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Case Report

Transposition of the inferior vena cava with hemiazygos continuation: A rare case report ☆

Sara Ez-zaky, MD, Ihssan Hadj Hsain, MD, Sanae Jellal, MD, Sara Essetti, MD, Jamal El Fenni, PhD, Rachida Saouab, PhD, Ouijdane Zamani, MD*

Radiology Department, Mohamed V Military Training Hospital, Mohammed V University, Rabat, Morocco

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ABSTRACT

Hemiazygos continuation of a left-sided inferior vena cava (IVC) is an extremely rare developmental anomaly. We present the case of a male patient in whom this condition was incidentally discovered during a thoraco-abdominopelvic CT scan. With the widespread use of contrast-enhanced computed tomography and magnetic resonance angiography, the detection of congenital IVC anomalies has become more accessible. These anomalies are often found incidentally during imaging performed for other clinical reasons. Awareness of this anomaly is crucial before any surgical or endovascular procedures to avoid potential complications.

Introduction

Developmental variations of the inferior vena cava (IVC) are found in the general population, with a prevalence ranging from 0.07% to 8.7% [1]. IVC anomalies can be classified into prerenal anomalies (interrupted IVC), renal anomalies (retroaortic renal vein and circum-aortic venous collar), and postrenal anomalies (duplicated IVC, left-sided IVC, and retrocaval ureter) [2]. Left-sided IVC and azygos or hemiazygos continuation of the IVC have prevalences of 0.2% to 0.5% and 0.6%, respectively, and are typically asymptomatic, incidental findings [3]. The exact prevalence of the combined occurrence of left-sided IVC with hemiazygos continuation is unknown due to the rarity of reported cases. With the advent of advanced imaging techniques such as CT and MRI, it has become imperative to understand the various anatomical variations of the inferior vena cava and the azygos system, as these techniques provide valuable insights into these structures, including the variation described in our case. A thorough understanding of the congenital anomalies of the IVC and the azygos system is essential for radiologists to prevent diagnostic errors and to aid in preoperative planning. We report a rare case of transposition of the inferior vena cava with azygos continuation, associated with polysplenia and a short pancreas.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: sara92ezzaky@gmail.com (S. Ez-zaky).

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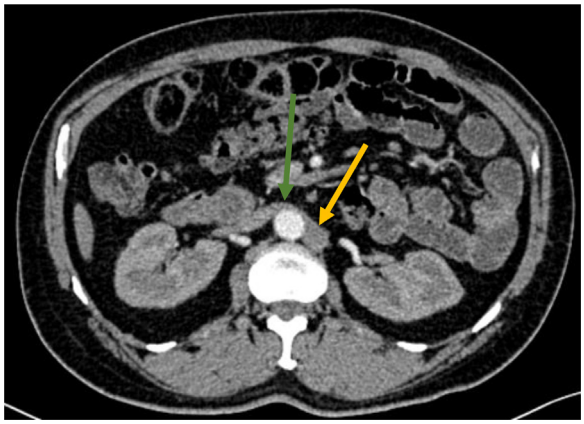

Fig. 1 – Axial section of an abdominal CT scan after injection of contrast medium into an arterial beat showing a left inferior vena cava (yellow arrow) in relation to the aorta (green arrow).

Case report

This is a 60-year-old man with no significant medical or surgical history and no notable family history, who presented with epigastric abdominal pain for the past month, which later became diffuse.

A blood test was conducted, revealing a lipase level of 60 IU/L and a CRP level of 7 mg/L.

An abdominal ultrasound was performed, revealing 2 hepatic lesions that could not be accurately characterized by the ultrasound. A thoraco-abdomino-pelvic CT scan was requested for better characterization and to search for a primary tumor.

The contrast-enhanced CT scan incidentally revealed a left-sided inferior vena cava with hemiazygos continuation:

- The 2 iliac veins drain into the left-sided inferior vena cava (Fig. 1), which then joins the hemiazygos vein (Fig. 2A) before converging with the azygos vein (Fig. 2B), which drains into the superior vena cava via the azygos arch (Fig. 3).

