Chronic occlusion mimicking agenesis of the inferior vena cava in patients with iliofemoral deep vein thrombosis

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ABSTRACT

Agenesis of the inferior vena cava (IVC) has been described as a risk factor for proximal deep vein thrombosis (DVT). We have described the cases of two patients with iliofemoral DVT who had developed post-thrombotic syndrome (PTS). Both cases were misdiagnosed as IVC agenesis on routine imaging; however, an IVC lumen was successfully identified and recanalized during venography with significant improvement in the patients' PTS symptoms. Patients with iliocaval segment DVT with a misdiagnosis of IVC agenesis might have chronic occlusion imitating agenesis. It is worthwhile to attempt percutaneous recanalization of the IVC lumen in these patients and restoring normal venous flow to prevent the development of PTS. (J Vasc Surg Cases Innov Tech 2021;7:746-9.)

Keywords: Deep vein thrombosis; IVC agenesis; Post-thrombotic syndrome

Deep vein thrombosis (DVT) occurs with an incidence of ~1 per 1000 annually in adult populations.[1] Post-thrombotic syndrome (PTS) is a long-term complication of DVT and can occur in 20% to 50% of patients after the first episode of DVT.[2,3] Inferior vena cava (IVC) agenesis or atresia is well described as a risk factor for DVT occurrence, especially in younger populations.[4-8]

In the present report, we have described two cases of iliofemoral DVT in patients with a misdiagnosis of IVC agenesis on routine imaging. Both patients had developed PTS despite treatment with anticoagulation medication and compression. They were revaluated with percutaneous venography, and an IVC lumen was identified and successfully recanalized, resulting in improvement of their PTS symptoms. Both patients provided written informed consent for the report of their case details and imaging studies.

CASE REPORT

Patient 1. A 25-year-old man was admitted to the hospital with an extensive left iliofemoral DVT. He had a history of a traumatic pretibial wound washout and splint immobilization of the same leg 2 weeks earlier. His medical history included fibromyalgia and chronic pain. He did not have a personal or family history of thrombophilia and was not taking any regular medications.

He had presented with acute pain and swelling in his left groin, with associated dyspnea and chest tightness. His vital signs were

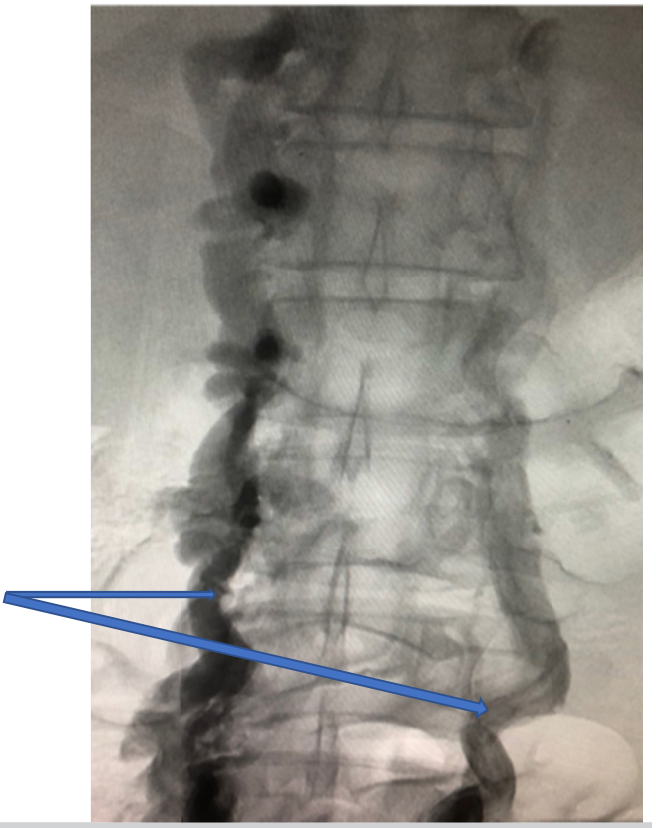

Fig 1. Diagnostic venogram of patient 1 showing prominent paravertebral channels providing venous drainage (*arrows*).

