thrombectomy of this proximal iliac segment was attempted. Unfortunately, there was minimal improvement in the appearance of the iliac thrombus despite multiple attempts, and further suction thrombectomy was aborted due to concern for iatrogenic hypovolemia. The decision was then made to treat the residual thrombus with overnight catheter directed thrombolysis. A 5 French 135 cm Cragg McNamara infusion catheter (Medtronic, Minneapolis, MN) with a 50 cm infusion length was chosen, allowing infusion along the entire treated length of the patient's leg. Tissue plasminogen activator was initiated at 1 mg per hour overnight through the catheter, and heparin was administered at 400 units per hour through the side port of the popliteal vein sheath. The patient was admitted to the intensive care unit for overnight observation (Fig. 8).

Within hours of the mechanical thrombectomy procedure the patient reported significant improvement in pain. The following morning after approximately 20 hours of overnight catheter directed thrombolysis the patient denied any lower extremity pain and color changes had resolved. The existing 8 Fr sheath was utilized to perform a repeat venogram which showed a widely patent popliteal and common femoral vein with residual clot within the left common iliac vein near the bifurcation of the IVC (Fig. 9). Repeat suction thrombectomy resulted in decreased clot burden, however, due to a persistent filling defect near the IVC bifurcation, a single round of angioplasty was performed at the persistent luminal narrowing without significant improvement in flow. Completion imaging revealed restoration of flow in the iliofemoral venous segment to the level of the popliteal vein with the exception of a small remaining thrombus at the IVC confluence (Fig. 9). Postprocedurally, and in conjunction with the facial surgery team, the patient was started on rivaroxaban therapy.

Follow up

The patient was seen approximately six weeks postoperatively for planned IVC filter removal and repeat venogram. The IVC filter was removed without incident via a right internal jugular approach. Repeat venogram of the distal IVC and left common iliac vein showed no evidence of persistent thrombus (Fig. 10). Intravascular ultrasound (IVUS) demonstrated significant compression of the left common iliac vein by the right common iliac artery, diagnostic of May-Thurner syndrome (images not shown). Based on IVUS imaging a 16 mm × 60 cm Venovo stent (Bard, Tempe, AZ) was selected and advanced into appropriate position based on fluoroscopic

Fig. 7 – (Left) Penumbra Engine vacuum canister with large amount of thrombus collected in the filter (circle). (Right) Pieces of soft thrombus measuring approximately 15 cm in total length.

Fig. 8 – Photograph of the patients' lower extremities demonstrating decreased d swelling and improved color of the left lower extremity following mechanical thrombectomy and approximately 20 hours of catheter directed thrombolysis. An 8-Fr vascular sheath is seen in the left popliteal vein through which the second round of mechanical thrombectomy was performed.

landmarks, with the proximal portion extending into the IVC. The stented vein was treated with a 14 mm angioplasty balloon with good radiographic result. Repeat IVUS demonstrated the minimum diameter of the vein after treatment was 10 × 15 mm. Repeat venogram demonstrated brisk flow through the treated segment into the IVC (Fig. 11).

Discussion

PCD generally involves the lower extremities with 45% occurring on the left lower extremity, 29% on the right, and 26% bilaterally [1,2]. These thrombi can present anywhere from the IVC distally. Reported mortality ranges from 25% to 40%, depending on the stage of the disease, underlying causes, and patient risk factors. PCD can lead to significant morbidity and even death if appropriate management is not urgently performed. A serious complication of PCD is pulmonary emboli. In the literature, PE has been associated with a mortality rate of 50% [1], with an incidence between 12% and 40% [3].

If the main and collateral drainage of the limb is involved, hydrostatic pressure within the vessel overcomes the oncotic pressure and fluid seeps into the surrounding tissue, leading to swelling [4]. The increase in pressure results in decreased blood flow to the affected areas, inducing ischemia. The ischemia leads to increased vascular permeability with more fluid escaping the vessel. The continuation of this process may lead to the development of secondary compartment syndrome, requiring fasciotomy to relieve the pressure. Thrombus extension to the capillaries often marks irreversible progression of the disease and marks the beginning of irreversible ischemia and venous gangrene [5]. Once this occurs, amputation of the limb is required, and the goal of treatment

