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Hypoplasia of the inferior vena cava with hypertrophic *azygos/hemiazygos* and collateral venous circles of the abdomen: a case report

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ABSTRACT

Hypoplasia of the *inferior vena cava* is a rare congenital vascular defect with various forms; thus, identifying a specific anatomical variant in the literature is challenging. In some cases, the *inferior vena cava* may also be interrupted. Herein, we present a unique case of of an unknown subrenal hypoplasia of the *inferior vena cava* with *azygos* and *hemiazygos* hypertrophy and the creation of several collateral circles, particularly in the anterior wall of the abdomen, in an asymptomatic 75-year-old man. This report not only describes this unusual instance but also quickly demonstrates the variations of the venous system in the abdomen, particularly on the right side, and the *inferior vena cava* and the *azygos* system, and explains the significance of imaging in recognizing vascular anomalies. The case was explored using a multiphase computed tomography technique, which correctly identified this complex vascular anomaly. The patient had never experienced symptoms associated with the same vascular defect previously. Moreover, his symptoms did not appear to be related; therefore, a periodic follow-up was recommended.

Keywords: hypertrophic azygos; inferior vena cava; inferior vena cava hypoplasia; collateral venous circles; venous system.

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Гипоплазия нижней полой вены, сопровождающаяся гипертрофией непарной и полунепарной вен и образованием сети коллатеральных вен в брюшной полости: клинический случай

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АННОТАЦИЯ

Гипоплазия нижней полой вены — это редкая врождённая сосудистая аномалия, которая отличается разнообразием форм. В некоторых случаях нижняя полая вена прерывается. Именно поэтому поиск описания определённых анатомических вариантов данной аномалии в литературе — это достаточно трудная задача. В настоящей статье представлен уникальный случай бессимптомной гипоплазии инфраренального сегмента нижней полой вены, сопровождающейся гипертрофией непарной и полунепарной вен, а также формированием сети коллатеральных вен на передней брюшной стенке. Сосудистая аномалия выявлена случайно у мужчины 75 лет. Помимо описания клинического случая, в статье кратко охарактеризованы сопутствующие изменения венозной системы брюшной полости, особенно выраженные с правой стороны, а также изменения нижней полой вены и системы непарной вены. В работе также приведено обоснование важности проведения визуализационных исследований для выявления сосудистых аномалий. В представленном клиническом случае визуализация выполнена с помощью компьютерной томографии с многофазным контрастированием, что позволило точно определить наличие сложной сосудистой аномалии. У пациента ранее никогда не возникало симптомов, указывающих на наличие данной аномалии, а проявлявшиеся симптомы, по-видимому, не были с ней связаны, поэтому пациенту рекомендовано периодическое наблюдение.

Ключевые слова: гипертрофия парной вены; нижняя полая вена; гипоплазия нижней полой вены; коллатеральные вены; венозная система.

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下腔静脉发育不全伴奇静脉和半奇静脉肥大及腹腔侧支静脉网络形成：临床病例

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摘要

下腔静脉发育不全是一种罕见的先天性血管异常，其解剖变异形式多种多样，有时甚至导致下腔静脉的中断。文献中对这些具体解剖变异的描述较少，因此该领域的研究仍面临挑战。本文报道了一例 75岁男性患者 的独特病例，其 无症状性下腔静脉肾下段发育不全 伴有奇静脉和半奇静脉肥大，以及 腹前壁侧支静脉网络 的形成。发现方式：该血管异常是在患者无相关症状的情况下，通过 多期对比增强计算机断层扫描（CT） 偶然发现的。影像学表现：患者 右侧腹腔静脉系统 显著异常；下腔静脉中断及奇静脉系统代偿性肥大；腹前壁形成了明显的 侧支静脉网络，提示血液回流路径的重组。临床观察：患者没有既往相关症状，观察到的解剖异常未与临床症状相关。本文强调 影像学检查，特别是 多期对比增强CT，在检测血管异常中的关键作用。在该病例中，影像学技术成功识别了复杂的血管异常，提供了清晰的解剖学细节。鉴于患者既往无症状，建议定期影像学随访，以监测异常血管结构的潜在进展及可能引发的并发症。

关键词：奇静脉肥大；下腔静脉；下腔静脉发育不全；侧支静脉；静脉系统。

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INTRODUCTION

Hypoplasia or the absence of the *inferior vena cava* (IVC) is a rare congenital condition that causes venous return from the lower body through the *azygos* or *hemiazygos* venous system [1,2]. The IVC may be hypoplastic or absent and is termed "interrupted" in this case: this term refers to total agenesis.

