CHEST

A 27-Year-Old Man With Acute Severe Low Back Pain and Bilateral Leg Swelling That Prompted Renting a Wheelchair for Mobility

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A 27-year-old man with OSA, posttraumatic stress disorder, and chronic mechanical back pain presented with a 3-day history of acute atraumatic worsening of his low back pain as well as right groin numbness that was exacerbated by walking. He also complained of bilateral leg "heaviness," pain, and swelling, all becoming so severe that he rented a wheelchair for mobility.

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Physical Examination Findings

His temperature was 97°F, his pulse was 108 beats per minute, his blood pressure was 139/88 mm Hg, his respiratory rate was 16 breaths per minute, and oxygen saturation as measured by pulse oximetry (Spo_2) was 94% on room air. He was in no respiratory distress. The examination was notable for 3 to 4+ pitting edema of both legs with tenderness to palpation over the medial aspect of the left thigh.

Diagnostic Studies

Initial laboratory studies revealed a white blood cell count of 13.3 × 10^9/L, hemoglobin value of 13.7 g/dL, platelet count of 124 × 10^9/L, erythrocyte sedimentation rate of 29 mm/h, and C-reactive protein level of 122 mg/L.

MRI of the lumbar spine without contrast showed no bony abnormalities or masses but revealed a thrombosed infrarenal inferior vena cava (IVC). CT of the abdomen and pelvis with contrast revealed dilatation and nonopacification of the distal IVC (Fig 1, asterisk) and bilateral iliac and femoral veins consistent with extensive DVT. The suprarenal IVC (Fig 1, small arrow) appeared narrowed. CT angiography of the chest demonstrated two pulmonary emboli in the right lower lobe.

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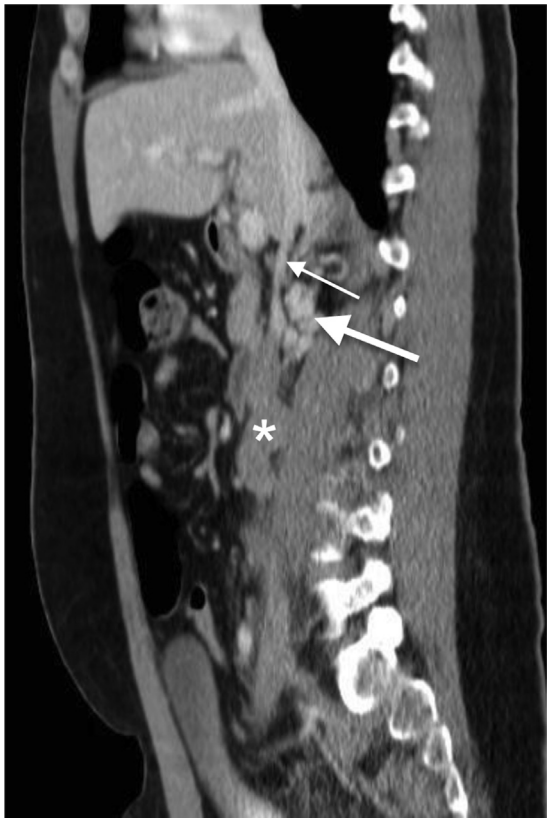

Figure 1 – *Abdominal CT scan. Sagittal CT image showing the narrowed suprarenal inferior vena cava (IVC) (small arrow), dilated nonopacified distal IVC (asterisk), and dilated venous collaterals (large arrow).*

What is the diagnosis?

Diagnosis: IVC atresia complicated by extensive DVT of the lower extremities and the numerous enlarged paraspinal venous collaterals

Discussion

IVC atresia is an uncommon congenital abnormality due to embryonal malformation of the IVC and venous system, with an estimated prevalence of 0.5% to 0.6% in the general population.

The embryogenesis of the IVC is complex and involves the sequential anastomosis of three pairs of embryonic veins—the posterior cardinal veins, subcardinal veins, and supracardinal veins—between the fourth and eighth weeks of gestation. Because of the complexity of this process, various anomalies of the IVC may occur, including IVC atresia (also known as IVC agenesis or interruption), isolated left IVC, double IVC, and retroaortic left renal vein.

Congenital dysgenesis is thought to be the central cause of IVC atresia. Complete IVC agenesis likely indicates defective embryogenesis involving the aforementioned three pairs of embryonic veins. However, intrauterine or perinatal thrombosis of the IVC can also result in IVC malformations. As a result of IVC blockage, lumbar venous collaterals develop to drain venous blood from the lower extremities. The origin of these diverting collateral veins can arise from the iliac and hypogastric veins of the pelvis or bridging collateral veins from the IVC; proximally, these collaterals may drain into the azygos venous system.

