Phlegmasia cerulea dolens secondary to an aortoiliac aneurysm

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ABSTRACT

Phlegmasia cerulea dolens is an uncommon entity. We present a case of phlegmasia cerulea dolens secondary to an aortoiliac aneurysm that compressed the common iliac vein. Catheter-directed thrombolysis was not considered to be a suitable option, because the patient needed an urgent fasciotomy. The aneurysm was treated with a bifurcated stent graft and the thrombosed veins were opened with pharmacomechanical thrombectomy and recombinant tissue plasminogen activator. The reopened iliac veins, including an aneurysmal external iliac vein, were stented and fasciotomy was performed. Pharmacomechanical thrombectomy can be performed with a low dose of recombinant tissue plasminogen activator and allows for subsequent surgery. (J Vasc Surg Cases and Innovative Techniques 2019;5:278-82.)

Keywords: Phlegmasia cerulea dolens; Pharmacomechanical thrombectomy; Abdominal aortic aneurysm; Venous obstruction; Compartment syndrome

Phlegmasia cerulea dolens (PCD) is an uncommon entity. Clinical signs include a sudden presentation of pain, edema, and bluish discoloration of the extremity owing to a massive deep venous thrombosis that impedes venous outflow and may result in venous gangrene, shock, and loss of limb or life.[1] Peripheral pulses may be present on presentation, but is likely to be absent if the disease has progressed to venous gangrene.[2] PCD is associated with malignancy, venous stasis, and a hypercoagulable state.[2] Catheter-directed thrombolysis (CDT) has emerged as the treatment of choice, but may be not be feasible if synchronous surgery is needed. We present a case of PCD where CDT was contraindicated owing to the need for adjunctive emergency surgery. The patient has consented to publication.

CASE REPORT

A 68-year-old man with known hypertension was admitted with a 1-hour history of intense pain and massive swelling of the left leg. The leg was cold, bluish in color, and motor function and sensation were reduced. The ankle-brachial index was normal.

Contrast-enhanced computed tomography (CT) scans in arterial and venous phases revealed an abdominal aortic aneurysm and a left common iliac artery aneurysm measuring 54 and 45 mm, respectively and a left hydronephrosis (Fig 1, *A-C*). Furthermore, a CT scan revealed edema and dilated and thrombosed left common and external iliac and femoral veins (Fig 1, *D-F*). The left external iliac vein was aneurysmal (39 mm) and the left common iliac vein was compressed by the aortoiliac aneurysm, several enlarged collateral veins were present.

The clinical diagnosis was PCD secondary to an aortoiliac aneurysm compressing the common iliac vein and causing extensive venous thrombosis.

Intervention was commenced with the patient in general anesthesia. The aortoiliac aneurysm was treated percutaneously with a standard bifurcated stent graft after coil embolization of the left internal iliac artery (Fig 2).

With the patient in the supine position, the patent popliteal vein was punctured and an 8F introducer inserted. The thrombosed veins were reopened with pharmacomechanical thrombectomy using the CLEANER XT Rotational Thrombectomy System (Argon Medical Devices, Frisco, Tex) combined with recombinant tissue plasminogen activator (tPA) (Alteplase, Boehringer-Ingelheim, Ingelheim am Rhein, Germany; Fig 3, *A* and *B*). A small amount of tPA was infused into a segment of the thrombus and after a delay of 2 to 3 minutes the cloth was macerated by the rotating sinusoidal wire of the CLEANER XT, the procedure was stepwise repeated cranially until the veins were reopened. The total dose of tPA was 10 mg. The compressed and thrombosed common iliac vein was predilated to a diameter of 14 mm with an ATLAS PTA Dilatation Catheter (Bard Peripheral Vascular Inc, Tempe, Ariz). Subsequently, the common and external iliac veins were covered by in total four Zilver Vena Venous Self-Expanding Stents (Cook Medical LLC, Bloomington, Ind) with a diameter ranging from 14 to 16 mm (Fig 3, *C* and *D*). The aneurysmal part of the external iliac vein was covered by a dual layer of Zilver Vena stents. Finally, a percutaneous left nephrostomy and medial and lateral fasciotomy of the calf and thigh was performed.

