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Case report

A remarkable case report of an interrupted inferior vena cava with hemiazygos and transhepatic continuation

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Abstract. Inferior vena cava (IVC) interruption with azygos/hemiazygos continuation is an extremely uncommon congenital vascular anomaly, which may present with multiple variants. As a result, it is challenging to find in the literature the same anatomical variant. We report a unique case of an interrupted IVC with hemiazygos and transhepatic continuation in an 83-year-old female patient. The case was evaluated by performing Computed Tomography (CT) as imaging modality, with a multiphase protocol, able to detect accurately this complex vascular anomaly. The purpose of this case report is not only to present this remarkable case but also to briefly show the types of interrupted IVC, starting from the anatomy and the embryology of the IVC and the azygos system, and to discuss the value of imaging in detecting the vascular anomaly. (www.actabiomedica.it)

Key words: hemiazygos continuation, inferior vena cava, interrupted IVC, transhepatic continuation

Introduction

The Inferior Vena Cava (IVC) is a wide retroperitoneal vein that drains deoxygenated blood from the lower limbs, pelvic, and abdominal viscera to the heart's right atrium (1).

The azygos venous system is a paired paravertebral venous pathway in the posterior thorax that provides collateral circulation between the Superior Vena Cava (SVC) and the IVC. The azygos, hemiazygos, accessory hemiazygos, and left superior intercostal veins form an H-shaped arrangement which is typical of the azygos venous system (2).

Interruption of IVC is a rare congenital anomaly in which venous return from the lower body occurs via azygos or hemiazygos venous system (3). The term "interrupted" is used to indicate a complete agenesis as well as an anomalous course.

The anomaly primarily results from abnormal regression or persistence of embryological veins (anterocardinal, post-cardinal, sub-cardinal, supra-cardinal, and vitelline veins) that formed the five embryologic segments of the final structure of the IVC (iliac, subrenal, renal, suprarenal, hepatic including suprahepatic and retrohepatic) (1,3).

In most cases, the interruption of the IVC occurs because of a failed anastomose between the right subcardinal vein and the vitelline vein, resulting in the agenesis of the infrarenal IVC and the interruption at the suprarenal segment. In this circumstance,

the suprarenal IVC reroutes to drain via the azygos vein, while the hepatic IVC only receives the hepatic veins (4).

In other cases, the suprahepatic IVC may be absent or hypoplastic, resulting in direct drainage into the right atrium. In this circumstance as well the azygos system would be enlarged.

Subrenal interruption or absent IVC also has been reported but is less common (2).

These anatomical variants represent an asymptomatic condition if associated with well-developed azygos/hemiazygos continuation, but they may present with recurrent deep vein thrombosis (DVT) of the lower limbs, leg swelling, leg pain, varices of lower extremities, abdominal pain, and rarely hematochezia.

The interruption of the IVC may be isolated or associated with other anomalies, such as congenital cardiac disease, polysplenia (left isomerism), abnormal abdominal situs, and azygos lobe.

In an asymptomatic condition it is typically detected incidentally in the early to middle ages of life (5,6).

To study the vascular anomaly, imaging modalities include echocardiographic techniques, Color-Doppler imaging, CT-angiography, and ICV-angiography, that help to detect interrupted ICV, to identify the anomalous course of the main vessels and to evaluate the dilatation of the azygos system caused by increased flow. Especially, angiography is important to recognize the exact anatomy of the drainage for surgical purposes. Therefore, detection of these venous anomalies is significant because it would hamper right heart catheterization via the femoral vein approach as well as cardiopulmonary bypass surgery and pacemaker placement (7).

Case presentation

An 83-year-old Caucasian woman came for the first time to our department to undergo a Total-Body CT with medium contrast administration for a lung tumor follow-up. Other CTs were already performed in other clinics, but reports were not brought to view at the time of the investigation.

She referred to not having a prior history of coronary artery disease, congestive heart failure, hypertension, diabetes, or a history of deep venous thrombosis.

