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SHORT REPORT

Catheter Directed Thrombolysis for Pulmonary Embolism in Patients With Rare Inferior Vena Cava Variants

Cheng-Hsuan Tsai, Yen-Hung Lin, Jen-Kuang Lee *

Department of Internal Medicine, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan

Introduction: Congenital left inferior vena cava (IVC) with hemiazygos continuation and drainage into the superior vena cava (SVC) is a rare IVC variant and is associated with venous thrombo-embolism. This is the report of a case of hemiazygos continuation of the left IVC receiving catheter directed thrombolysis (CDT) for pulmonary embolism.

Report: A 72 year old woman presented with progressive dyspnoea, and CT images confirmed the diagnosis of pulmonary embolism. The unusual route of the IVC was observed during the right heart catherisation and CDT. CDT was then performed with a left femoral venous approach. The patient tolerated the procedure well, and the follow up pulmonary angiogram showed no residual thrombus.

Discussion: The use of CDT for pulmonary embolism in patients with left IVC with hemiazygos continuation and drainage into the SVC has been reported rarely. Awareness of this diagnosis is critical during right heart catheterisation to prevent devastating complications. However, with careful manipulation, right heart catheterisation and CDT can be achieved successfully. Pulmonary embolism in patients with left IVC with hemiazygos continuation and drainage into the SVC can be treated with CDT safely and effectively with caution. It is wise to remember the different anatomical variations during IVC catheterisation.

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INTRODUCTION

Congenital left IVC with hemiazygos continuation and drainage into the superior vena cava (SVC) is a relatively rare inferior vena cava (IVC) variant with an incidence of less than 1%.[1] Embryogenesis of the IVC is a complicated process, and disruption of this process can result in IVC abnormalities.[1] Although left IVC and other variants can be found in asymptomatic patients, several papers have reported increased risk of venous thrombosis from venous stasis in patients with left IVC.[2–4] CDT therapy, especially ultrasound assisted CDT (UACDT) therapy for venous thrombosis, has received increasing attention in recent years. UACDT can restore right ventricular haemodynamics in cases of pulmonary embolism more rapidly than anticoagulation alone.[5,6] At the study institution, however, CDT for pulmonary embolism has primarily been performed using a femoral venous approach that may encounter obstacles when applied to patients with IVC abnormalities. Here, the case is reported of a patient with left IVC with hemiazygos continuation presenting with sub-massive pulmonary embolism who underwent successful UACDT.

CASE REPORT

A 72 year old woman with hypertension and rheumatoid arthritis presented with progressive dyspnoea for two weeks. She had no history of venous thrombosis. The electrocardiogram showed sinus rhythm with the S1Q3T3 pattern. Chest Xray showed enlargement of the pulmonary trunks and right ventricle. Echocardiogram revealed a dilated right heart and McConnell's sign. The vascular duplex showed no deep vein thrombosis. Pulmonary CT angiography revealed thrombus in the left main pulmonary artery and bilateral lobar pulmonary arteries (Fig. 1). UACDT was performed because of progressive dyspnoea even under systemic anticoagulation.

Echo guided right femoral venous cannulation was performed first under local anaesthesia, and a 6F sheath was inserted. However, it was difficult to pass a 5F multipurpose diagnostic catheter through the normal IVC route. The venogram showed a tortuous right femoral vein and abnormal venous drainage route through a left IVC with hemiazygos continuation then drainage to the SVC (Figs. 2 and 4). Because of the tortuosity of the right femoral

* Corresponding author. National Taiwan University College of Medicine and Hospital, No. 7, Chung-Shan South Road, Taipei, 100, Taiwan.
E-mail address: b85401104@gmail.com (Jen-Kuang Lee).

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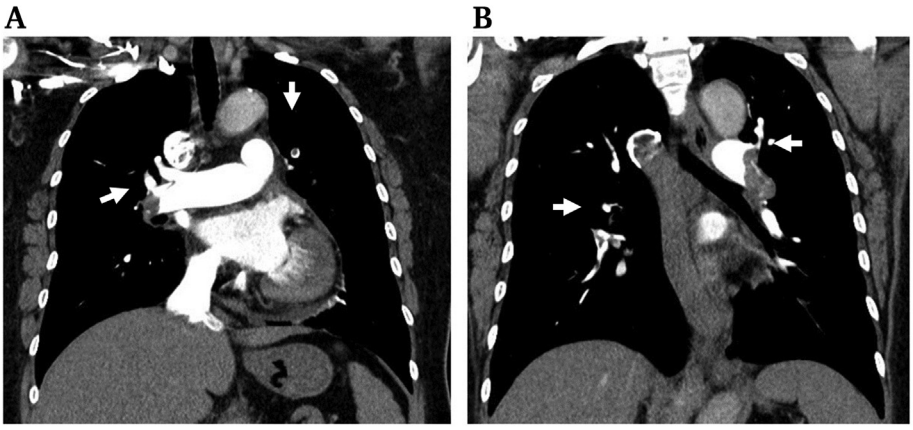