In most cases, this vascular anomaly is due to some embryologic mechanisms, and a failed anastomosis exists between the right subcardinal vein and the vitelline vein, resulting in hypoplasia or, in some cases, agenesis of the infrarenal/subrenal IVC and an interruption at the suprarenal venous segment [3].

In some cases, the newborn suprahepatic IVC could be missing or hypoplastic, resulting in direct outflow into the right atrium [4]. In this case, the small suprarenal IVC in the hepatic hilum drains through the *azygos* vein, while the hepatic IVC exclusively receives the hepatic veins. These anatomical variants are asymptomatic if the *azygos/hemiazygos* continuation is well-developed and the venous collateral loop is intact [5]. However, recurrent deep vein thrombosis of the lower limbs, leg swelling, leg pain, varices of the lower extremities, abdominal pain, and hematochezia in rare cases may present in the future [6,7]. Asymptomatic conditions are frequently discovered in the early to middle years of life, as in this case.

DESCRIPTION OF THE CASE

Anamnesis

A 75-year-old Caucasian man presented to the emergency department after a referred fall and underwent his first contrast-enhanced computed tomography (CT). A multiphase examination was performed using a 64-detector scanner, beginning with an unenhanced scan and progressing to postcontrast scans of the arterial and portal venous phases.

Diagnostic assessment and differential diagnosis

CT did not detect fractures, and no consequences visible under a radiological examination were observed. However, during imaging, the radiologist detected an unknown venous anomaly in the chest and abdomen.

The patient was not aware of this variant in the vascular anatomy and had never had symptoms related to it (Fig. 1).

