Double superior vena cava: presentation of two cases and review of the literature

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Double superior vena cava: presentation of two cases and review of the literature

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ABSTRACT

Introduction: Various anomalies in the development of the great thoracic veins of the embryo can be incidentally discovered in the normal adult. Duplication of superior vena cava (SVC) is a rare abnormality, but the most common thoracic venous congenital anomaly.

Case reports-methods: We present two cases in the intensive care unit of our hospital, of asymptomatic patients who underwent an uneventful central line placement in the left subclavian vein. The track of the catheter, as shown in the X-ray, was misplaced to the left of the aorta and further investigation with computed tomography angiography confirmed a persistent left SVC. In both cases the vein drained into the coronary sinus and then to the left atrium. In the second case the echocardiography revealed a dilated coronary sinus.

Conclusions: Double SVC can be fortuitously discovered during catheter insertion, thoracic or cardiac imaging and surgery. In most cases it drains into the left atrium, through the coronary sinus. This entity is significant to the physician because of its importance in differential diagnosis as a cause of a widened mediastinum, as well as any difficulty that can occur in the placement of a central venous catheter or a pacemaker.

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KEYWORDS

Left superior vena cava; coronary sinus; congenital venous anomalies; widened mediastinum

Introduction

Congenital abnormalities of superior vena cava (SVC) can present as incidental findings either in childhood, be related to underlying cardiac conditions or be syndromic in origin [1]. During the eight week of the embryological period, an oblique anastomosis is formed between the two anterior cardinal veins, which becomes the left brachiocephalic vein. Then the caudal part of the left anterior cardinal vein occludes. Right anterior cardinal vein and right common cardinal vein form SVC [2].

The SVC drains blood from the head and the upper limbs, into the right atrium. Normally, the right side is drained through the right brachiocephalic vein, whereas the left side through the left brachiocephalic vein. In some instances, a double SVC may be encountered. These vessels are referred to as the right and the left SVC [3].

Persistent SVC is a rare anomaly with a prevalence of 0.3–0.5% in general population. On the contrary, this percentage varies between 10–11% in patients with congenital heart disease [4].

In most cases a persistent left vena cava drains into the right atrium, through the coronary sinus [5]. We hereby present two cases of a double superior vena cava and a short review of the existing literature.

Case 1

A 62-year-old woman was admitted to the intensive care unit (ICU) of our hospital intubated, under sedation and mechanical ventilation, because of spontaneous subarachnoid hemorrhage. The patient underwent an uneventful central vein catheterization in her left subclavian vein. The X-ray revealed an abnormal position of the catheter, which appeared to descend into the aorta (Figure 1). A chest computed tomography was performed with 3D reconstruction and revealed the catheter tracking into a left superior vena cava with the tip inside the coronary sinus, which was confirmed with intravenous contrast (Figures 2–4). Further investigation with echocardiography did not reveal any other cardiac anomalies.
Case 2

A 58-year-old woman was immediately intubated and transferred to the ICU of our hospital, after a pulmonary infection and hypoglycemic coma. A guided-wire central catheter was placed in the left subclavian vein. The X-ray showed an increased cardiac index and a misplacement of the catheter left to the aorta (Figure 5). A computed tomography angiography (CTA) revealed a left superior vena cava with an uncertain track. Because of an

Figure 1. Central vein catheter tracking left to the aorta. Widened mediastinum. The arrow indicates the presence of the catheter.

Figure 2. Computed tomography angiography (CTA). The central line catheter inside the left superior vena cava. Injection of a contrast media through the catheter reveals the tip inside the coronary sinus.

Figure 3. The CTA in Figure 2 depicts a more detailed view of the contrast media draining into the coronary sinus and the right atrium.

Figure 4. A coronal view. Absence of the left innominate vein. The central vein catheter inside the persistent left superior vena cava.

Figure 5. Widened mediastinum. Increased cardiac index (white arrows). The central vein catheter is misplaced to the left of the mediastinum (black arrow).

Figure 6. Echocardiography. Dilated coronary sinus (CS). RA: right atrium; RV: right ventricle; LV: left ventricle.
unexplained sinus tachycardia, a cardiac ultrasound was performed. The patient suffered from a serious pulmonary hypertension with a dilated coronary sinus (Figure 6). The patient’s status deteriorated and no further intervention could be accomplished.

**Discussion**

The cardinal veins include the anterior cardinal vein (draining the cephalic portion of the body) and the posterior cardinal vein (draining the remainder of the body of the embryo). The anterior and posterior cardinal veins on each side join to

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**Figure 7.** The cardinal veins on the day 28 of embryo. ACV: anterior cardinal vein; CCV: common cardinal vein; PCV: posterior cardinal vein; SV: sinus venosus.

**Figure 8.** The rise of SVC. ACV: anterior cardinal vein; Sub.V: subclavian vein; PCV: posterior cardinal vein; SVC: superior vena cava; SV: sinus venosus.
form the common cardinal vein before entering the sinus venosus (Figure 7).

The right anterior and common cardinal veins normally give rise to the SVC. An anastomosis that forms between the right and left anterior cardinal veins become the left innominate (brachiocephalic) vein. The anterior cardinal veins regresses except for a small terminal portion that persist as superior intercostals veins (Figure 8) [6]. When the left anterior cardinal vein regresses, a ligament remains that joins the left superior intercostals vein with the coronary sinus. It is called the ligament of left SVC or Marshall's ligament (Figure 9). If the left

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Table 1. Cases reported with double SVC (persistence left SVC).
innominate vein fails to develop, the left anterior cardinal vein persists and continues to drain the left brachiocephalic veins. In this situation, it becomes the left SVC. The persistent SVC usually drains into the sinus venosus, which ultimately becomes the coronary sinus [7].

The clinical significance of left SVC varies and depends on the existence or lack of symptoms and other congenital heart disease. It can affect the clinicians in imaging diagnosis, central venous catheterization and surgery. Since McCotter first reported three cases in 1916 [8], there were several cases published (Table 1). Steinberg et al. suggested a classification of persistence left superior vena cava into three groups: (1) bilateral superior vena cava without congenital cardiac anomalies, (2) bilateral superior vena cava with associated congenital cardiac anomalies and (3) absence of the right superior vena cava [8].

In our case reports, the central line catheter was seen on the left side of the mediastinum, which was also widened. To an inexperienced physician this can lead to false conclusions, even to the assumption of a major complication. The stability of the patients allowed further investigation with CT scan. In both cases the left innominate vein was absent. Webb et al. reported that double SVC in most cases was associated with regression of the left innominate vein [9]. Widened mediastinum, can be incidentally found in adults and duplication of SVC should be considered as a differential diagnosis [10].

When a dilated coronary sinus is revealed in echocardiography, it may imply the presence of a persistent SVC [11]. Cooper et al. suggested that the gold standard for imaging of persistent left SVC is invasive angiography [12]. However, Ucar et al. proposed 3D reconstructions of CT images for better visualization, as it was performed in our first case [13].

Conclusions

Duplication of SVC is a rather rare entity but extremely important to the awareness of any physician. It can be suspected in several cases when imaging findings contradict the clinical presentation of the patient. Thus, in asymptomatic patients, a widened mediastinum, a dilated coronary sinus or a misplacement of a left internal jugular or subclavian venous catheter in the X-ray, may raise the suspicion of the presence of a left superior vena cava. In that case, there might be technical difficulty in placement of a catheter from the left side of the patient. Further investigation with CTA imaging and echocardiography is essential for diagnosis.

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Consent for publication
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