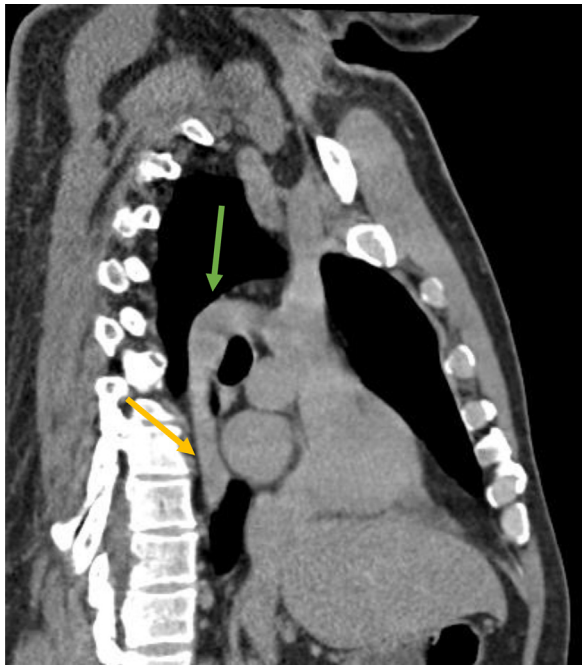

Fig. 3 – Sagittal reconstruction of a thoraco-abdomino-pelvic CT with contrast injection showing the drainage of the azygos vein (yellow arrow) into the superior vena cava via the arch of the azygos vein (green arrow).

- The hepatic veins drain directly into the right atrium (Fig. 4).
- Associated anomalies include polysplenia (Fig. 5) and a short pancreas (Fig. 6).

The CT scan confirmed that the hepatic lesions initially observed on the ultrasound were hepatic hemangiomas. No abnormalities were found on the scan to explain the patient's abdominal pain. The patient was treated with symptomatic management, leading to good clinical improvement.

In conclusion, there was a fortuitous discovery of a transposition of the inferior vena cava (IVC) with hemiazygos continuation.

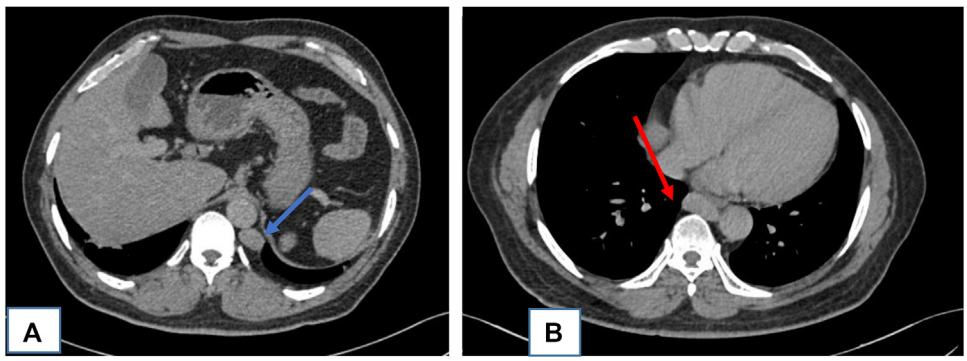

Fig. 2 – Axial slice of a thoraco-abdominal CT scan with contrast injection in the portal phase, showing the inferior vena cava draining into the hemiazygos vein (blue arrow), which then joins the azygos vein (red arrow).

