Multisystem

Hemiazygous-accessory hemiazygous continuation of double inferior vena cava

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ARTICLE INFO

Article history:
Received 1 July 2017
Received in revised form 22 August 2017
Accepted 28 August 2017
Available online

Keywords:
Hemiazygous-accessory
Hemiazygous continuation of double IVC

ABSTRACT

We report a case of hemiazygous-accessory hemiazygous continuation of a double IVC with absent azygous vein, incidentally discovered in an adult patient.

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Introduction

Whereas congenital abnormalities of the infradiaphragmatic systemic venous return, such as a double inferior vena cava (IVC) or an azygous continuation of the interrupted IVC, are relatively common, certain anatomic variants are exceedingly rare. Here we report a case of an extremely rare variant of a hemiazygous-accessory hemiazygous continuation of a double IVC with an absent azygous vein in an adult female patient. Recognition of such venous anomaly is important as it may simulate pathology on imaging, may have significant implications for central venous catheterization and surgical planning, and may be sometimes associated with congenital heart disease, heterotaxy, and increased risk of venous thromboembolism.

Case presentation

An adult woman, known for Parkinson disease and a surgical history of remote cholecystectomy, hysterectomy, and left breast lumpectomy with left axillary dissection, underwent various imaging studies in our institution between 58 and 64 years of age. At the age of 67, the patient was diagnosed with acute myeloid leukemia, which led to more diagnostic imaging and radiological monitoring of the disease and treatment-related complications.

A contrast-enhanced computed tomography of the abdomen and pelvis performed for the investigation of lower abdominal pain, when the patient was 64 years old, demonstrated a double IVC, with a right-sided IVC interrupted at the right renal vein, a retroaortic communication between the infrarenal right...
Fig. 1 – Contrast-enhanced axial computed tomography of the abdomen and pelvis (A, B) and coronal oblique reformations (C, D) demonstrate double IVC, with retroaortic communication (red arrows) between the right IVC (yellow arrows) and the left IVC (green arrows), the latter continuous with the enlarged hemiazygous vein (blue arrow). IVC, inferior vena cava.

Fig. 2 – Axial computed tomography pulmonary angiography (A, B), coronal oblique (C) and sagittal (D) reformations demonstrate HAz-AccHAz continuation of the left IVC, draining into the dilated SupInt, which in turn drains into the LBCV and then into the normal right-sided SVC. AccHAz, accessory hemiazygous; HAz, hemiazygous; IVC, inferior vena cava; LBCV, left brachiocephalic vein; SupInt, superior intercostal vein; SVC, superior vena cava.
and left IVCs, and a hemiazygous continuation of the left IVC (Fig. 1).

Computed tomography pulmonary angiography performed subsequently for the investigation of dyspnea, showed the absence of the azygous vein, and a hemiazygous-accessory hemiazygous continuation of the left IVC draining into the left brachiocephalic vein via a dilated superior intercostal vein, which in turn drained into the normal right superior vena cava and the right atrium (Fig. 2).

A schematic representation of the normal central venous anatomy and of the venous anomalies seen in our patient is shown in Figure 3.

Discussion

The IVC and the azygous-hemiazygous venous system develop between weeks 6 and 8 of embryogenesis through an ordered sequential formation and regression of 3 pairs of embryonic veins—posterior cardinal, subcardinal, supracardinal, and the extensive anastomoses between them [1]. In normal development, the infrahepatic IVC arises from the right subcardinal vein and the lower portion of the right supracardinal vein; the hepatic portion of the IVC arises from the vitelline vein, which connects to the infrahepatic IVC by the formation of the subcardinal-hepatic anastomosis; the azygous and hemiazygous veins arise from the upper portions of the right and left supracardinal veins, respectively [1].

The middle and lower portions of the left supracardinal vein normally involute. Failure of the latter will result in a double IVC, with the left-sided IVC typically ending at the left renal vein, which drains into the right-sided IVC [2].

In our patient, it is the right-sided IVC that ended at the right renal vein; the right and left IVCs were connected by a retroaortic vascular bridge, and the left-sided IVC then continued as a hemiazygous vein (Figs. 1 and 3). Such an arrangement is postulated to result from the persistent left supracardinal vein and the simultaneous failure of formation of the right subcardinal-hepatic venous anastomosis [1].

Furthermore, the azygous vein was absent in our patient. Thus, the venous return from the lower body reached the normal right-sided superior vena cava and the right heart successively via the hemiazygous vein, the accessory hemiazygous vein, the superior intercostal vein, and the left brachiocephalic vein (Figs. 2 and 3).
An isolated double IVC variant is relatively common, with an estimated incidence of 0.2%-3.0% [1]. A hemiazygous-accessory hemiazygous continuation of the left IVC is rare; our search of the English-language medical journal databases has yielded only 2 reports of such cases [2,3]. Only 1 report of findings similar to our case could be found, describing a double IVC with a retroaortic right renal vein and a hemiazygous-to-azygous continuation of the left IVC in a 2-year-old patient with spinal dysraphism [1]. To our knowledge, this is the first report of a combination of double IVC and a hemiazygous-accessory hemiazygous continuation of the IVC with an absent azygous vein in an adult patient.

Recognition on radiological imaging of anatomic venous variants such as the one reported here, is important for a number of reasons. The presence of some variants may signal possible concomitant cardiovascular anomalies, especially congenital heart disease and heterotaxy syndromes. The associated dilation of the azygous or accessory hemiazygous and superior intercostal veins may result in an unusual radiographic appearance of the mediastinal contours (Figs. 2 and 4), which can be misinterpreted as lymphadenopathy, abnormally dilated lymphatics, or aortic dissection or aneurysm. In addition, the radiological assessment post insertion of central venous lines can be quite confusing in such patients as illustrated in Figure 4. An aberrant venous return can lead to venous insufficiency of the lower limbs with potential for thromboembolic disease [4]. Finally, the recognition of aberrant venous anatomy can have important implications during surgery or endovascular procedures, as illustrated by a previously reported fatal outcome after ligation of the aberrant vessel in a patient with a hemiazygous continuation of the left IVC [5].

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