Anticoagulation was commenced with intravenous heparin. Creatinine kinase spiked to 59,915 U/L, he developed anuric renal failure and was on continuous venovenous hemodialysis for

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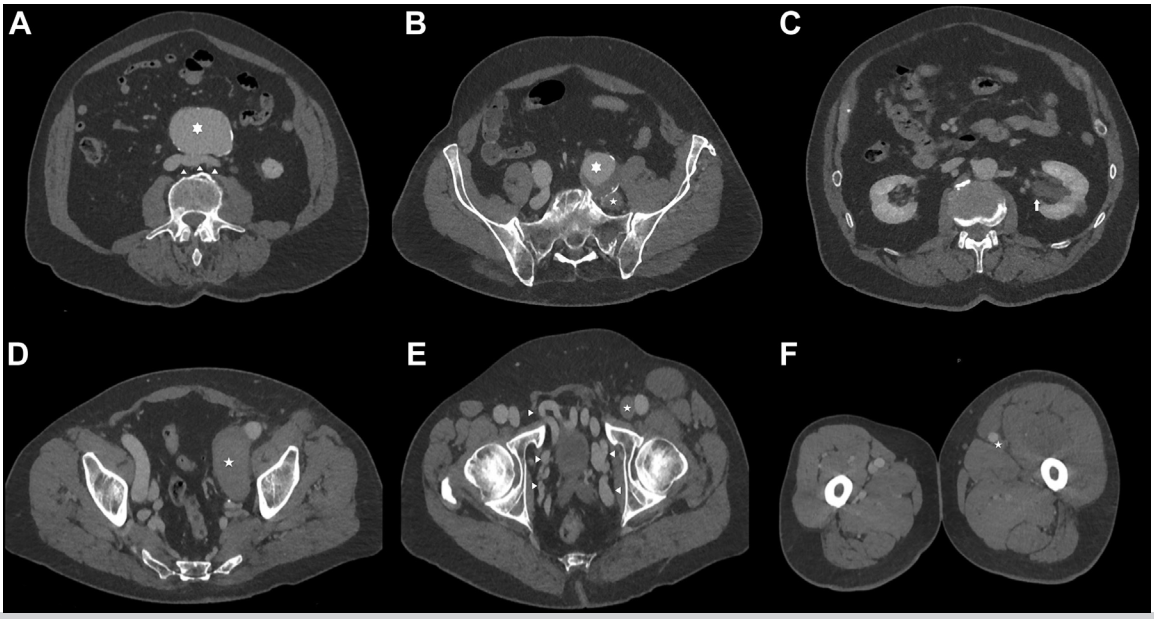

Fig 1. Axial contrast-enhanced computed tomography (CT) scans the in venous phase from the level of the kidneys to the proximal thigh. **A,** Abdominal aortic aneurysm (*), left renal vein (*arrowheads*). **B,** Aneurysm of the left common iliac artery (large *) and thrombosed common iliac vein (small *). **C,** Left hydronephrosis (*arrow*). **D,** Aneurysmatic and thrombosed left external iliac vein (*). **E,** Thrombosed left common femoral vein (*) and collateral pelvic veins (*arrowheads*). **F,** Thrombosed left superficial femoral vein (*) and a significant edema.