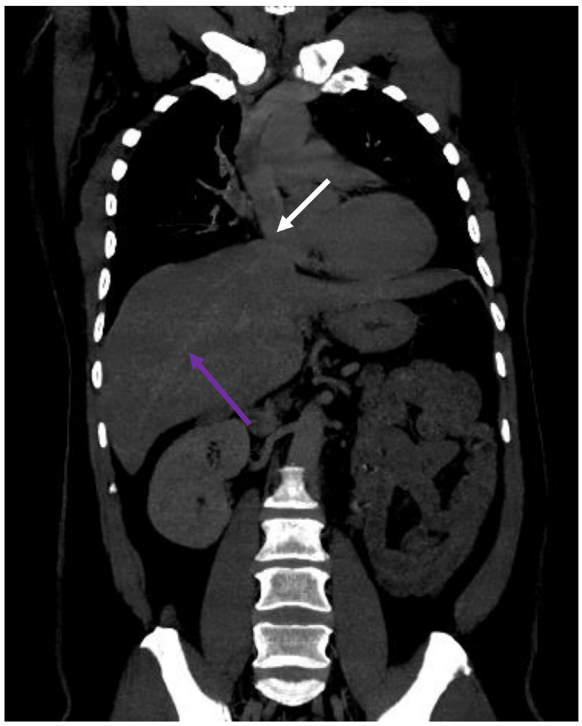

Fig. 4 – Coronal reconstruction of a thoraco-abdomino-pelvic CT showing the drainage of the hepatic veins (Violet arrow) into atrium (White arrow).

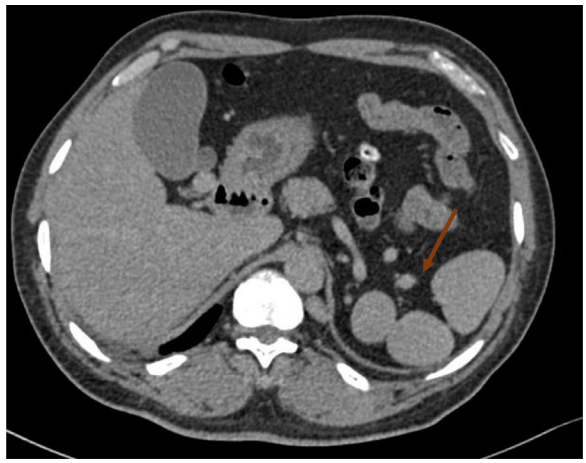

Fig. 5 – Coronal reconstruction of a thoraco-abdomino-pelvic CT showing polysplenia (brown arrow).

Discussion

The inferior vena cava (IVC) is a single vein situated to the right of the abdominal aorta. Failure in the process of embryogenesis can lead to congenital anomalies of the IVC [4]. The development of the IVC is a complex process that occurs between the fourth and eighth weeks of gestation. It involves 3 pairs of primitive venous channels: the posterior cardinal veins, the subcardinal veins, and the supracardinal veins [5]. The normal IVC is composed of 4 segments: the hepatic, prerenal, renal, and postrenal segments. It collects blood from the lower limbs, pelvic region, and abdomen [6,7] .