The study protocol followed the investigation motivation as per medical request and consequently, it was not the most appropriate for the study of vascular anomalies. Therefore, it was performed a multiphase examination including a first unenhanced scan, followed by post-contrast scans including an arterial and portal venous phase, with a 64-detector scanner. In order to better evaluate the anomaly, post-processing reconstruction on all planes (axial, coronal, sagittal), also with MIP application, and 3D images were obtained.

At first look, the most evident imaging finding about the vascular anomaly concerned the non-visualized *IVC* in the usual right side of the aortae.

In this case, the paired common iliac vein joined a distended *hemiazygos vein* that received the left renal vein to end continuing in the accessory hemiazygos vein.

The enlarged *accessory hemiazygos* made a visualizing arch that followed externally the one of the aortae, to join in the end the SVC, which appeared enlarged as well (Figure 1).

The *suprahepatic veins* flew out directly into the right atrium, but one of them passed entirely through the liver and continued outside originating a *transhepatic continuation*, that joined the *confluence* between the azygos vein and the right renal vein. From the right common iliac vein, a wide atypical vein departed joining the confluence. In this way, the connection between the azygos and hemiazygos veins was obtained (Figure 2).

The *azygos vein* followed its typical connection with the SVC through its arch, but its origin was anomalous, considering it started after it overpassed at L1-L2 level the diaphragmatic pillar from the confluence between the right renal vein, the transhepatic vein, and an *anomalous vein*. (Figure 3)