Figure 1. Computed tomography pulmonary angiography showing high burden thrombus (white arrow) in the right interlobar artery (A) and left main pulmonary artery (B).

vein, echo guided left femoral vein punctures were performed and 90 cm sheath inserted for better support. After a pigtail catheter was inserted to the main pulmonary artery with a 0.035″ 300 cm Radifocus stiff wire support, conventional pulmonary angiography was performed at designated EkoSonic endovascular UACDT catheter placement sites. Two UACDT catheters were placed bilaterally to the common basal pulmonary trunks, respectively (Fig. 3). Urokinase infusion via the UACDT catheters was then started immediately, and the dyspnoea improved dramatically within 24 hours. The follow up conventional pulmonary angiogram showed a patent pulmonary artery without residual thrombus. The catheters were then removed, and the procedure was successfully completed without complications.

DISCUSSION

The development of the IVC is a complex process, with the IVC originating from three paired embryonic veins including supracardinal veins, posterior cardinal veins, and subcardinal veins[1] (Fig. 4). A developmental error during any step in the formation of the IVC will cause different types of IVC abnormalities, including IVC agenesis, double IVC, and left IVC.[1] A left IVC results from regression of the right supracardinal vein with persistence of the left supracardinal vein. Normally, the left IVC extends from the left renal vein and then joins the right IVC.[1] However, there may be variations in this arrangement. In the present case, the right IVC was atrophied and the left IVC was connected to the hemiazygos vein and then drained to the SVC via the azygos vein (Fig. 4). Left IVC with hemiazygos continuation and drainage into the SVC is one of the rare IVC variants, having an incidence of $<1\%$.[1] Most individuals with this condition are asymptomatic, but others have non-specific abdominal or lower back discomforts, abdominal wall venous distension, venous claudication, and other symptoms associated with venous stasis.[3] The drainage of the lower extremities through the azygos and hemiazygos veins may be

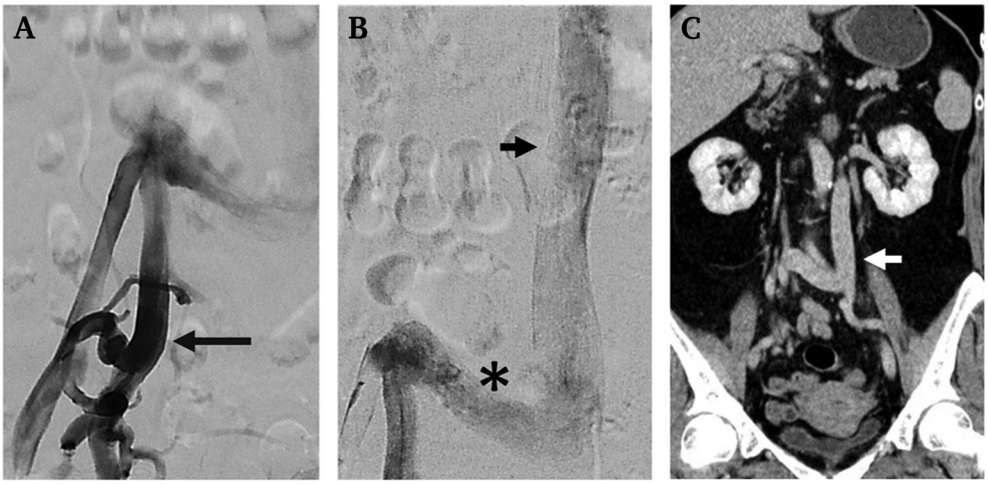

Figure 2. Venogram from the right common femoral vein showing tortuous right femoral vein (long black arrow) (A) and abnormal venous drainage route through intersupracardinal anastomosis (black asterisk) to the left inferior vena cava (IVC) (black short arrow) (B). The CT image demonstrated the venous drainage route of the left IVC (white arrow) (C).