IVC anomalies may present with vague and nonspecific symptoms, such as abdominal or low back pain, or may be asymptomatic and found incidentally during radiologic examinations. Spontaneous and often extensive DVT of the lower extremities and pelvis is a known complication of IVC atresia and is hypothesized to be due to a frank or relative low-flow state that occurs when the demands for venous drainage exceed the capacity of the venous collaterals and the azygos system. Such increased demands for venous drainage may occur during growth spurts and periods of increased physical activity which may, in part, account for why IVC-associated DVT typically occurs in adolescents and young adults between the second and fourth decades of life. Indeed, it is estimated that approximately 5% of individuals $<$ 30 years who present with spontaneous DVT have IVC atresia. CT and MRI with contrast are considered the best tests to diagnose IVC atresia.

There are no definitive evidence-based treatments for IVC atresia. Placement of an IVC filter for IVC-atresia-associated DVT is difficult, as the position of the filter must be below the origin of collateral veins to effectively prevent clot migration. A filter may be considered in the low infrarenal IVC, but if this position is not technically feasible, bilateral iliac vein filters may be warranted. In the setting of bilateral ileofemoral DVT, lumbar collateral thromboses, and IVC atresia, an IVC filter cannot be effectively placed to prevent clot migration. Placing several filters in the multiple collaterals would be unwieldy and, moreover, an IVC filter may further compromise blood return by exacerbating the low-flow state.

Local continuous catheter-directed infusion of a thrombolytic agent has been recommended in patients with dolens cerulens or extensive DVT, although this approach has not been systematically studied. One regimen is tissue plasminogen activator (tPA) infusion at 0.5 to 1 mg/h (or 0.01 mg/kg/h) with a maximum of 20 to 40 mg total cumulative dose over a 24-hour period, as well as infusion of "subtherapeutic" unfractionated heparin (typically 200 to 500 units/h). tPA infusion is generally limited to $<$ 96 hours and is guided by resolution of thrombi, as determined by daily venography. Obtaining daily fibrinogen levels and holding tPA when the fibrinogen level is $<$ 100 mg/dL is recommended. Duration of anticoagulation for IVC-atresia-associated DVT is not standardized. Some have recommended at least 6 months of treatment, but because the low-flow state caused by IVC atresia is permanent, lifelong anticoagulation is appropriate but should be individualized. Surgical grafts to replace the atretic IVC have been reported to be beneficial, although the venous collaterals are generally sufficient for venous drainage once the superimposed thromboses are treated.

Clinical Course

The abdominal and pelvic CT also revealed large innumerable paraspinal venous collaterals at the level of L2 (Fig 1, large arrow) with bilateral intraluminal filling defects, consistent with thromboses of the paraspinal venous collaterals. The left renal vein drained into one of the paraspinal veins, and the right renal vein emptied into the intrahepatic IVC. The extremely small infrahepatic IVC was poorly visualized. MRI with contrast corroborated the CT findings, revealing the

right ascending lumbar collaterals as multiple varices in the right retroperitoneum adjacent to the right kidney. The azygos and hemiazygos veins were both markedly engorged as collateral venous extensions of the ascending collateral lumbar veins. These findings are consistent with congenital atresia of the infrahepatic IVC.

The patient had no family history of venous thromboembolism. Anticardiolipin antibody and lupus anticoagulant tests were both negative, and the homocysteine level was normal. Examination and multiple imaging studies did not reveal evidence of a malignancy. Given the extensive clots in the collateral veins, it is conceivable that he had repeated formation of smaller clots resulting in a vicious cycle of progressive low-flow state and clot extension until the clot burden in the collaterals reached a "tipping point," leading to the acute back pain and bilateral leg swelling.

Bilateral popliteal venous catheters were placed, with the catheter tips terminating in the ascending lumbar collateral veins. tPA at 0.5 mg/h and heparin at 200 units/h were infused through each of the two catheters. On the second day after tPA was initiated, mechanical thrombolysis of the dominant thrombi in the iliac and superficial femoral veins was performed using the CLEANER XT device (Argon Medical Devices) and balloon venoplasty. By the third day of thrombolysis, his back and leg pain as well as some of the lower extremity swelling had resolved. Venograms obtained over the 3 days of thrombolysis revealed a progressive decrease in clot burden (Fig 2A-F). tPA was discontinued on the third day of lysis and IV heparin was transitioned to long-term anticoagulation treatment.