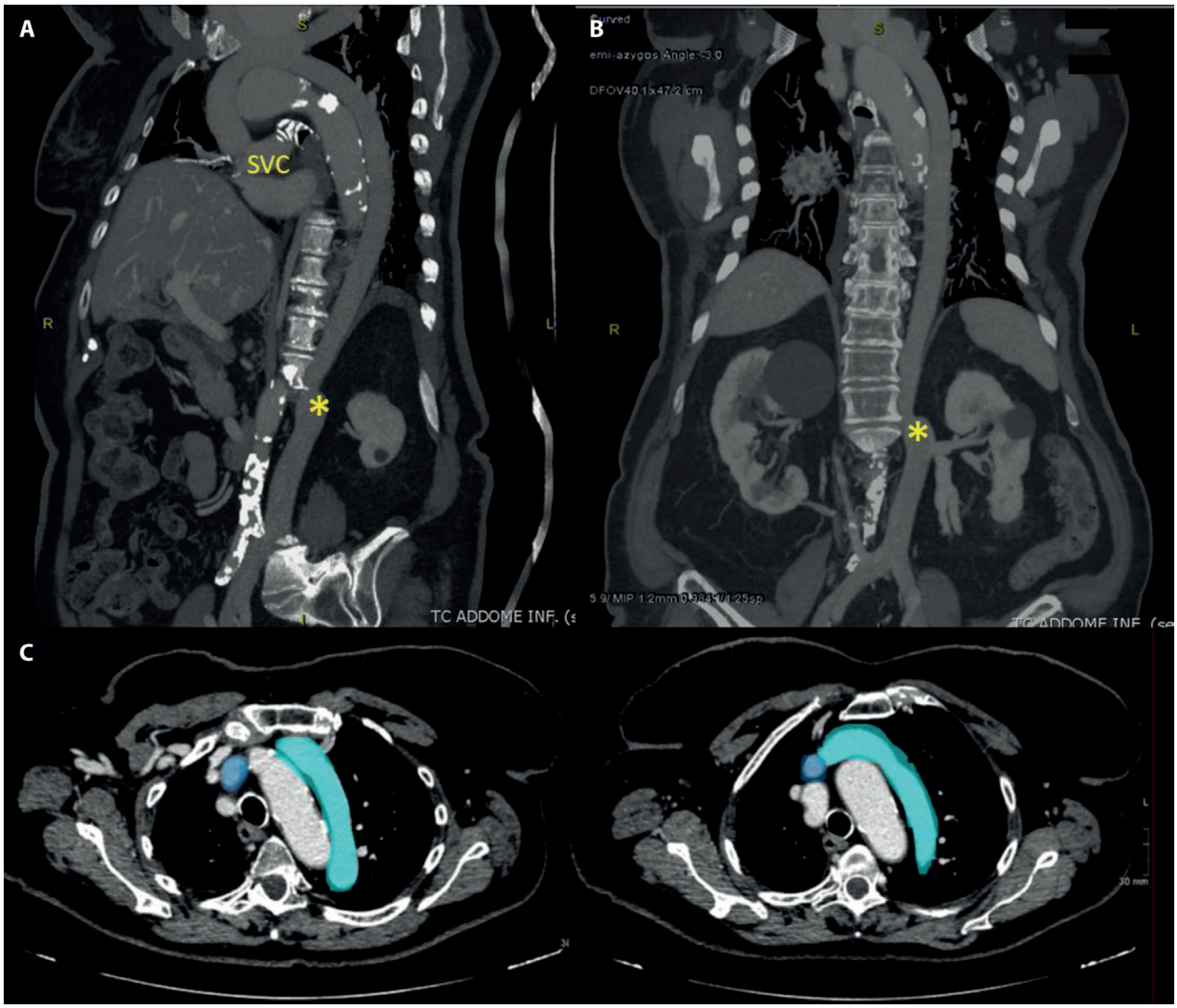

Figure 1. Portal phase of a Whole Body-CT scan with MIP application. a) sagittal plane showed the accessory hemiazygos arch from the Superior Vena Cava (SVC) and the continuation of the hemiazygos vein. b) coronal plane showed the accessory hemiazygos arch, the entire course of the hemiazygos vein flowing at the left side of the vertebral column, its origin from the common iliac veins. At the L1-L2 level, the hemiazygos overpassed the diaphragmatic pillar (*) c) axial plane images showed the entire arch on the left side of the aortae and the accessory hemiazygos arch joining the Superior Vena Cava. Blue: Superior Vena Cava; Light blue: accessory hemiazygos arch.

Discussion

The interruption of the ICV with azygos system compensation is a complex and rare vascular anomaly that presents multiple variants.

In the case presented the patient had got: an intrahepatic interrupted ICV with the suprahepatic veins flowing out directly in the right atrium; a transhepatic continuation of one suprahepatic vein connected to the azygos vein; an enlarged hemiazygos system as main

