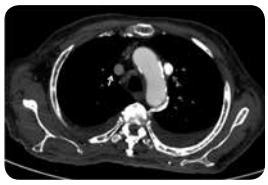

Persistent Left Superior Vena Cava: Case Report

Persistencia de la vena cava superior izquierda: Presentación de un caso

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Palabras clave (DeCS)
Vena cava superior
Seno coronario
Venas cavas

Summary

Persistent left superior vena cava is a rare anatomical variant, which should be known by physicians, since it is relevant for some procedures such as insertion of catheters and pacemakers. Described in the literature since 1950, it has been associated with several cardiac anomalies. This venous structure drains towards several places, which must be known. Although most of the time it is an incidental finding, it has been associated with stroke and death. Imaging modalities such as CT, MRI and echocardiography are helpful for its diagnosis.

Resumen

La vena cava superior izquierda persistente es una variante anatómica poco común, pero su conocimiento por parte de los médicos es importante para algunos procedimientos, como inserción de catéteres, entre otros. Se ha descrito desde 1950 y se asocia con anomalías cardiacas. Esta estructura venosa tiene varios sitios donde drenar los cuales se deben conocer. Casi siempre es un hallazgo incidental, pero se ha asociado a accidente cerebrovascular y muerte. Para su diagnóstico se han utilizado varias modalidades de imagen, como ecocardiografía, tomografía computarizada (TC) y resonancia magnética (RM).

Introduction

The function of the superior vena cava (SVC) is to carry deoxygenated blood from the upper body to the right atrium (1). In this structure the occurrence of congenital abnormalities, which are discovered as incidental findings, is frequent, may be associated with other cardiac abnormalities; acquired causes have also been described (1). Bubble echocardiography, computed tomography (CT), and magnetic resonance imaging (MRI) have been used to visualize and characterize SVC (1).

Case presentation

A 74-year-old male patient, who consults for weight loss and anemia. Studies are carried out in search of endoscopic and endoscopic digestive bleeding, which were normal, angio-CT of the thorax and abdomen in which, in the axial cut to the height of the supraaortic vessels, five vascular structures are identified (Figure 1) and in the aortic arch, a vascular structure on the left side of the mediastinum (Figure 2). In coronal reconstruction, a vessel is seen parallel to the right superior vena cava (Figure 3) that drains into the right atrium through the coronary sinus (Figure 4). In addition, some blast lesions secondary to prostate cancer were identified (Figure 5).

Embriology

In order to understand the persistent left SVC variant, it is necessary to first understand its embryology. In the fifth week of fetal life, there are 3 pairs of cardinal veins that drain the embryo, include the anterior cardinal vein (1, 2) that drains the cephalic portion of the embryo (1, 3), the posterior cardinal vein that drains the portion caudal (1, 3) and common

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cardinal veins that drain blood into the sinus vein (1). For week 8 of gestation (2) anastomosis is formed between the right and left system causing the flow to go from left to right (1, 2), then giving rise to right SVC from the proximal portion of the right anterior cardinal vein, right common cardinal vein (1, 2), and the right horn of the venous sinus (1), while the right posterior cardinal vein forms the azygos vein (1). On the left side, the anterior cardinal vein forms the left superior intercostal vein and the left brachiocephalic vein (1), the other structures involute and form the Marshall›s ligament (1, 3, 4).

The left SVC is formed from the left anterior cardinal vein, the left common cardinal vein and the left venous sinus horn (1, 3).

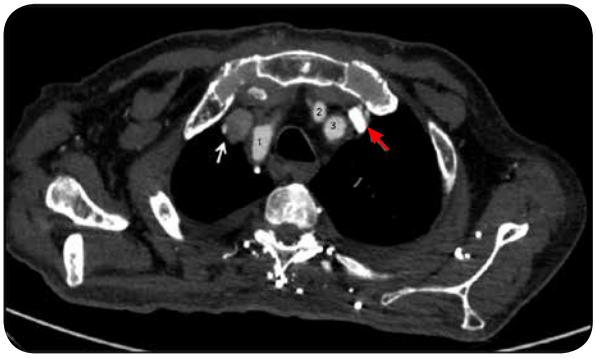

Figure 1. Angio CT, axial section. Five vascular structures at supra-aortic vessels are identified: right superior vena cava (white arrow), left superior vena cava (red arrow), brachiocephalic trunk (1), left common carotid artery (2) and left subclavian artery).

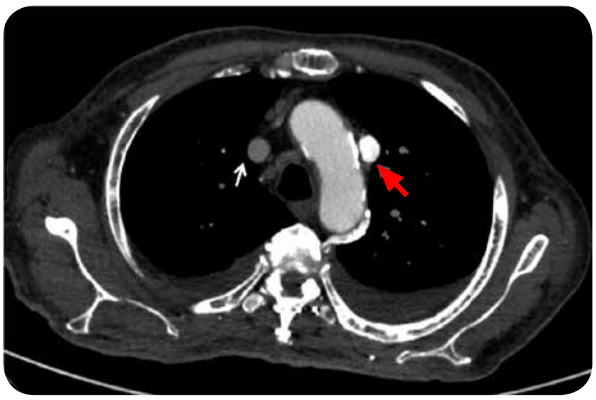

Figure 2. Angio CT, axial section. At the level of the aortic arch there are two vascular structures: right superior vena cava (white arrow) and left superior vena cava (red arrow) and bilateral pleural effusion.