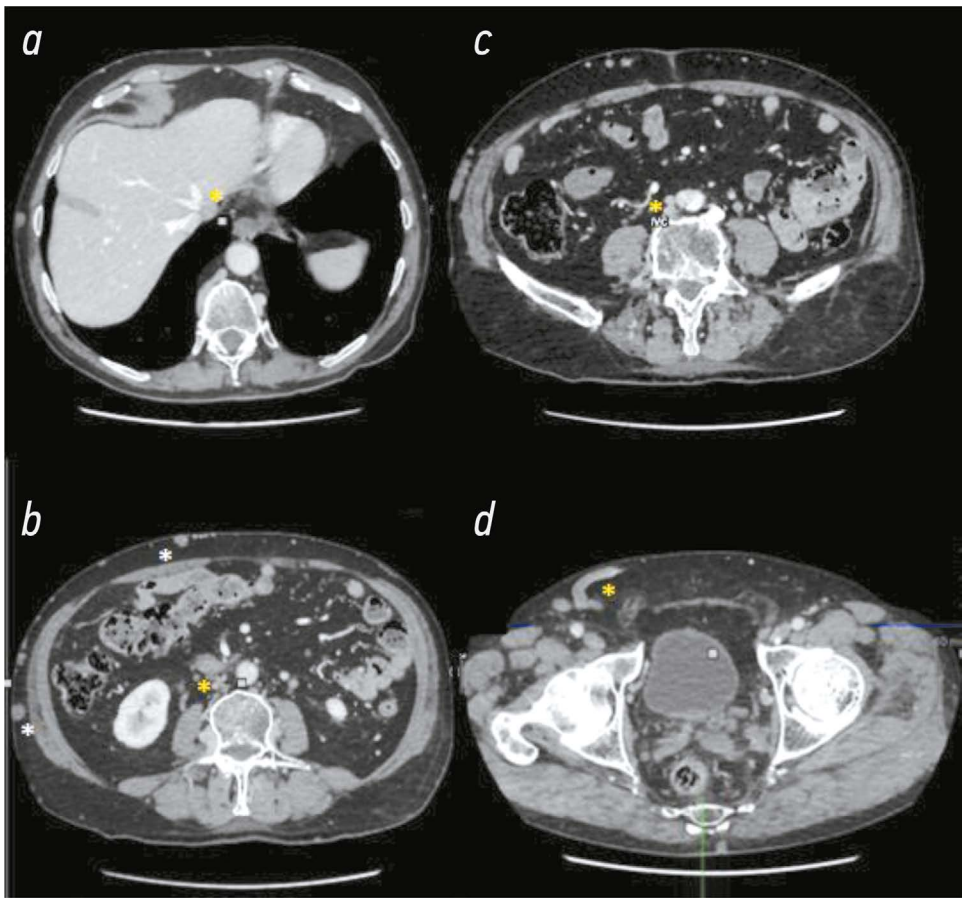

Fig. 1. Computed tomography images of the axial section of the abdomen (portal phase): *a*) The small suprarenal IVC is indicated by a yellow asterisk. *b*) Lower in the abdomen, the yellow asterisk indicates the hypoplastic IVC, and the hypertrophic collateral venous circles in the anterior abdomen wall on the right are indicated by a white asterisk. *c*) Another lower axial section showing the small IVC (yellow asterisk). *d*) The drainage on the right side is accomplished by a constant iliac vessel (yellow asterisk).

Interventions

To adequately study this vascular anomaly, postprocessing reconstruction was conducted on all planes (axial, coronal, and sagittal) using the MIP program, and 3D images were generated.

At first glance, the most evident imaging finding of the vascular anomaly was the presence of multiple collateral venous circles on the anterior wall of the abdomen, particularly on the right side, and the IVC under the kidneys was hypoplastic (Fig. 2).

Certain distended azygos and hemiazygos veins received blood from the abdomen. The *azygos* vein connected with the *superior vena cava* (SVC) through its arch; however, its dimensions were abnormal. It began from D7 and extended to D10–11, from the confluence of the right renal vein, transhepatic vein, and an aberrant vein (Fig. 3 and Fig. 4).

Follow-up and outcomes

The patient had previously never experienced symptoms that could be correlated with the same vascular abnormality, and the symptoms he experienced did not appear to be related. Therefore, periodic follow-up was recommended.

