Difficult Temporary Pacing in a Patient with Interruption of Inferior Vena Cava and Hemiazygos/Accessory Hemiazygos Continuation

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ABSTRACT

Many cardiovascular procedures such as temporary pacemaker insertion, electrophysiological studies, and percutaneous transmitral commissurotomy are carried through inferior vena cava (IVC). Its malformations such as its interruption with azygous/hemiazygous continuation, left-sided IVC, and double IVC can make some difficulties for cardiologists during these procedures. In this case report, we present the case of a 75-year-old male with recurrent episodes of syncope and bradycardia. Due to interrupted IVC, temporary pacing lead was difficultly advanced from the IVC to the accessory hemiazygous vein, left brachiocephalic vein, superior vena cava, right atrium, and right ventricle. Temporary pacemaker insertion is an invasive procedure and should be performed under the guidance of fluoroscopy. However, rarely, IVC malformations such as interrupted IVC make it difficult to do procedure in a short period of time. Rupture of IVC and some complications due to severe bradycardia can be prevented by understanding rare IVC malformations.

Introduction

Inferior vena cava (IVC) is an important vein for cardiologists. When the two common iliac veins join together, IVC is formed. It is the largest vein which drains deoxygenated blood from the lower part of the body to the right atrium. Normally, it runs on the right side of the aorta and consists of three parts: hepatic, renal, and sacrocardinal (1).

In embryonic period, yolk sac blood drains into the sinus venosus through the vitelline (omphalomesenteric) veins. Gradually, the
proximal and distal parts of the left vitelline vein disappear, and the hepatic portion of the IVC is formed by the development of right vitelline vein. The right subcardinal vein and the right sacrocardinal vein develop into the renal and the sacrocardinal segments of the IVC, respectively. When renal and hepatic segments join together, IVC formation is completed. Right intercostal veins drain into the right supracardinal vein and left intercostal veins drain into the left supracardinal veins, so azygos on the right side and hemiazygos veins on the left side are formed. Azygos vein also receives blood from hemiazygos vein.

The accessory or superior hemiazygos vein created by joining the left middle intercostal veins connects to the azygos or hemiazygos vein, but rarely it ends in the left brachiocephalic vein (2).

Any deviation in the developmental process of IVC formation mentioned above may result in abnormal anatomical IVC pattern such as interrupted IVC with azygos/hemiazygos continuation, left-sided IVC, and double IVC. When the right subcardinal vein fails to anastomose with the hepatic sinusoids, the hepatic part of the IVC does not develop and it will be interrupted. In congenitally interrupted IVC, the azygos or hemiazygos veins and superior vena cava have the main role in returning blood to the right side of heart; in this situation, the role of azygos vein is prominent. Heterotaxy and situs ambiguous are other congenital abnormalities seen in patients with IVC abnormalities (1, 3, 4).

Case Report
A 75-year-old male referred to our hospital with recurrent episodes of syncope and falling for 2 days before. The electrocardiogram showed sinus bradycardia (heart rate = 30). On physical examination, no other significant abnormality was detected. He was taken to the catheterization laboratory unit for temporary pacemaker placement through transfemoral venous approach. Right femoral venous access was obtained; however, it was difficult to advance pacemaker lead from the IVC to the right atrium. Therefore, IVC venography by injection of contrast in the right femoral vein was done. Interrupted IVC and its continuation through the accessory hemiazygos vein to the left brachiocephalic vein were confirmed by angiography (Figure 1).

Then, a temporary pacing wire was advanced from the IVC to the accessory hemiazygos vein, left brachiocephalic vein, superior vena cava, right atrium, and right ventricle (RV). RV pacing was achieved successfully (Figure 2).

With the diagnosis of sinus node disease, a permanent pacemaker was placed. During
hospital admission, no new episode of syncope was detected, and the patient was discharged from the hospital with stable hemodynamics, and he is currently on follow-up.

Discussion
In this patient, failure of anastomosis between hepatic sinusoids and right subcardinal vein resulted in interrupted IVC. Hence, blood return to the heart depends on the presence of hemiazygos or azygos veins (5). This is the most common congenital anomaly of the IVC, and is usually found in patients with polysplenia. It may also have hemi-azygos/accessory hemi-azygos continuation. Previous studies have shown the incidence of IVC anomalies in 0.6-2 percent of patients with congenital heart disease, and < 0.5% in normal population. Cyanotic heart diseases and left isomerism usually accompany this situation, but patients with isolated form of IVC interruption usually are not symptomatic and patients are not aware of the disease unless they need vascular intervention, coronary artery bypass graft, or imaging study. Phlebography, computed tomography scan, magnetic resonance imaging, and echocardiography are some modalities with acceptable sensitivity to diagnose this anomaly. A dilated azygos may be misdiagnosed as aortic aneurysm or mediastinal mass, and previously mentioned modalities are helpful in these situations (6).

Interrupted IVC may have association with deep vein thrombosis, sinus node disease, high-degree atrioventricular block, and atrial flutter. Many procedures such as temporary pacemaker insertion, electrophysiological procedures, and percutaneous transmtral commissurotomy for mitral stenosis are done through IVC (5, 7). When IVC ends below the hepatic veins, systemic venous flow has to find other route to carry blood from the lower extremities to the right atrium. In this situation, dilated azygos/hemiazygos veins through the superior vena cava make the venous blood flow possible (7, 8).

In conclusion, the IVC, due to its complex embryogenesis, may develop abnormally. The isolated interrupted IVC is often clinically asymptomatic and should be considered when a cardiologist encounters difficulty in reaching the right side of the heart through femoral vein approach.

Conflict of Interests
Authors have no conflict of interests.

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References