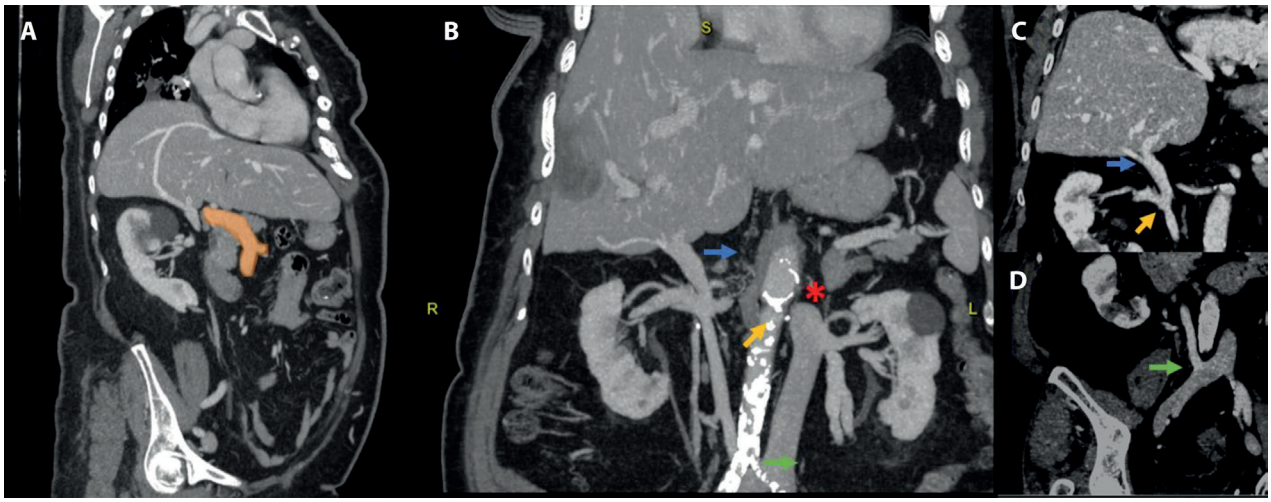

Figure 2. Portal phase of Whole-Body CT scan. a) curved reformatted image of the transhepatic continuation of one of the suprahepatic veins. There was no shunt with the portal system, as there was no communication with the splenic-portal axes (colored in orange) b) showed the anomalous vein originating from the right common iliac vein, the confluence of the right renal vein, and the azygos vein, the transhepatic continuation. c) and d) showed respectively the origin from the right common vein and the continuation in the liver. Green arrow: origin; Yellow arrow: confluence; Blue arrow: transhepatic continuation; Red asterisk: azygos vein.

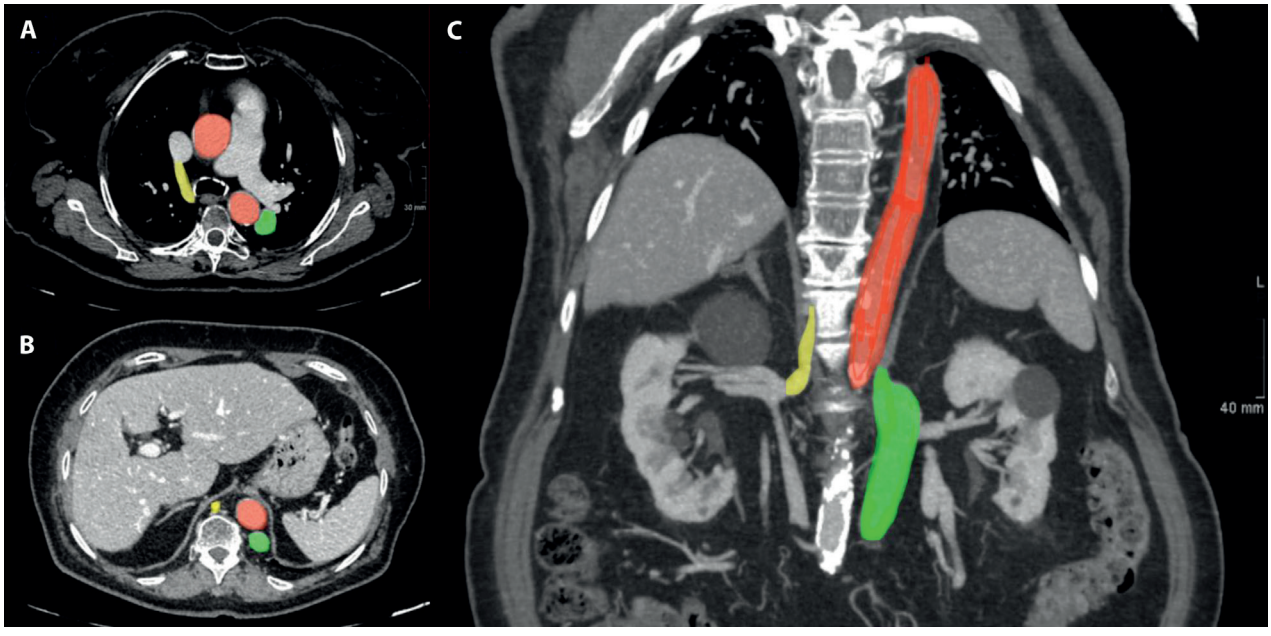

Figure 3. Portal phase of a Whole-Body CT scan with MIP application. a) axial plane showed the typical azygos arch. b) axial plane showed from right to left the azygos vein, the aortae, and the enlarged hemiazygos. c) coronal plane showed the azygos overpassing the diaphragmatic pillar at the L1-L2 level and its confluence with the right renal vein and the transhepatic continuation. Yellow: azygos; Red: aortae; Green: hemiazygos.

drainage system. All conditions are rare and unusually to find simultaneously (7).

The role of imaging in the detection of this type of vascular anomaly is essential: CT in the case presented allowead a good differentiation of all vessels and the detection of the type of variant as well as the enlargement of all the azygos system.