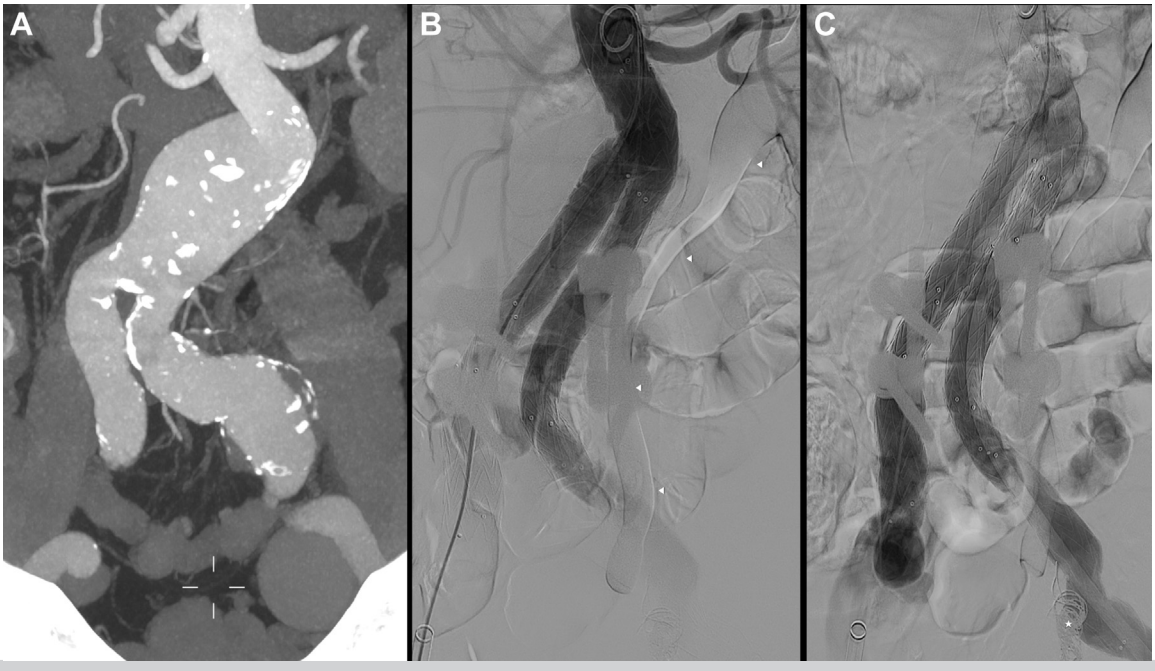

Fig 2. The aortoiliac aneurysm before and after treatment. **A,** Sagittal three-dimensional maximum intensity projection reconstruction of the aorta and common iliac arteries. **B,** Aortic angiogram after endovascular aneurysm repair (EVAR), upper part. Dilated left ureter (*arrowheads*). **C,** Aortic angiogram after EVAR, lower part. Coil embolized left internal iliac artery (*).

4 days until renal function returned. He remained intubated for 9 days owing to cardiac arrhythmias, overhydration, pain, and the need for multiple revisions of muscular necrosis in the lateral thigh compartment.

The calf and medial thigh fasciotomies were closed within the first week. The lateral thigh compartment was eventually covered by a split thickness skin graft and he was discharged to home 50 days after admission.

