Acta Clin Croat 2022; 61:145-148
doi: 10.20471/acc.2022.61.01.18

Case Report

LEFT-SIDED INFERIOR VENA CAVA ASSOCIATED WITH PROXIMAL DEEP VENOUS THROMBOSIS OF THE LEFT LOWER EXTREMITY

Duško Kardum[1], Tina Bečić[2], Mislav Vrsalović[3], Bruno Lukšić[4], Vedran Carević[5] and Damir Fabijanić[5]

[1]Department of Internal Medicine, Dubrava University Hospital, Zagreb, Croatia; Faculty of Medicine, Josip Juraj Strossmayer University of Osijek, Osijek, Croatia;
[2]Department of Emergency Medicine of Split-Dalmatia County, Split, Croatia;
[3]Department of Cardiology, Sestre milosrdnice University Hospital Center, University of Zagreb School of Medicine, Zagreb, Croatia;
[4]Department of Surgery, Split University Hospital Center, University of Split School of Medicine, Split, Croatia;
[5]Division of Cardiology, Split University Hospital Center, University of Split School of Medicine, Split, Croatia

SUMMARY – Left-sided inferior vena cava (IVC) is a rare congenital venous anomaly that is most frequently detected incidentally during abdominal computer tomography scanning. However, as in the case presented, the first clinical manifestation of this anomaly may be deep venous thrombosis (DVT) of lower extremities. Therefore, left-sided IVC should be kept in mind in case of inferior DVT, especially in young patients with no predisposing thrombotic risk factors.

Key words: *Deep venous thrombosis; Inferior vena cava; Anomalies*

Introduction

Anomalies of inferior vena cava (IVC), i.e., absent infrarenal IVC, left-sided IVC and double IVC, have been most frequently revealed as incidental findings during abdominal computed tomography (CT) scanning[1,2]. Because these anomalies are often asymptomatic, they represent a potential diagnostic and therapeutic pitfall particularly relevant for radiologist and surgeons. However, these rare anomalies, with an estimated prevalence of 0.07% to 8.7% in the general population, may be associated with thrombosis of the iliac and femoral veins, particularly in young patients with no predisposing risk factors[1,2].

Correspondence to: *Damir Fabijanić, MD, PhD*, Division of Cardiology, Split University Hospital Center, Spinčićeva 1, HR-21000 Split, Croatia
E-mail: damirfabijanic62@gmail.com
Received November 25, 2016, accepted November 27, 2016

We present a young male with left-sided IVC associated with deep iliofemoral vein thrombosis.

Case Report

A 27-year-old obese man (body mass index 34 kg/m^2) was admitted because of swelling of the left lower extremity. Duplex-ultrasound scan revealed thrombosis of the left iliac and femoral veins, with the permeable right venous system. Abdominal ultrasound and laboratory parameters suggestive of thrombophilia (protein C, protein S, antithrombin III, factor V Leiden, antiphospholipid antibodies) revealed no abnormalities. Because of venous collaterals revealed in the left iliac region (Fig. 1 A and B), the patient underwent abdominal CT scan, which demonstrated partially calcified occlusive thrombosis of the left-sided IVC (Fig. 2 A and B). The patient was treated with low-weight molecular heparin (enoxaparin in a dose of