What makes this anatomical variant truly unusual can be summarized as follows:

- the interruption of the ICV was intrahepatic and not at a suprarenal level, which is the most common variant.
- the compensating main vessel corresponded to the accessory hemiazygos and the hemiazygos vein, with the latter that totally substitutes the absent ICV. Therefore, it originated from the two paired common iliac veins.
- the arch formed by the accessory hemiazygos vein embracing that of the aorta.
- the continuation of the accessory hemiazygos vein directly with the hemiazygos vein.
- the transhepatic continuation of one of the suprahepatic veins, originating an intrahepatic shunt.
- the confluence among the azygos vein, the transhepatic continuation, and the right renal vein.
- the connection through a collateral vein between the confluence above-mentioned and the right common iliac vein, originating a shunt between the azygos and the hemiazygos system (Figure 4).

Hemiazygos' continuation of an interrupted IVC has several variations, including three possible paths for blood in the hemiazygos vein to reach the right atrium. In the first paths, the hemiazygos vein joins the azygos vein at T8–T9. In this route, the findings at more cranial levels are like azygos continuation with enlargement of the distal azygos vein, as well as the enlargement of the hemiazygos vein itself. The second path involves a persistent left SVC, and blood flows from the hemiazygos vein into the accessory hemiazygos vein and left SVC and then into the coronary sinus, all of which are dilated. The third path, which is the one reported in this case, the hemiazygos vein drains to the accessory hemiazygos vein to join into an usual right SVC (2,8).

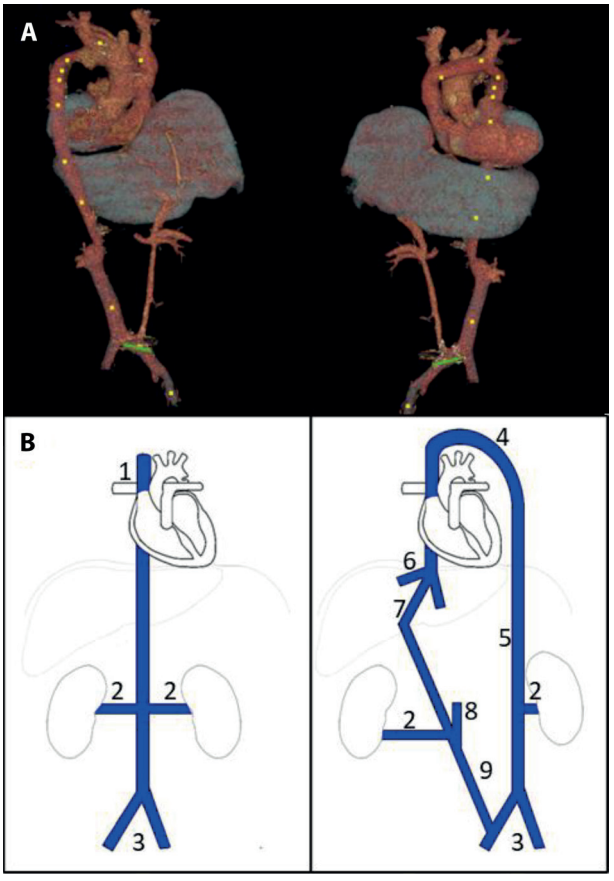

Figure 4. 3D reconstruction of the main vessels involved in this vascular anomaly from a posterior and anterior view (a) compared to a schematic drawing of normal anatomy and the aberrant abdominal venous pathways of this reported case (b). a) showed the arch, the hemiazygos continuation until the division in the common iliac veins and the wide vein from the right common iliac vein connecting the transhepatic continuation, the right renal vein, and the azygos vein. In b) veins are numbered as follows: 1 inferior vena cava IVC; 2 renal vein; 3 common iliac vein; 4 accessory hemiazygos arch; 5 hemiazygos vein; 6 suprahepatic veins; 7 transhepatic continuation; 8 azygos vein; 9 wide vein connecting the right common iliac vein with the confluence of 2, 7 and 8.

