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

Duško Kardum¹, Tina Bečić², Mislav Vrsalović³, Bruno Lukšić⁴, Vedran Carević⁵ and Damir Fabijanić⁵

¹Department of Internal Medicine, Dubrava University Hospital, Zagreb, Croatia; Faculty of Medicine, Josip Juraj Strossmayer University of Osijek, Osijek, Croatia; ²Department of Emergency Medicine of Split-Dalmatia County, Split, Croatia; ³Department of Cardiology, Sestre milosrdnice University Hospital Center, University of Zagreb School of Medicine, Zagreb, Croatia; ⁴Department of Surgery, Split University Hospital Center, University of Split School of Medicine, Split, Croatia; ⁵Division 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 computer tomography (CT) scanning². 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 factors¹,².

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/m²) 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
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 development1. 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 rare1.

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 surgeons1,2. On the other hand, the main diagnostic significance of the left-sided IVC is its potential to be misdiagnosed as left para-aortic lymphadenopathy1,2. 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 aorta1. 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 variation1,4.

The potentially main pathophysiological mechanism of DVT in patients with an anomaly of the IVC is inadequate blood return in spite of prominent venous collaterals5. 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 patients6. This prevalence of bilateralism is in contrast to the reported incidence of less than 10% in patients with DVT with normal IVC6. Also, some authors suggest that an interaction between IVC anomalies and thrombophilia contributes to the pathogenesis of DVT in patients with these anomalies and thrombophilia7. 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 pain7.

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 veins8. 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 alone8. 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 alone9. 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.

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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. Naičešće se otkriva slučajno, prilikom pretrage trbuha kompjutoriziranjom 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 mladih bolesnika s trombozom donjih udova, bez čimbenika rizika koji predisponiraju trombogenezu.

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