Deep Venous Thrombosis in a Patient with Inferior Vena Cava Agenesis

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ABSTRACT
The prevalence of congenital inferior vena cava anomalies is difficult to pinpoint and is estimated to occur in 0.05–8.7% of the population. These abnormalities are often asymptomatic and are incidentally identified in patients undergoing imaging investigations for other reasons. Inferior vena cava agenesis maybe a risk factor for deep venous thrombosis due to inadequate venous drainage. We describe the case of an 84-year-old female who was hospitalized due to unprovoked deep venous thrombosis and inferior vena cava agenesis was found in a further investigation.

Keywords: Inferior vena cava, deep venous thrombosis, thrombolytic therapy

INTRODUCTION
Congenital inferior vena cava (IVC) anomalies are rarely seen and their prevalence is estimated to be 0.07%–8.7% (1). Most patients with these anomalies are asymptomatic. Although enlarged superficial veins around the umbilicus are observed in some cases, this can also be associated with liver cirrhosis that develops secondary to portal hypertension. Generally, these patients are incidentally diagnosed while undergoing abdominal surgery or radiological investigation for other reasons (2). Some patients with recurrent spontaneous deep vein thrombosis (DVT) also have IVC agenesis (IVCA).

In this case report, a patient hospitalized with spontaneous DVT and found to have underlying IVCA during further investigations is presented.

CASE PRESENTATION
An 84-year-old female patient was admitted to our clinic with complaints of swelling and pain in the left lower extremity lasting for 2 days. The patient had no history of previous DVT, surgical intervention, long-term bedrest, or travelling. She had no known malignancy and she did not receive hormone replacement therapy. Except a coronary angiography process, which had been performed 3 years ago, she did not have a history of vascular intervention in the femoral area.

Her physical examination revealed that she had a difference in the diameter of the left lower extremity, and the result of Homan’s test was negative. Other findings of physical examination were evaluated as normal. In the second physical examination performed after the detection of VCIA, the presence of collateral veins and appearance of caput medusa were investigated, particularly in the abdomen. However, no consistent finding was observed.

In the venous Doppler ultrasonography (DUSG) of the left lower extremity, the augmentation of the left popliteal vein was completely assessed, but acute thrombus was detected in venous structures in the proximal areas.

Based on anamnesis and ultrasonography findings of the patient, pharmacomechanical thrombectomy was planned for the patient due to the diagnosis of acute DVT in the left lower extremity. The patient was taken into the operating room. To insert the vena cava filter from the contralateral extremity, a puncture was created in the right femoral vein using the Seldinger technique, but the guiding wire could not be advanced after a certain point. The view obtained by giving opaque substance with a puncture needle under a scope revealed a serious narrowing in the proximal region of the right common iliac vein, and IVC could not be monitored. The patient was scheduled to undergo the venous phase of computed tomography (CT) angiography, but the process was not performed.

In the venous phase of CT angiography, IVC was found to be agenetic, following the involvement of the right renal vein (Figure 1), and it continued as a fibrotic band (Figure 2). Moreover, recanalized thrombus sequelae was detected in the bilateral iliac veins of the patient (Figure 3), and iliac veins were found to flow into the suprarenal segment of IVC through the paravertebral veins.

The patient was administered with lifelong oral anticoagulant therapy because of stasis in the iliac arteries. The patient was discharged with optimum INR value. DUSG examination performed for control in the third month revealed recanalized flow in the left lower common femoral vein and in the superficial femoral vein.

For 6 months, the patient had followed up by giving oral anticoagulant therapy for the targeted INR value of 2–2.5, and no recurrent DVT developed. Written informed consent was obtained from patient who participated in this study.
DISCUSSION

A normal VCI develops in the 6th and 8th weeks of embryological period, and it consists of four segments including hepatic, suprarenal, renal, and infrarenal ones (1). In infrarenal segment agenesis, iliac veins drain into the vena azygos and hemiazygos owing to collaterals in the paravertebral region and umbilicus.

IVCA causes inappropriate drainage and stasis of iliac veins and poses a risk factor for DVT (3). It is interesting that our patient was an 84-year-old woman although this condition is generally seen in male patients at the age of ≤30 years. In the series of Lambert et al., which consisted of 72 patients, 59 (82%) were males (4). In another similar series on 5 patients, it was reported that IVCA was more common among men (5). When spontaneous DVT, particularly involving iliac veins, is detected through ultrasonography, IVC must definitely be examined through CT or magnetic resonance imaging (MRI). Obernosterer et al. reported that 31 of 97 patients had occlusion even in iliac veins and IVCA was found in 5 of these 31 patients (3).

While the prevalence of DVT is 5% in patients with IVCA, it varies between 0.5% and 0.6% in those at the same age with normal IVC (6, 7). At present, while some techniques such as long-term (3–6 months) anticoagulant therapy, mechanical thrombectomy, and pharmacomechanical thrombectomy are used in the treatment of DVT, the optimum treatment method for IVCA patient with DVT has not been concretized yet. In a series of 72 cases conducted by Lambert et al. in 2010, almost all patients were given long-term anticoagulant therapy and none of the patients had recurrent DVT (4). In our case, the patient received lifelong anticoagulant therapy and recurrent DVT was not found during follow-up.

CONCLUSION

In conclusion, our patient was a geriatric female patient having DVT associated with a rare congenital cause. Our case was deemed worthy of presenting because it is a rarely seen case and it shows that unnecessary interventions and DVT recurrence could be avoided with accurate diagnosis and with lifelong anticoagulant therapy.
Informed Consent: Written informed consent was obtained from patient who participated in this study.

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REFERENCES