The patient was essentially symptom free at 2 months of follow-up while continuing treatment with enoxaparin. A venogram performed at that time demonstrated that the atretic infrahepatic IVC and the vessels inferior to the IVC continued to remain patent.

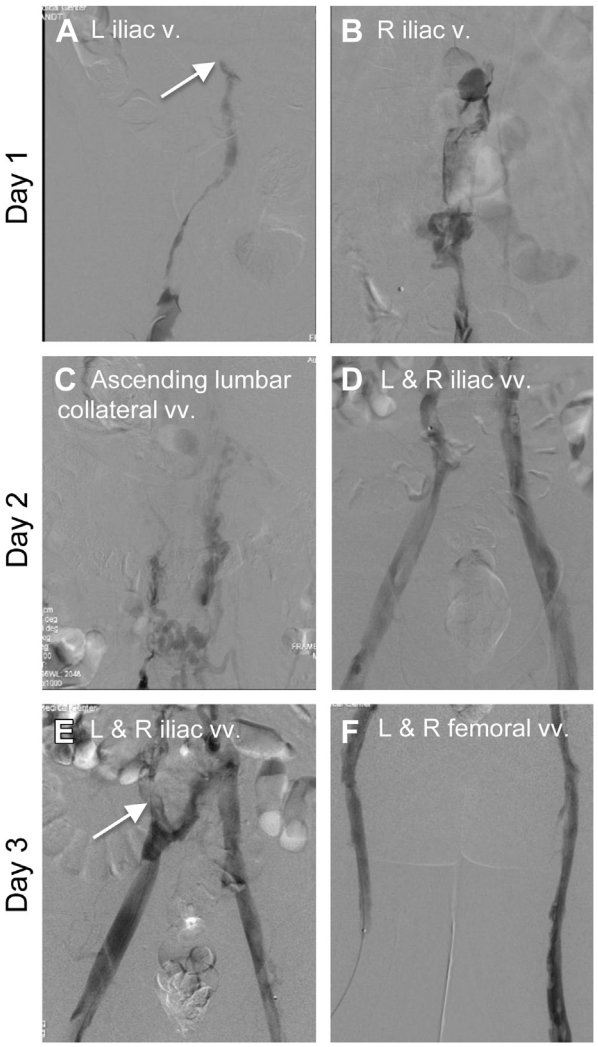

Figure 2 – *Venograms of the lower extremity veins showing progressive resolution of the DVT with catheter-directed thrombolytic infusion (all venograms are posterior views taken with the patient in the prone position). A, Complete thrombosis of the left iliac vein with no significant venous outflow after 21 hours of catheter-directed thrombolysis (arrow). B, Complete thrombosis of the right iliac vein with no significant venous outflow after 21 hours of catheter-directed thrombolysis. C, The ascending lumbar collaterals are the only patent pelvic outflow veins, as the common iliac venous confluence and IVC remain thrombosed after 52 hours of catheter-directed thrombolysis. D, Bilateral iliac veins showing marked reduction in clot burden after mechanical thrombolysis was performed in addition to the ongoing infusion of tissue plasminogen activator (tPA) at about 52 hours. E, Bilateral iliac veins showing marked reduction in clot burden at 74 hours of catheter-directed thrombolysis. Note the large left ascending lumbar collateral (arrow) arising before confluence of the iliac veins. F, Bilateral superficial femoral veins now nearly free of thrombus after 74 hours of ongoing catheter-directed thrombolysis.*

Clinical Pearls

1. *IVC atresia is due to embryonal malformation of the IVC or is secondary to thrombosis of a portion of the IVC during the intrauterine or perinatal period.*
2. *DVT is a complication of IVC atresia, typically presenting by age 40 years; therefore, IVC atresia should be considered as a potential cause of spontaneous and extensive bilateral lower-extremity DVT, particularly in a young otherwise healthy person.*
3. *Although IVC atresia acts like a natural IVC filter, the presence of large paraspinous venous collaterals can serve as conduits for pulmonary emboli. As a result, placement of an IVC filter may not prevent pulmonary emboli and could unintentionally exacerbate the low-flow state.*

4. *Catheter-directed thrombolysis is the treatment of choice for limb-threatening or extensive bilateral DVT (or both) and can decrease the risk of postphlebitic syndrome developing. Although at least 6 months of anticoagulation is recommended for IVC-atresia-associated DVT, an argument can be made for lifelong anticoagulation treatment given the ongoing risk of thrombosis.*

5. *IVC atresia is not usually surgically corrected, since venous collaterals bypass the IVC obstruction; however, in cases recalcitrant to medical treatment, surgical grafts may provide symptomatic relief.*

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Suggested Readings

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