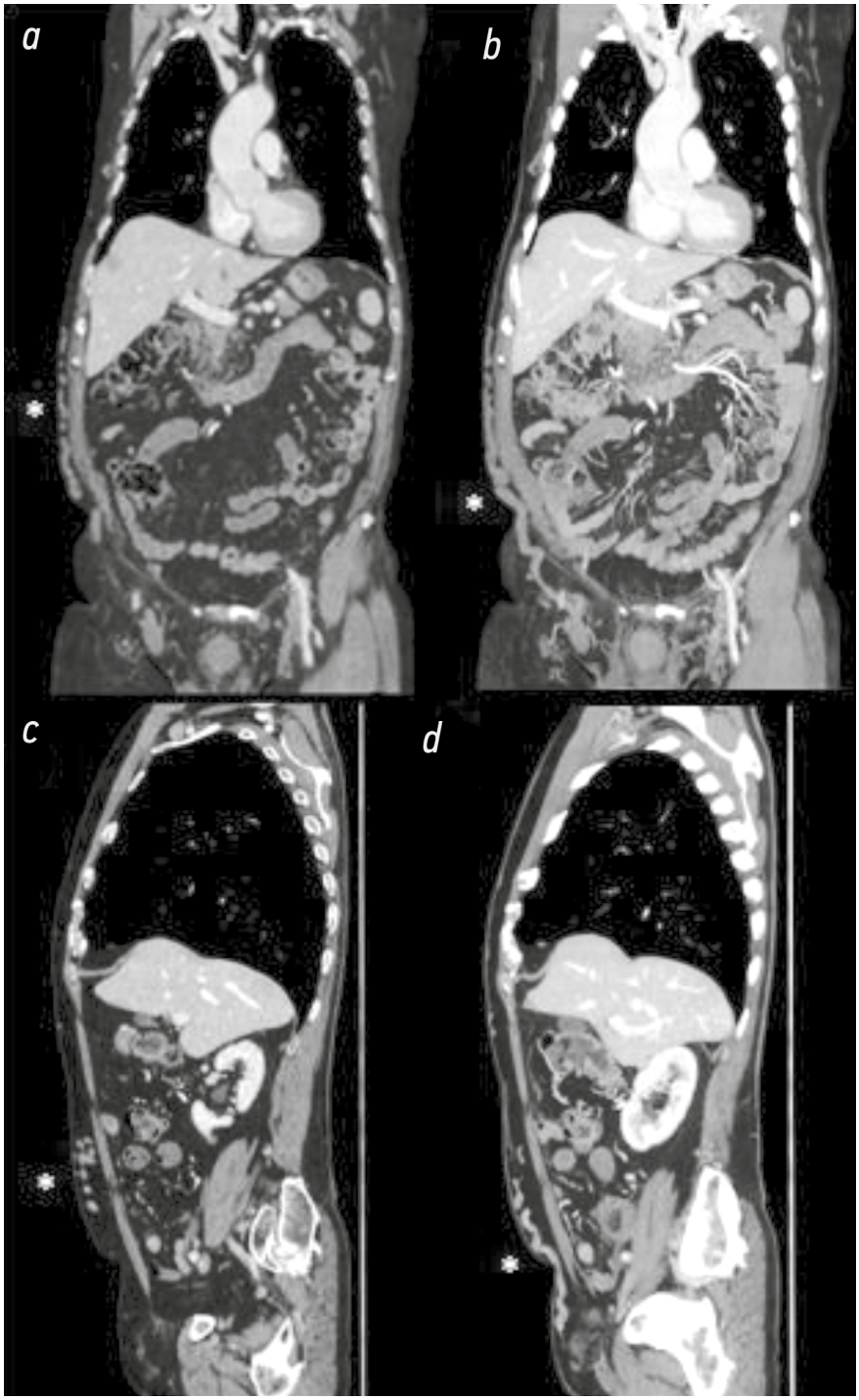

Fig. 2. Computed tomography images of the coronal (up) and sagittal (down) sections in the portal phase of the chest and abdomen: *a*) Right abdominal wall: consistent venous collateral circles are visible (white asterisk). *b*) The same image with a rising MIP value indicates venous collateral rings, particularly on the patient's right side. *c*) and *d*) Hypertrophic venous collateral circles in the sagittal section at different levels.