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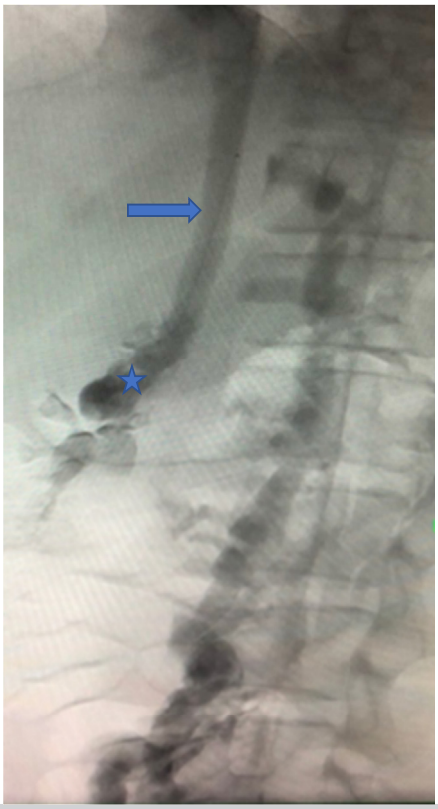

Fig 2. Left anterior oblique venogram of patient 1 with access via the right internal jugular vein and bilateral mid-thigh femoral veins. No infrarenal inferior vena cava (IVC) was visualized, with a normal suprarenal IVC (*arrow*) and only the right renal vein (*star*) visualized.

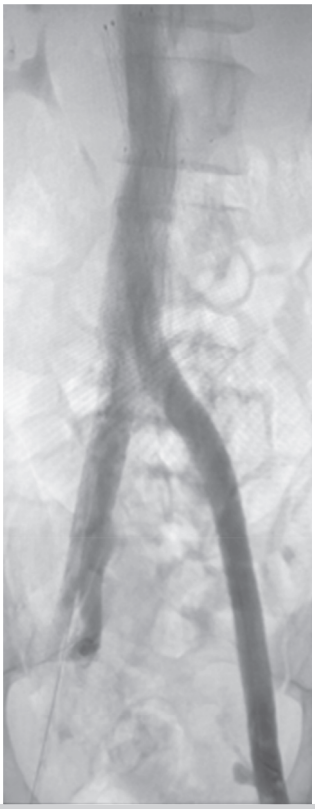

Fig 3. Imaging study of patient 1 showing IVC and bilateral common iliac veins stented with good flow achieved.

within normal parameters, and he was afebrile. His biochemistry test results revealed normal renal function, a full blood count, and normal levels of inflammatory markers. A duplex venous ultrasound revealed fresh thrombus in the left common femoral vein with proximal extension into the common iliac vein. A computed tomography (CT) pulmonary angiogram was performed because of his symptoms of dyspnea and chest tightness. However, no pulmonary embolus was identified. Oral rivaroxaban was started for treatment of the iliofemoral DVT, in addition to compression therapy.

A thrombophilia screening test was positive for heterozygous factor V Leiden mutation. The patient underwent interval ultrasound scans of his leg for surveillance of the thrombus, with a 10-month follow-up scan showing further propagation of the clot. His anticoagulation regimen, which was managed by a hematologist, was modified to apixaban, with the addition of aspirin. Repeat ultrasound during the modified anticoagulation regimen revealed continued clot propagation in the popliteal vein, and the patient was switched, yet again, to warfarin. CT venography was performed at this stage, and an absent infrahepatic IVC was reported. Congenital IVC agenesis was presumed to be the underlying etiology. At 1 year after his initial DVT, the patient developed severe PTS (CIVIQ [chronic venous disease quality of life questionnaire] score, 20-75; Villalta scale score, 16; venous clinical severity score [VCSS], 10).

The patient sought a second opinion regarding the diagnosis of IVC agenesis 19 months after his initial DVT and subsequently underwent diagnostic venography. The procedure was performed with the patient under general anesthesia, with percutaneous access achieved through the bilateral mid-thigh femoral vein and right jugular vein.

The diagnostic venogram revealed occlusion of the IVC from the infrahepatic area to the junction of the iliac veins with prominent paravertebral collateral veins (Fig 1), both common iliac veins were occluded at the origin, with stenosis of the left external iliac and femoral veins. Both iliac veins and the IVC were recanalized, predilated with a balloon, and stented (Sinus Venous and Sinus XL; Pyramed, Belrose, New South Wales, Australia; and Zilver Vena; Cook Medical Australia, Eight Mile Plains, Queensland, Australia) ranging from 14 to 25 mm based on the size of the vessel, with good flow obtained through the reconstructed veins. The patient continued warfarin therapy postoperatively. The 6-month follow-up review showed significant improvement in his PTS (CIVIQ score, 20-74; Villalta score, 8; VCSS, 5; Figs 2 and 3).