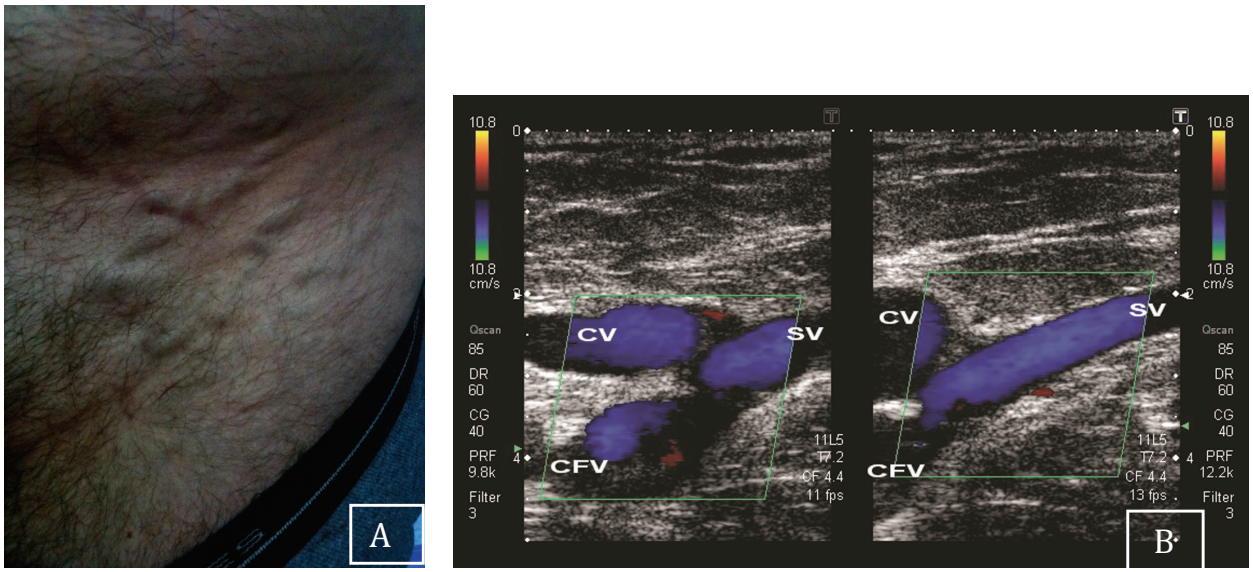

Fig. 1. Clinical examination (A) revealed visible subcutaneous veins at the left inguinal region. Duplex-ultrasound scan (B) revealed thrombosis of the left iliac and common femoral vein (CFV), with intensive vein flow through the saphenous vein (SV) and subcutaneous collaterals (CV).

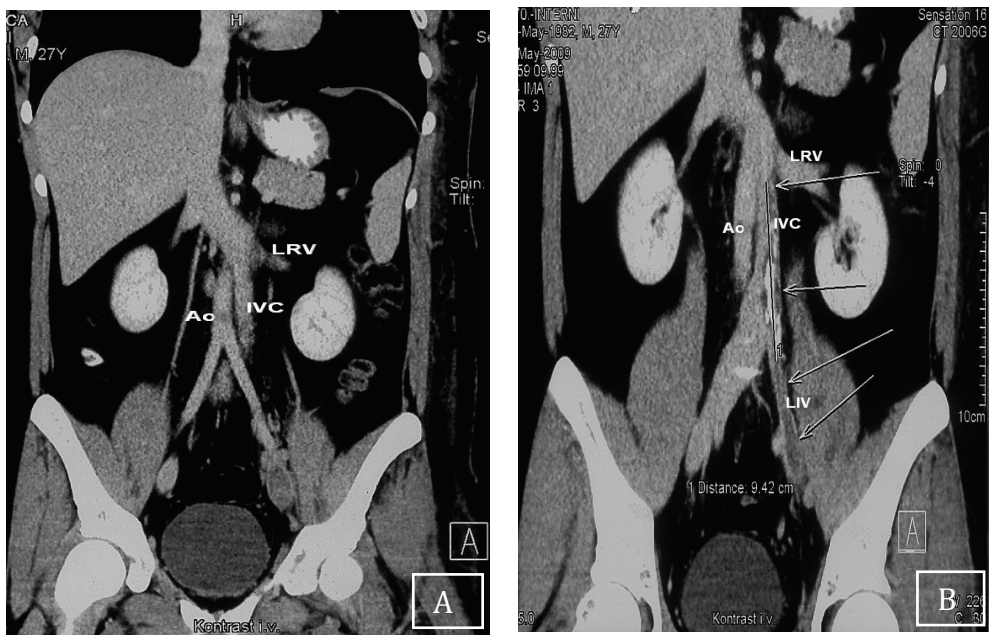

Fig. 2. Coronal reconstruction of the abdominal computed tomography scan: (A) left-sided infrarenal inferior vena cava (IVC) crosses the aorta (Ao) immediately after joining the left renal vein (LRV); (B) occlusive partially calcified thrombosis of the IVC occurred at the level of crossing aorta and expands into the left iliac vein (LIV).

100 mg bid), followed by oral anticoagulant (warfarin) with the International Normalized Ratio maintained between 3 and 3.5. After 6 months of warfarin therapy, complete resolution of thrombi in the left external iliac and femoral veins was revealed, whereas IVC thrombosis was unchanged. Two months after warfarin therapy

discontinuation, the patient experienced re-thrombosis of the left femoral vein. After thrombus resolution, lifelong thromboprophylaxis with dabigatran (2x150 mg bid) was recommended.