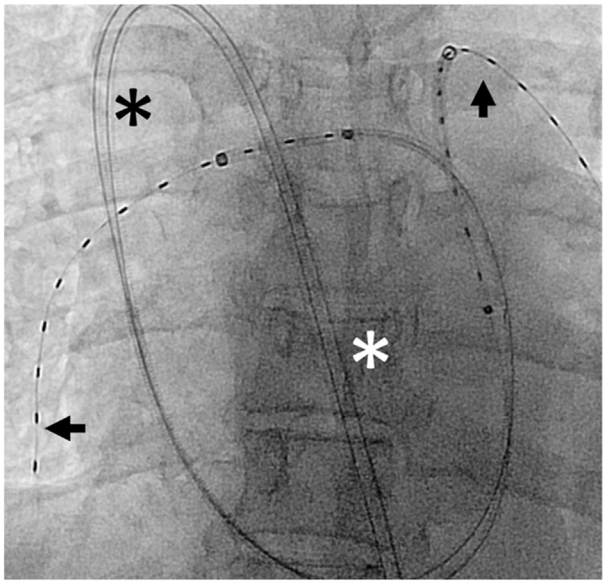

Figure 3. Fluoroscopic image showing the route of venous drainage via the thrombolysis catheter from the hemiazygos vein (white asterisk) to the superior vena cava (black asterisk) then to the right heart. There were two EkoSonic endovascular thrombolysis catheters placed in the bilateral common basal trunks of the pulmonary arteries (black arrow).

PE. The trial showed that UACDT results in decreased RV dilation, decreased pulmonary hypertension severity, and decreased anatomical thrombus burden with minimised intracranial haemorrhage.[9,10] However, the abnormal IVC variants may lead to technical CDT difficulties, especially with a femoral approach. For interventional cardiologists, being aware of the variant of left IVC with hemiazygos continuation is critical during CDT for pulmonary embolism or other right heart procedures to prevent devastating complications. However, this variant can easily be missed without a careful review of CT images. For patients with this IVC variant, the ideal vascular access sites for CDT or other right heart procedures will be the internal jugular veins. Furthermore, the left femoral vein can also be a suitable vascular access site for procedures performed with careful manipulations. The present study demonstrates that UACDT and right heart catheterisation can be achieved successfully via a left femoral venous approach without complications.

Congenital left IVC with hemiazygos continuation and drainage into the SVC is an unusual cause of pulmonary embolism. Accordingly, it is wise to remember the different anatomical variations during catheterisation of the IVC.

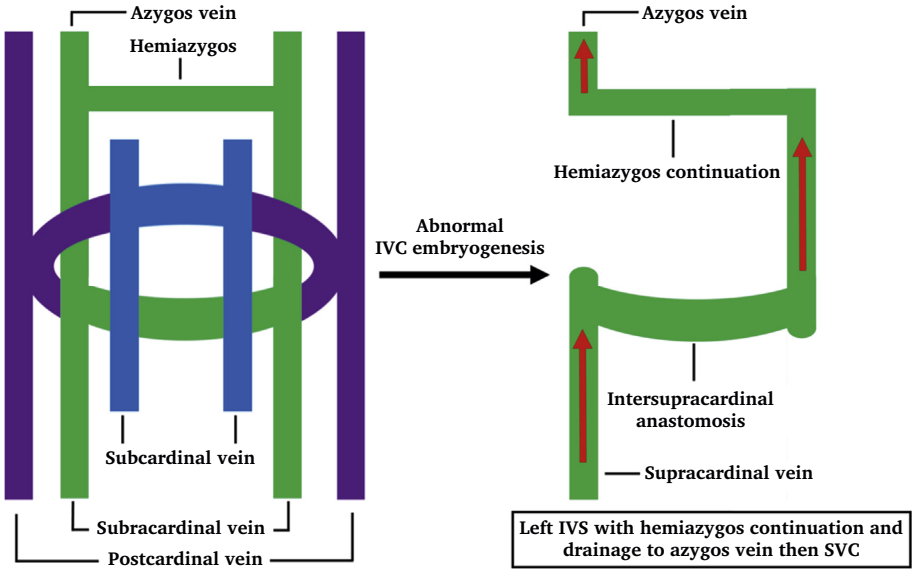

Figure 4. The inferior vena cava (IVC) originates from three paired embryonic veins. In this case, the left IVC resulted from regression of the right supracardinal vein with persistence of the left supracardinal vein. The left IVC was connected to the hemiazygos vein then drained to the superior vena cava (SVC) via the azygos vein.

insufficient, causing venous stasis and eventually thrombosis.[2,7] This case presented with sub-massive pulmonary embolism complicated by progressive dyspnoea even with adequate anticoagulation. As such, UACDT was used as a rescue therapy.

CDT and UACDT are mainly used to treat sub-massive or massive pulmonary embolism.[8] The prospective, single arm, multicentre SEATTLE II trial evaluated the efficacy and safety of UACDT for patients with sub-massive or massive

CONFLICTS OF INTEREST

None.

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