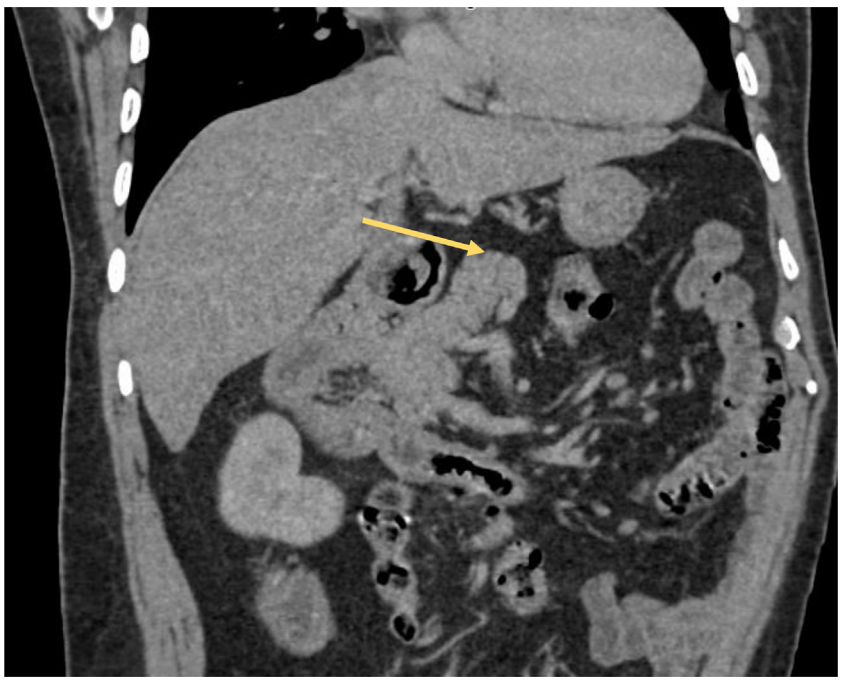

Fig. 6 – Coronal reconstruction of a thoraco-abdominal CT scan showing a short pancreas (yellow arrow).

Infrahepatic interruption of the IVC with azygos or hemiazygos continuation is a rare condition, most likely resulting from agenesis of an IVC segment or failure of fusion between the adrenal and hepatic segments of the IVC [8]. In such cases, the hepatic veins drain directly into the right atrium (Fig. 3), and venous blood from the lower body is redirected into the azygos or hemiazygos vein. The azygos vein then drains into the superior vena cava, ultimately reaching the right atrium [9]. During early embryogenesis, venous drainage from the left and right sides of the body occurs independently. After the regression of most of the left supracardinal veins and the interconnections between the sacrocardinal veins, the entire venous drainage from the left lower limb shifts to the right side, forming the IVC. Disruptions in this developmental process can result in anatomical variants, such as a left-sided IVC [10]. An isolated left-sided IVC ascends on the left and may drain into the right atrium through various pathways. In the first scenario, the left-sided IVC crosses the midline anterior to the aorta after receiving the renal veins, then continues on the right side to drain into the right atrium. In the second scenario, the IVC crosses the midline but is interrupted and continues as the azygos vein, which then connects to the superior vena cava before draining into the right atrium. Lastly, in less common cases like ours, the left-sided IVC may continue caudally on the left as the hemiazygos vein [11]. Azygos and hemiazygos continuation of the IVC is often associated with complex malformation syndromes, such as congenital heart defects, polysplenia, and situs anomalies [12]. In our case, it is associated with polysplenia and a short pancreas.

Noninvasive imaging techniques like contrast-enhanced computed tomography and magnetic resonance imaging are the most reliable methods for identifying these anomalies in asymptomatic patients [8]. Ultrasound is a noninvasive, radiation-free method often used for initial evaluation; however, it is an operator-dependent modality, and the examination of the infrarenal part of the IVC is often limited by gas and adipose tissue [13]. In pediatric patients, ultrasound is generally the first-line modality for cross-sectional imaging. If ultrasound is insufficient due to operator or patient-related factors, magnetic resonance imaging is sensitive for detecting IVC anomalies [14]. CT is commonly used to assess the IVC; its advantages include excellent spatial resolution and the availability of postprocessing techniques such as multiplanar reconstruction of volume-rendered images. Routine imaging is performed between 60 and 70 seconds (portal venous phase) after contrast administration [14]. Another advanced imaging technique for IVC evaluation is magnetic resonance imaging (MRI). MRI, particularly 3-dimensional T1-weighted imaging enhanced with a contrast agent during breath-hold and balanced steady-state free precession techniques [13], offers detailed assessment. Magnetic resonance venography (MRV) is an effective method for identifying developmental anomalies of the IVC. It can be performed with or without contrast agents (gadolinium) [15].

IVC anomalies generally do not require treatment but can be significant for planning vascular or surgical interventions [2].

Conclusion

Left-sided IVC with hemiazygos continuation is an extremely rare anomaly. It is crucial to recognize and identify this anatomical variant before planning any vascular or interventional procedures involving the IVC. CT is the imaging modality of choice for detecting this variation. In every abdominal or thoraco-abdominal CT scan, it is essential to analyze the vascular structures, especially veins, and describe any anatomical variations.

Patient consent

Written informed consent was obtained from the patient for the publication of this case report.

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