Discussion

The left-sided IVC is a rare congenital venous malformation with a prevalence of 0.2%-0.5%[1,2]. This anomaly develops from persistence of the left instead of the right supracardinal vein, which occurs in normal embryonic development[1]. The left-sided infrarenal IVC typically joins the left renal vein before it crosses the aorta to form a normal right-sided suprarenal IVC. Complete transposition of the IVC to the left with hemizygous continuation is extremely rare[1].

The left-sided IVC is occasionally asymptomatic and most frequently is detected incidentally during CT scanning performed before abdominal surgery; in this case, this anomaly has a significant practical implication for surgeons[1,2]. On the other hand, the main diagnostic significance of the left-sided IVC is its potential to be misdiagnosed as left para-aortic lymphadenopathy[1,2]. There have also been reports of spontaneous rupture of abdominal aortic aneurysms into the left-sided IVC and of the left-sided IVC that caused chronic intermittent mesenteric angina due to its compression of the celiac trunk while crossing in front of the aorta[1]. However, as in our case, thromboembolic complication may be the first clinical manifestation of this anomaly, especially in young people in whom up to 5% incidence of idiopathic deep venous thrombosis (DVT) may be related to IVC anatomic variation[3,4].

The potentially main pathophysiological mechanism of DVT in patients with an anomaly of the IVC is inadequate blood return in spite of prominent venous collaterals[5]. This inadequate blood return may increase blood pressure in the veins of lower extremities, with ensuing venous stasis and subsequent DVT, which is bilateral in more than 50% of patients[6]. This prevalence of bilateralism is in contrast to the reported incidence of less than 10% in patients with DVT with normal IVC[6]. Also, some authors suggest that an interaction between IVC anomalies and thrombophilia contributes to the pathogenesis of DVT in patients with these anomalies and thrombophilia[7]. However, it seems that, in our patient, thrombosis started in the IVC, at the site where IVC crosses aorta. It is possible that pulsations of the aorta, at the site of their contact, caused damage to the IVC endothelium and stimulated development of thrombosis. It is likely that in our patient, contact of the aorta and IVC was further strengthened by extreme abdominal obesity.

Dilated abdominal wall collateral veins are an important clinical sign suggesting the possibility of IVC thrombosis. Namely, in patients with the IVC anomaly associated with lower extremity DVT, a significant part of the venous outflow from the legs is taking place *via* venous collaterals. Using contrast CT, Gayer *et al*. revealed prominent collateral flow through the azygos and hemiazygos systems, ascending lumbar and internal paravertebral veins, and also anterior abdominal wall veins. Moreover, these authors report that in some patients, the markedly dilated paravertebral veins caused lower back pain[7].

So far, there are no standard guidelines for the treatment of IVC anomaly associated DVT. Patients are most frequently initially treated with low-molecular weight heparin followed by oral anticoagulation. Some authors described the efficacy and potential advantage of catheter directed thrombolysis that resulted in rapid thrombus removal, especially in patients with acute thrombosis of iliofemoral veins[8]. It seems that catheter directed thrombolysis provides immediate symptom relief and significantly decreases thrombus burden that may otherwise take weeks to resolve with systemic anticoagulation alone[8]. In patients with extensive iliofemoral DVT, pharmacomechanical catheter directed thrombolysis (a combination of mechanical thrombectomy and catheter directed thrombolysis) has also been shown to significantly decrease the thrombus burden, incidence of recurrent DVT, and incidence of post thrombotic syndrome compared to systemic anticoagulation alone[9]. According to the radiologically revealed calcification of the thrombosed IVC segment, we assumed that the IVC thrombosis in our patient was a chronic process. Therefore, instead of the IVC catheter directed thrombolysis, we decided to treat our patient with low-weight molecular heparin (enoxaparin) followed by oral anticoagulant (warfarin) in order to treat acute thrombosis of iliofemoral veins. Due to early re-thrombosis despite unproven hereditary thrombophilia, we suggested our patient to take permanent, most probably lifelong anticoagulation. In that context, because of proven efficacy and ease of administration, we prescribed a new oral anticoagulant dabigatran (direct thrombin inhibitor)[10,11]. The patient was also strongly advised to

wear elastic stocking support and leg elevation, as well as to significantly reduce his body weight.