ICV interruption with hemiazygos continuation is a benign condition and, in this case, as vascularization was well compensated, no treatment was needed. Otherwise, the knowledge of the condition was crucial for the patient in case of necessary surgical intervention (9).

In addition, a misdiagnosis could have been done considering that this anomaly at chest x-ray in the posterior anterior film could present as an enlargement of the mediastinal shadow (that might be confused with adenopathy of aortic pathology) while, in the lateral film, ICV shadow should be absent; or at Transesophageal Echocardiography (TEE) the dilated azygos or hemiazygos vein lies adjacent to the descending aorta might mimic an aortic pathology (5,10).

Considering that interrupted ICV is commonly associated with other congenital anomalies, especially in the cardiac field, a prompt search for additional pathologies was evaluated. Moreover, excluding portosystemic shunting was important for determining management as persistent congenital portosystemic shunts could be associated with significant morbidity. In the case reported, there was no correlation with other congenital anomalies and there was no evidence of an association with the tumor affecting the patient. The two conditions can be considered independent.

In conclusion, we reported a case of a rare venous anomaly characterized by an interrupted ICV with a developed hemiazygos and transhepatic continuation. Both conditions are extremely rare, and their combination is poorly described in the literature.

The presented case gave the occasion to emphasize the importance of imaging in the detection of complex vascular anomalies, such as the one described. Physicians should be aware of this unusual vascular anomaly both to avoid misdiagnosis and to aid surgical intervention.

Inform Consent: Consent to participate and for publication. Written informed consent for publication was obtained from the patient.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

References

1. Li SJ, Lee J, Hall J, Sutherland TR. The inferior vena cava: anatomical variants and acquired pathologies. Insights Imaging. 2021;12(1):123. doi:10.1186/s13244-021-01066-7.
2. Demos TC, Posniak HV, Pierce KL, Olson MC, Muscato M. Venous anomalies of the thorax. Am J Roentgenol. 2004;182(5):1139-1150. doi:10.2214/ajr.182.5.1821139.
3. Ghandour A, Partovi S, Karuppasamy K, Rajiah P. Congenital anomalies of the IVC—embryological perspective and clinical relevance. Cardiovasc Diagn Ther. 2016;6(6): 482-492. doi:10.21037/cdt.2016.11.18.
4. Vignesh S, Bhat TA. Unique medley of cardinal veins: duplicated superior and inferior venae cavae with left renal agenesis and hemiazygos continuation of left inferior vena cava with drainage into left atrium. Vasc Endovascular Surg. 2022;56(3):330-334. doi:10.1177/15385744211051493.
5. Liu Y, Guo D, Li J, et al. Radiological features of azygos and hemiazygos continuation of inferior vena cava: a case report. Medicine (Baltimore). 2018;97(17):e0546. doi:10.1097 /MD.0000000000010546.
6. Chen SJ, Wu MH, Wang JK. Clinical implications of congenital interruption of inferior vena cava. J Formos Med Assoc. 2022;121(10):1938-1944. doi:10.1016/j.jfma.2022.01.021.
7. Sahin H, Pekcevik Y, Aslaner R. Double inferior vena cava (IVC) with intrahepatic interruption, hemiazygos vein continuation, and intrahepatic venous shunt: a rare variant of IVC anomalies. Vasc Endovascular Surg. 2017;51(1):38-42. doi:10.1177/1538574416687734.
8. Koc Z, Oguzkurt L. Interruption or congenital stenosis of the inferior vena cava: prevalence, imaging, and clinical findings. Eur J Radiol. 2007;62(2):257-266. doi:10.1016/j .ejrad.2006.11.028.
9. Mandato Y, Pecoraro C, Gagliardi G, Tecame M. Azygos and hemiazygos continuation: An occasional finding in emergency department. Radiol Case Rep. 2019;14(9): 1063-1068. doi:10.1016/j.radcr.2019.06.003.
10. Holemans JA. Azygos, not azygous. Am J Roentgenol. 2001;176(6):1602-1602. doi:10.2214/ajr.176.6.1761602b.

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