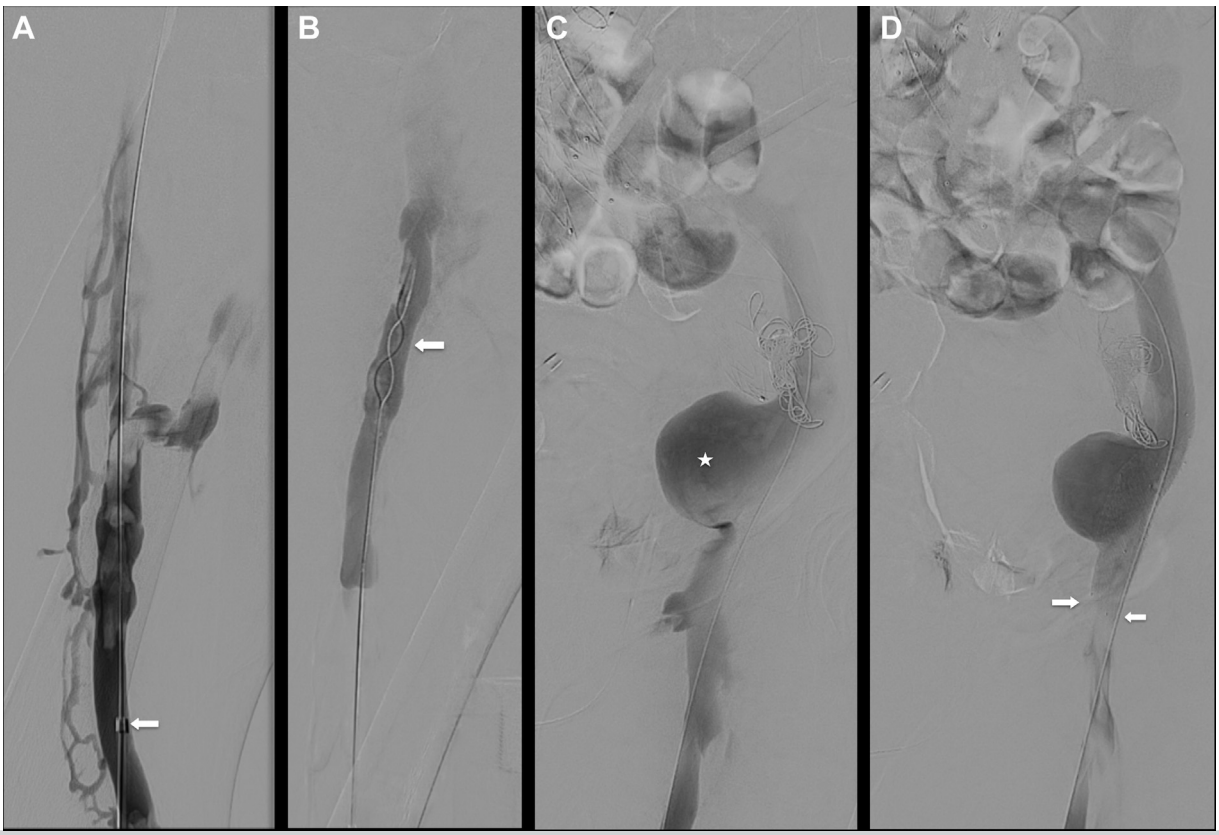

Fig 3. Venograms from the pharmacomechanical thrombectomy and stenting of the external and common iliac veins. **A,** Superficial femoral vein with thrombus. Distal marker of the 8F introducer sheath from the popliteal vein (*arrow*). **B,** The CLEANER XT thrombectomy system clearing the thrombus in the thigh. The sinusoidal shaped wire macerating the thrombus (*arrow*). **C,** The aneurysmatic left external iliac vein (*) before stenting and **(D)** after stenting (*arrows* marking the distal end of the stents).

A follow-up CT scan 2 months after the initial procedure revealed an increase in the size of the aortic component to 61 mm and a proximal type I endoleak and a large type II endoleak from a persistent inferior mesenteric artery. The type I endoleak was treated by a balloon-expandable stent graft and the inferior mesenteric artery was occluded with coil embolization. On a subsequent CT scan and contrast-enhanced ultrasound examination 14 months after the initial treatment the size of the aneurysm was stable (59 mm). The treated endoleaks were resolved, there was a persistent, small type II endoleak from a patent lumbar artery (Fig 4). Anticoagulation therapy has been discontinued and at 17 months after surgery he has no symptoms of post-thrombotic syndrome; the leg has regained full motor and sensory function.

DISCUSSION

No consensus exists on how to treat PCD, but it may be treated in several different ways: anticoagulation alone, CDT, aspiration thrombectomy, surgical thrombectomy, or fasciotomy. In the case of a threatened limb, anticoagulation alone is not considered to be a good option, although in a patient with disseminated and terminal cancer, it may well be the only option.

Several factors had to be taken into account when considering the treatment options. It has previously been reported that extrinsic pressure on the iliac veins owing to an asymptomatic or ruptured AAA may cause PCD[3,4] and in such cases the compressed vein has to be addressed as well. If the aneurysm is not ruptured or symptomatic in the sense of impending rupture, the deliberation is whether to leave the aneurysm and only treat the PCD and the compressed vein. It seems reasonable to treat the causative factor, for example, the aneurysm, in the same setting as persisting pressurization of the aneurysm may cause the venous stents to collapse. In addition, in the present case the aneurysm was of such a size that treatment was warranted.

CDT with or without mechanical adjuncts may be considered the treatment of choice today for PCD with excellent reported outcome in smaller series.[2,5] Contraindications to CDT do exist, especially recent intracerebral bleeding and/or tumor and the need for subsequent or synchronous surgery. CDT was considered in this case, but fasciotomy was indicated and the risk of substantial bleeding was weighed to be too high. Before CDT, surgical venous thrombectomy was the only treatment