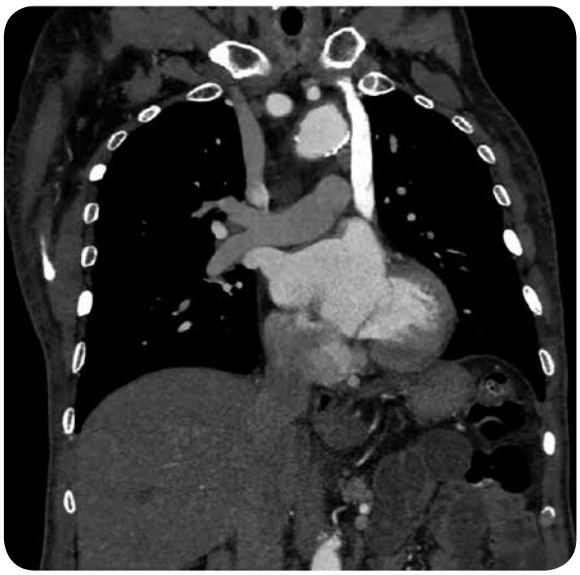

Figure 3. Coronal reconstruction showing two tubular vascular structures parallel to each side of the mediastinum.

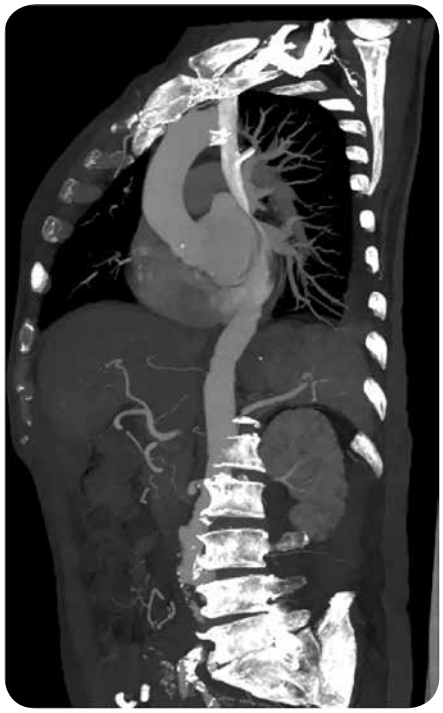

Figure 4. Reconstruction MIP: Curve showing how persistent left SVC drains to the coronary sinus

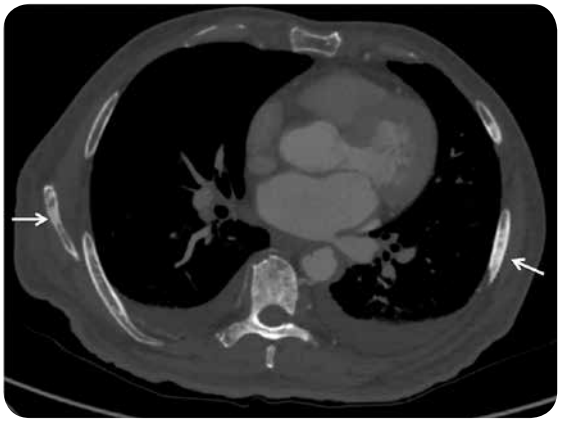

Figure 5. CT angiography, axial cut in bony window: Blast lesions (white arrows) are seen in the sacral arches secondary to prostate cancer.

Persistence of the superior left vena cava

Although a rare congenital anomaly (5), left SVC is the most common venous system of the thorax (1, 2, 4, 6, 7). It was first described in 1950 by Drs Edwards and DuShane (3, 6). It has a prevalence of 0.3 to 0.5% in the general population (1, 5-7) and 4.4 to 12% in patients with congenital heart disease (1, 3, 6, 7). These patients may have a normal right SVC, small or not have it (1, 5, 7). In the latter scenario, cardiac abnormalities are more common (1, 5). When the right SVC exists it can be connected to the left SVC (5, 6). When the two veins cavas are present in 65% of cases the left unnamed vein does not exist (4, 7) or is diminished in size (4).

The most frequently described congenital anomalies associated with absence of right SVC are atrial and ventricular septal defects, tetralogy of Fallot, bicuspid aortic valve, coarctation of the aorta and mitral atresia (1, 3-5). The most common associated malformation is the atrial septum defect (7).

The normal course of this structure is lateral to the aortic arch. In the middle of its course, the vessel is located anterior to the left hilum, then crossed by the Marshall's ligament and drained into the right atrium through the coronary sinus (1, 4-6), which occurs in 90% of the cases (2, 3). In some cases, it does not drain into the right atrium, but rather it drains into the left (3, 5), creating a right-to-left short circuit (1, 3, 6), known as Raghib syndrome (1). Drainage has been reported in the left superior pulmonary vein (7).

When it drains into the coronary sinus, it is larger (3, 6), so it should be considered in the differential diagnosis (3, 6), when this finding is present (6)

Clinical Presentation

These patients are usually asymptomatic and the finding is made by imaging studies performed for another reason (1-3, 5, 6) or when a procedure is performed in the left subclavian vein (4). In some cases these patients may have systemic embolization (1, 2) with symptoms of stroke, brain abscess (1) or death (2).

Another clinical implication of this anomaly is that it may create some difficulty in the implantation of pacemakers, cardiodesfibrillators and other catheters (1-3, 6). It has also been associated with anatomical and architectural abnormalities of the sinus node (5), which predisposes arrhythmias, the most common atrial fibrillation (4).

Diagnostic Imaging

Echocardiography: Coronary sinus dilation is observed with confirmation of left SVC when saline solution is used (5). Axial imaging modalities, such as CT and MRI, aid the diagnosis because an abnormal vessel (5) is evidenced with the expected path for left SVC. In addition, it is useful because it helps to know where it drains, whether or not there is SVC right and if there is communicating vessel between the two cava veins.

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