Patient 2. A 35-year-old man had been referred because of PTS affecting his right lower limb with evidence of healed ul-

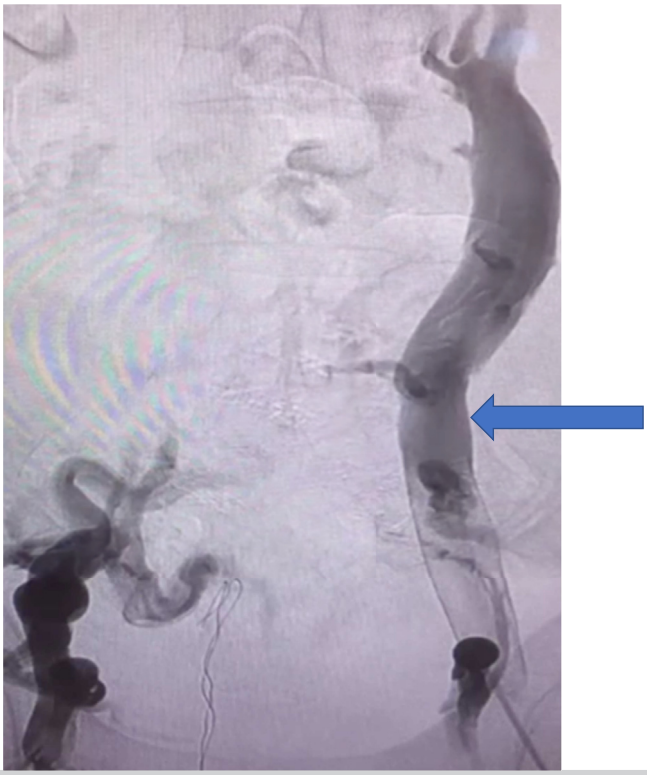

Fig 4. Intraoperative venogram of patient 2 showing complete occlusion of the inferior vena cava (IVC) and large left ascending lumbar vein as a collateral vessel (*arrow*).

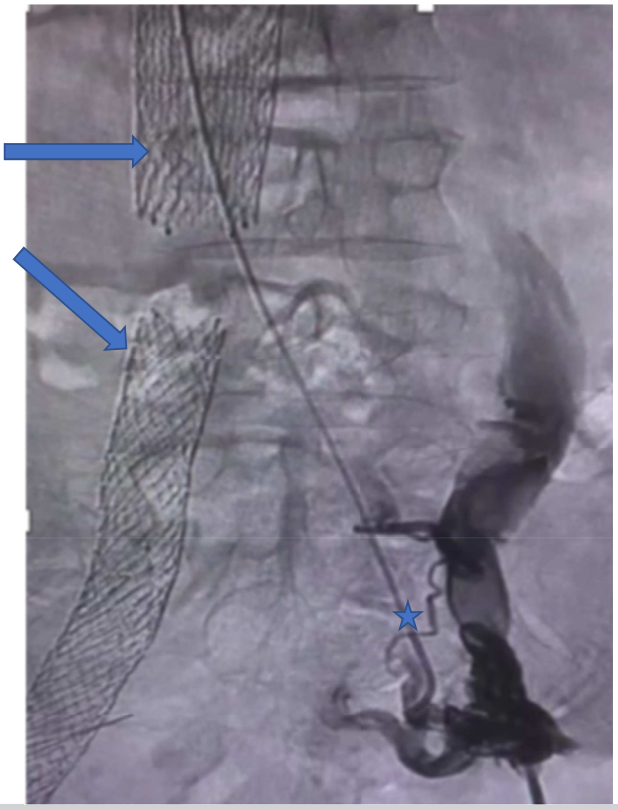

Fig 5. Intraoperative venogram showing successful recanalization in patient 2 of the inferior vena cava (IVC) from right internal jugular vein (*IJV*) and bilateral mid-thigh femoral vein access. The IVC and right common iliac vein were reconstructed with venoplasty and stenting (*arrows*). *Star* indicates a catheter from the right IJV in the left common iliac vein before venoplasty and stenting.

ceration (CEAP [clinical, etiologic, anatomic, pathophysiologic] class 5, VCSS, 7; Villalta scale score, 8). At the examination, the patient had had PTS for 10 years, after he had developed a right-sided iliofemoral DVT at 7 months after open varicocele surgery. He had no other medical history and no family history of thrombophilia. Complete hematologic and neoplastic screening performed during the initial diagnosis of DVT had not shown any evidence of malignancy or thrombophilia. His symptoms had been managed with compression stockings.