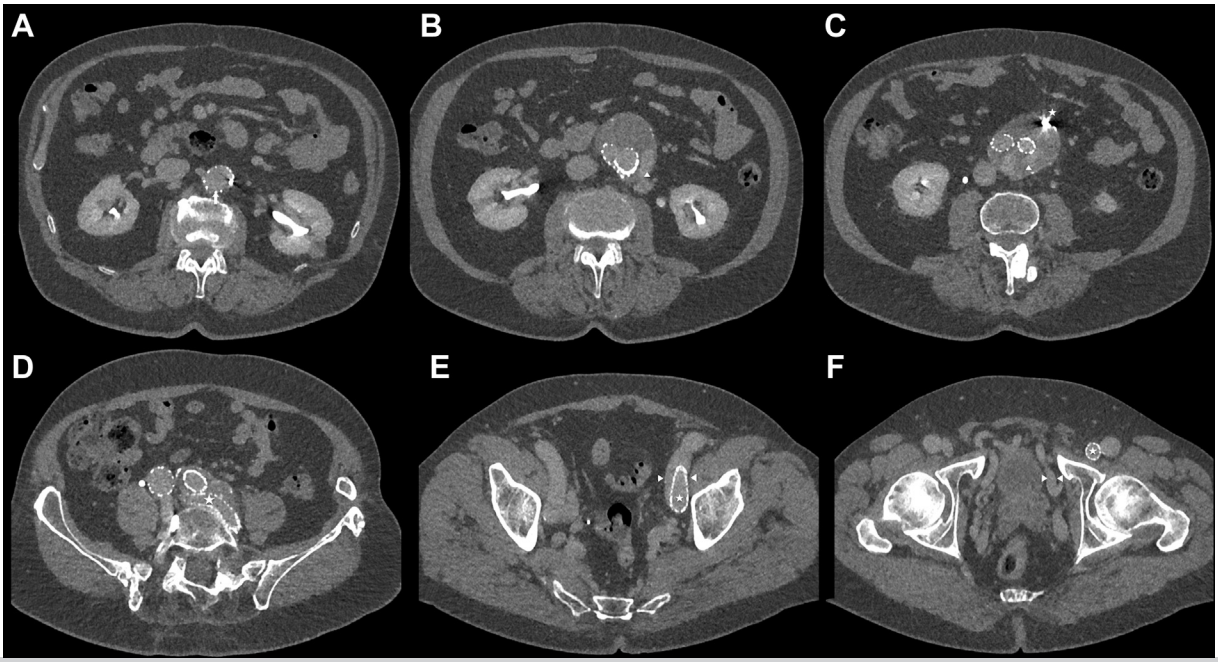

Fig 4. Follow-up axial contrast-enhanced computed tomography (CT) scans in the venous phase 14 months after surgery. **A,** Main body of the stent graft at the level of the right renal artery (*arrow*). **B,** Abdominal aortic aneurysm with the two stent graft legs, contrast enhancement in the sack of the aneurysm (*arrowhead*) indicating a type II endoleak. **C,** Abdominal aortic aneurysm with the two stent graft legs, contrast enhancement in the sack of the aneurysm (arrowhead) and coiled inferior mesenteric artery (*star*). **D,** Right and left common iliac arteries with stent grafts. Open left common iliac vein (*) with stents. **E,** Open left externa iliac vein and stents (*) at the level of the venous aneurysm. Arrowheads indicating the present diameter of the aneurysm. **F,** Open left common femoral vein with a stent (*). Persistent dilated pelvic veins (*arrowheads*).

option that could clear out the thrombus. It is still in use,[6] although several authors report less favorable outcome than after CDT; it is considered to be a difficult procedure with less clearance of thrombus, a higher rate of occurrence, and a higher rate of post-thrombotic syndrome owing to valvular incompetence.[2,5]

Pharmacomechanical thrombectomy using the CLEANER XT in combination with thrombolytic medication may be a safe alternative. The indication for this treatment is iliofemoral and femoropopliteal deep vein thrombosis and published data indicate a complete lysis in 73% to 82% of patients.[7,8] Successful treatment of PCD with the CLEANER XT has previously been reported in one case.[9] The plasma half-life of tPA is less than 5 minutes; thus, subsequent surgery may be performed without an increased risk of bleeding.

The venous aneurysm was covered by a dual layer of Zilver Vena stents with the intention to prevent possible embolization to the pulmonary circulation. The stents were sized according to the diameter of the vessel diameter proximal and distal to the aneurysm. Few reports on aneurysmal iliac veins have been published, to our knowledge none on endovascular treatment.[10] The diameter of the venous aneurysm decreased from 39 to 24 mm during follow-up and may be related to decreased venous pressure after reopening the veins and/or an effect from the dual stent layers similar to the flow diverter stents used to treat arterial aneurysms.[11]

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