Conclusion

The case presented suggests that rare IVC anomalies should be kept in mind in case of inferior DVT in a young patient with no predisposing thrombotic risk factors. Because of the high risk of re-thrombosis, patients with DVT associated with IVC anomalies also need life-long anticoagulation.

References

1. Malaki M, Willis AP, Jones RG. Congenital anomalies of the inferior vena cava. Clin Radiol. 2012;67:165-71. doi: 10.1016/j.crad.2011.08.006
2. Sirlak M, Cakici M, Inan MB, *et al*. Coexistence of left-sided inferior vena cava, deep vein thrombosis of the upper and lower extremities and prothrombotic polymorphisms in a young patient: a case report. Blood Coagul Fibrinolysis. 2008;19:443-5. doi: 10.1097/MBC.0b013e3282f9ae35
3. Nanda S, Bhatt SP, Turki MA. Inferior vena cava anomalies – a common cause of DVT and PE commonly not diagnosed. Am J Med Sci. 2008;335:409-10. doi: 10.1097/MAJ.0b013e318155748d
4. Lavens M, Moors B, Thomis S. Deep venous thrombosis in a young woman with hypoplastic inferior vena cava. Ann Vasc Surg. 2014;28:1036.e5-7. doi: 10.1016/j.avsg.2013.06.044.
5. Obernosterer A, Aschauer M, Schnedl W, Lipp RW. Anomalies of the inferior vena cava in patients with iliac venous thrombosis. Ann Intern Med. 2002;136:37-41. doi: 10.7326/0003-4819-136-1-200201010-00009
6. Chee YL, Culligan DJ, Watson HG. Inferior vena cava malformation as a risk factor for deep venous thrombosis in the young. Br J Haematol. 2001;114:878-80. doi: 10.1046/j.1365-2141.2001.03025.x
7. Gayer G, Luboshitz J, Hertz M, *et al*. Congenital anomalies of the inferior vena cava revealed on CT in patients with deep vein thrombosis. AJR Am J Roentgenol. 2003;180:729-32. doi:10.2214/ajr.180.3.1800729
8. Broholm R, Jorgensen M, Just S, Jensen LP, Bækgaard N. Acute iliofemoral venous thrombosis in patients with atresia of the inferior vena cava can be treated successfully with catheter directed thrombolysis. J Vasc Intervent Radiol. 2011;22;801-5. doi: 10.1016/j.jvir.2011.01.449
9. Comerota AJ, Gravett MH. Iliofemoral venous thrombosis. J Vasc Surg. 2007;46:1065-76. doi: 10.1016/j.jvs.2007.06.021
10. Balaban M, Stančić V, Rinčić G, *et al*. Recurrent venous thrombosis despite 'optimal anticoagulation therapy' for antiphospholipid syndrome – could new oral anticoagulants solve the problem? Acta Clin Croat. 2010;49:469-77. PMID: 21830460
11. Burnett AE, Mahan CE, Vazquez SR, Oertel LB, Garcia DA, Ansell J. Guidance for the practical management of the direct oral anticoagulants (DOACs) in VTE treatment. J Thromb Thrombolysis. 2016;41:206-32. doi: 10.1007/s11239-015-1310-7

Sažetak

LIJEVOSTRANA DONJA ŠUPLJA VENA POVEZANA S PROKSIMALNOM DUBOKOM VENSKOM TROMBOZOM LIJEVE NOGE

D. Kardum, T. Bečić, M. Vrsalović, B. Lukšić, V. Carević i *D. Fabijanić*

Lijevo pozicionirana donja šuplja vena je rijetka prirođena anomalija venskog sustava. Najčešće se otkriva slučajno, prilikom pretrage trbuha kompjutoriziranom tomografijom. Ipak, kao u prikazanom slučaju, prva klinička manifestacija ove anomalije može biti tromboza dubokih vena donjih udova. Stoga o lijevo pozicioniranoj donjoj šupljoj veni treba razmišljati u mlađih bolesnika s trombozom donjih udova, bez čimbenika rizika koji predisponiraju trombogenezu.

Ključne riječi: *Tromboza dubokih vena; Donja šuplja vena; Anomalije*