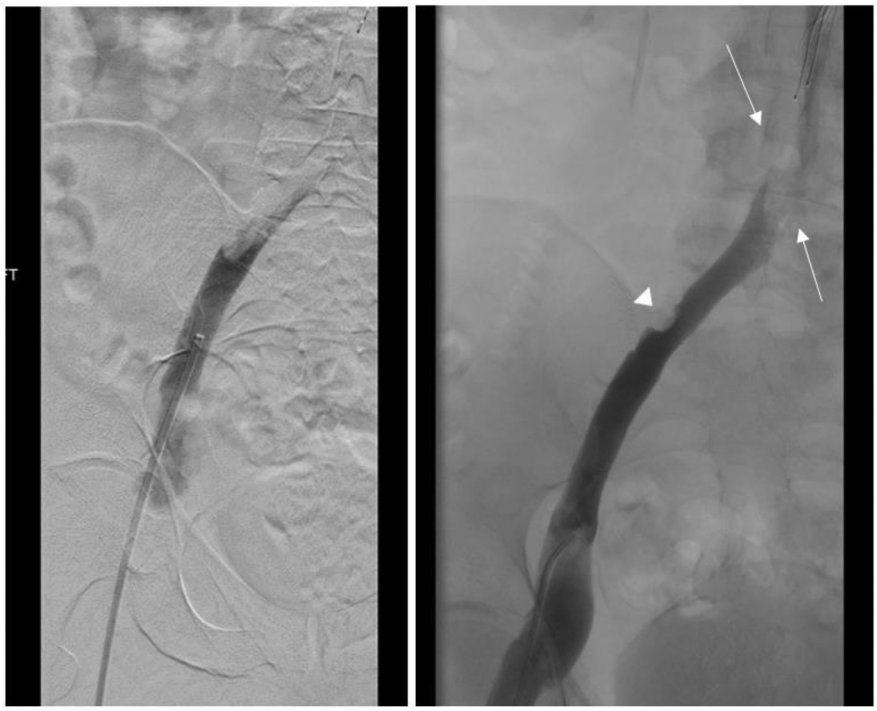

Fig. 9 – (Left) DSA demonstrating persistent thrombus in the left common iliac vein following mechanical thrombectomy and CDT. (Right) Band of nonopacified common iliac vein (arrows) corresponds with traversing right common iliac artery seen on CT. Findings likely represent May-Thurner anatomy. A small amount of residual thrombus is noted in the common iliac vein (arrowhead).

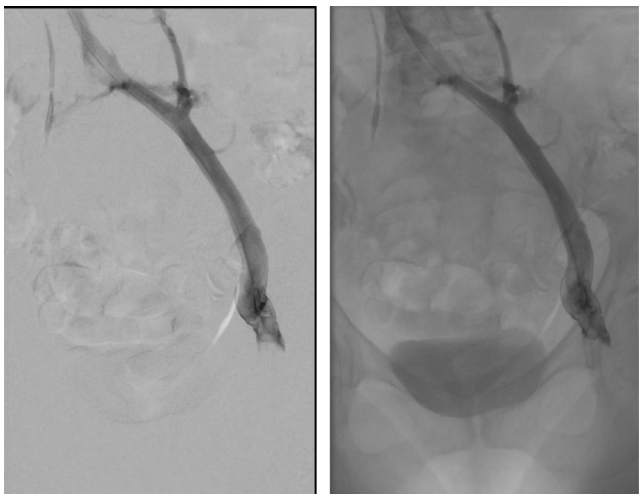

Fig. 10 – DSA of the left external and common iliac veins which are patent with a subtle narrowing of the proximal left common iliac.

is to preserve as much of the limb as possible. The extent of the amputation required will vary among cases, with mortality ranging from 22% to 68%, with the higher percentages belonging to patients requiring above-the-knee amputations [3].

Doppler ultrasonography is recommended as the diagnostic modality with CT also being used, as it was in the case of our patient [6]. Clinical presentation will include swelling, acute pain, and pathognomonic cyanosis of the affected extremity. Diminished pulses, blistering, and neurological deficits may also be present, but are not required for the diagnosis [1]. The most common risk factors for PCD include malignancy, hypercoagulable state, venous stasis, contraceptive agents, and May-Thurner syndrome, with 16% of cases being reported in patients lacking identifiable risk factors. In the case presented, the development of PCD is likely attributable to significant venous stasis from prolonged surgery accentuated by underlying anatomical compression of the left common iliac vein.

As of now, no treatment has been established as a standard of care. Thrombolysis with tissue plasminogen activator and heparin [3,7,8], thrombectomy [1,3], or a mix of both have been reported in the literature with success. Venous thrombectomy alone has reported improvement in 55% of cases, with another 22% requiring amputation. Thrombolysis alone reported 78% improvement with 14% requiring amputation [1]. It should be noted that the severity of cases may have some impact on the choice of intervention, with more severe cases justifying more invasive options. With the advent of minimally invasive techniques, thrombectomy has become more popular treatment option for rapid dissolution of the clot. Historically, cases requiring surgical intervention, fasciotomy or open thrombectomy, are associated with greater mortality, most likely due to the extensive nature of the disease which required the intervention. Minimally invasive procedures, as reported in this case report, offer rapid dissolution of thrombus with a much lower risk profile [9]. It should be noted the

Fig. 11 – DSA of the left external and common iliac veins after stenting which show a widely patent iliac venous system with brisk flow demonstrated on real-time fluoroscopy.

risk of PE from catheter manipulation has been described in the literature and therefore the use of a retrievable IVC filter is recommended, as was the case in our patient [10]. Minimally invasive procedures offer a relatively new approach to the treatment of PCD and therefore lack large sample size studies. In any case, emergent treatment must be initiated quickly to reverse the progression of the disease.

Early recognition and intervention, as achieved with this patient, is important in the management of PCD. We believe the presentation and management reported here are generalizable to most cases of early PCD. We hope this case report will contribute to the expanding material on this rare disease.

Disclaimer

The views expressed in this article are those of the authors and do not reflect the official policy of the Department of the Army, the Department of Defense, or the U.S. Government.

Patient consent

After reviewing the case, informed written consent was provided by the patient in the case titled "Successful thrombectomy and thrombolysis of deep vein thrombosis in a patient with acute phlegmasia cerulea dolens with May-Thurner anatomy."

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