A CT venogram was performed, which revealed an obliterated IVC with extensive collateral venous channels draining into the portal vein. Chronic thrombus was present in the right common femoral vein with proximal extension into the common iliac vein. The patient underwent diagnostic venography. A patent IVC lumen was identified and successfully reconstructed using Sinus XL and Sinus venous stents (Pyramed). He was instructed to take rivaroxaban and aspirin lifelong and to wear compression stockings. At the 6-month follow-up review, the patient was asymptomatic, with no swelling or pain in the affected leg (Villalta score, 1; VCSS, 5; Figs 4-6).

DISCUSSION

PTS is a relatively common complication of DVT and develops owing to a combination of reflux and obstruction causing venous hypertension. The incidence is ~50% in the first year despite anticoagulation therapy, and severe PTS will occur in 5% to 10% patients.[9] PTS has an annual direct cost of $200 million reported in the United States, in addition to the loss of work productivity. Patients experience pain, edema, and ulceration and have difficulty mobilizing, affecting their quality of life.[10]

Intrauterine or perinatal IVC thrombosis has been proposed as a theory for the etiology of IVC agenesis and atresia.[11] Ramanathan et al[12] further supported this after describing a young adolescent with an iliofemoral DVT and associated agenesis of the infrarenal IVC, with neonatal records showing IVC thrombosis in the same patient, suggesting chronic occlusion rather than embryologic agenesis. McDonald et al[13] reported a series of 10 patients with renal vein thrombosis, 3 of whom had had associated IVC thrombosis, whose infrarenal IVC was not visualized on subsequent surveillance scans.

Treatment with prolonged anticoagulation medication in conjunction with compression therapy has been well described. However, the incidence of PTS in these patients has not been well documented.[14] A small number of case series have described the successful use of catheter-directed thrombolysis for the treatment of

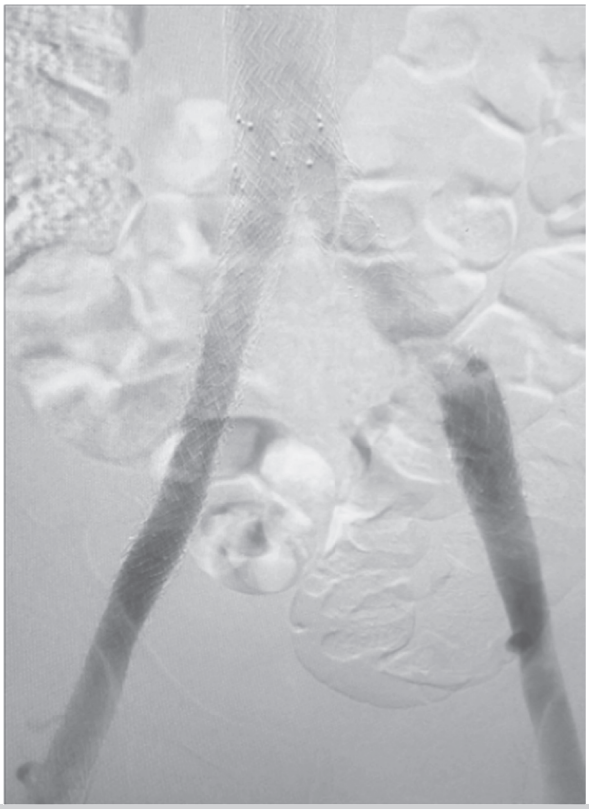

Fig 6. Completion venogram of patient 2 after stent deployment, showing good flow in the iliac veins and inferior vena cava (IVC). No flow was seen in the collateral venous channels.

venous thrombosis in association with apparent IVC atresia, with no recurrence of DVT or manifestation of PTS.[15,16] Two cases have been reported of acute iliofemoral DVT secondary to IVC atresia, with the IVC successfully recanalized, improving the patients' symptoms and highlighting the importance of restoring anatomic venous outflow.[17,18]

This paucity of evidence might have resulted from patients receiving a misdiagnosis of IVC agenesis from routine imaging (CT venography or magnetic resonance venography), in whom the IVC might be chronically thrombosed rather than truly absent. Lambert et al[14] highlighted the importance of differentiating between old thrombosis and true agenesis by seeking the caval lumen. They described magnetic resonance venography or CT venography as a suitable imaging modality to confirm a missing caval lumen.[14] However, our two cases were both misdiagnosed as agenesis on CT venography, with an identifiable lumen found during diagnostic venography.

IVC agenesis is a rare cause of iliofemoral DVT in the young population, with these patients at great risk of developing PTS. However, some of these patients could have a chronically thrombosed IVC that can be misdiagnosed as agenesis on routine imaging modalities. We believe it is worthwhile to perform diagnostic venography in these patients to seek out and recanalize the true caval lumen if present and restore normal venous outflow.

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