» Case Report «

Congenital Absence of Inferior Vena Cava with Azygos Continuation Revealed by Vascular Echo in a Patient with Pulmonary Thromboembolism and Deep Vein Thrombosis: A Case Report

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A 44-year-old man with an isolated anomaly of azygos continuation of the inferior vena cava (IVC) presented with dyspnea due to pulmonary thromboembolism (PTE) and deep-vein thrombosis (DVT). Sonographic examination disclosed not only pulmonary hypertension and DVT, but also infrahepatic interruption of the IVC with azygos continuation. A rare anomaly of azygos continuation of IVC could cause DVT and PTE. Vascular echo could play an important role in the examination of DVT and/or venous anomalies.

Keywords: vascular echo, deep vein thrombosis, azygos continuation

INTRODUCTION

Congenital absence of the inferior vena cava (IVC) with azygos continuation has an incidence of less than 1% and is occasionally comorbid with congenital heart disease.¹⁾ These anomalies have been reported to be associated with deep-vein thrombosis (DVT) and/or pulmonary thromboembolism (PTE).²⁾ Vascular echo has been recently used to evaluate DVT.³⁾ We report a case of infrahepatic interruption of IVC with azygos continuation presenting with DVT and PTE, in which vascular echo was key for the diagnosis.

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CASE REPORT

A 44-year-old man presented with worsening shortness of breath. He was afebrile, pulse 110/min regular, and blood pressure of 99/77 mmHg. On physical examination, neck veins were slightly dilated, and the heart had a regular rhythm with an increased S2. Extremities had bilateral pedal edema. The chest radiograph on admission revealed a right mediastinal mass just above the hilum. The coagulation increased at a D-dimer level of 11.8 µg/ml and fibrin degradation products of 25.0 ug/ml. Electrocardiogram showed a normal sinus rhythm, a right electro-axis deviation, and inverted Ts in II, III, and aVF leads.

Echocardiography disclosed a dilated right ventricle involving a deformity of the left ventricle in diastole and the estimated systolic pressure of 44 mmHg in the right ventricle. Vascular echo demonstrated a mural thrombus in the left popliteal vein (**Fig. 1**). The absence of IVC with a dilated azygos vein drained from renal veins (**Fig. 2A**), and hepatic veins were seen to drain directly into the right atrium (**Fig. 2B**). Enhanced computed tomography (CT), contrast-enhanced magnetic resonance imaging (MRI) and venography showed that the hepatic segment of the IVC was absent (**Fig. 3A**), and the azygos

Fig. 1 Vascular echo discloses thrombi (arrow) in the left popliteal vein.

Fig. 2 (A) Color Doppler echo illuminates a dilated azygos vein drained from the left renal vein (Az: azygos vein; R: renal vein). (B) Hepatic veins draining directly into the right atrium (arrow).

Fig. 3 (A) Contrast-enhanced magnetic resonance imaging (MRI), anterior view, shows that the hepatic segment of the inferior vena cava (IVC) is absent (arrow). (B) Venography shows the azygos continuation from renal veins drains to the superior vena cava.

continuation from renal veins drained to the superior vena cava (**Fig. 3B**). Multiple small thrombi in the bilateral pulmonary artery were observed on the enhanced CT (**Fig. 4**). After the intravenous administration of unfractionated heparin, right ventricular systolic pressure decreased from 4 mmHg to 3 mmHg. Dyspnea improved and thrombi in the pulmonary artery and left popliteal vein disappeared consequently. He has taken warfarin and was followed for 5 years without the recurrence of DVT and PTE.

Fig. 4 Reformatted postcontrast computed tomography (CT) image, anterior view, shows a pulmonary arterial thrombus (arrow) and hepatic veins draining directly into the right atrium.

DISCUSSION

Congenital absence of the IVC with azygos continuation is relatively rare, occurring with an incidence of less than 1%.¹⁾ Although this anomaly could be found incidentally in asymptomatic individuals, several papers have reported that this malformation could cause venous insufficiency of the lower limbs with a potential thromboembolic disease.^{4,5)} The present anomaly was reported to be associated with congenital heart disease.¹⁾ Sonographic evaluation for venous disease has progressed, and vascular echo has played an important role in the diagnosis of DVT. However, there had been no report that vascular echo had a key to the diagnosis of infrahepatic interruption of the IVC with azygos continuation comorbid with DVT and PTE.

We have assessed to venous thrombus including IVC in patients with suspect DVT using color Doppler and found infrahepatic interruption of the IVC with azygos continuation. We could propose that the routine IVC

search might be a necessary step for thromboembolic sources in patients with PTE because IVC anomaly represented the unusual condition that predisposes to DVT by inducing venous stasis.

Although the enhanced CT can be a gold standard for the evaluation of DVT and venous anomaly, it is rather difficult to use the enhanced CT as a screening tool for DVT. Noninvasive vascular echo could be a first imaging modality to examine DVT. The present case report might suggest the requirement of an evaluation for DVT, including IVC and/or iliac venous thrombi, using color-Doppler vascular echo in patients with possible DVT.

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DISCLOSURE STATEMENT

There are no conflicts of interest to declare.

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