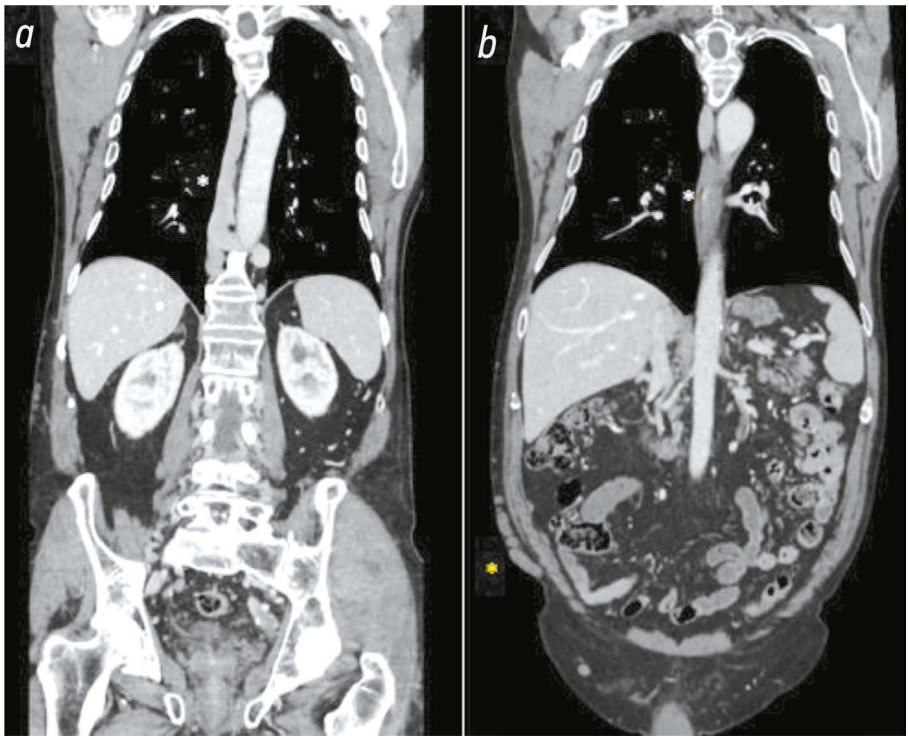

Fig. 3. Computed tomography images of the coronal section and portal phase of the chest and abdomen: *a*) The white asterisk indicates the confluence of the giant azygos, hemiazygos, and an aberrant vein. *b*) Same image at different sections. The yellow asterisk on the right side at the level of the anterior abdominal wall indicates marked collateral vein circles.

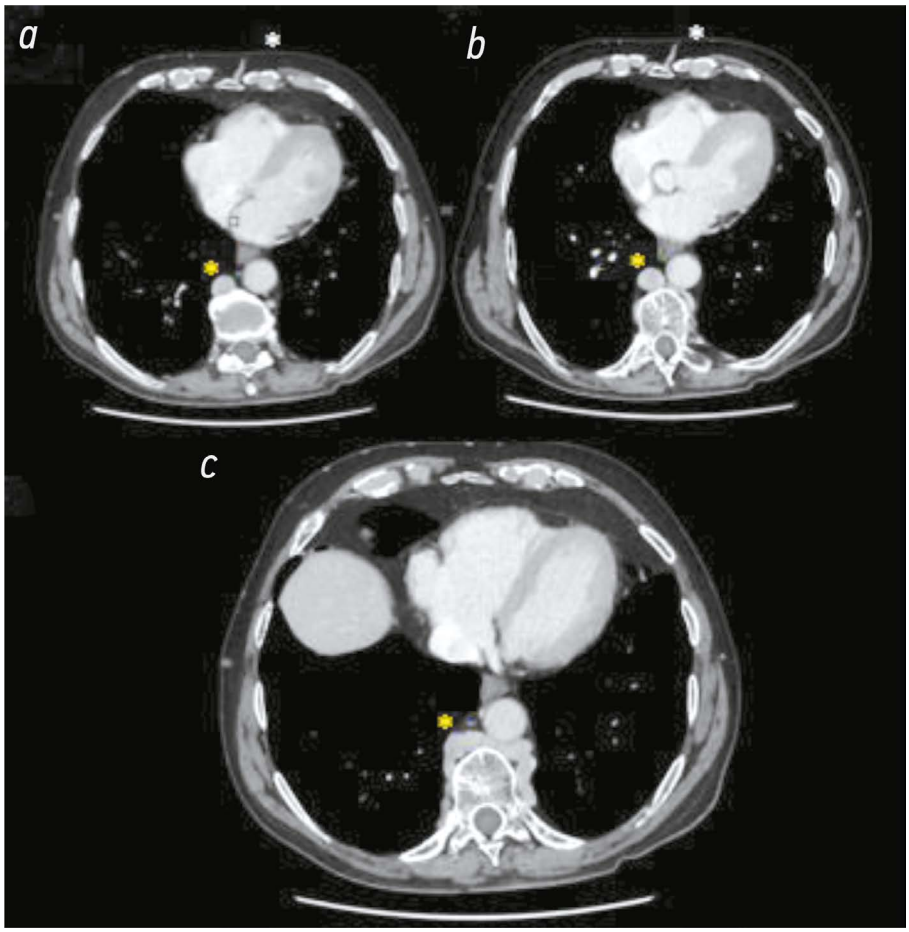

Fig. 4. Computed tomography images of the axial section of the chest: the confluence with the hypertrophic azygos and hemiazygos veins is seen in all images at different levels (from upper to lower). In *a*) and *b*), the white asterisk marks the confluence of the collateral circles of the abdomen and chest.

DISCUSSION

The IVC is a large retroperitoneal vein that transports deoxygenated blood from the lower extremities, pelvis, and abdomen to the right atrium. The *azygos* venous system is a paravertebral connection in the posterior thorax that connects the SVC to the IVC. The *azygos*, *hemiazygos*, accessory *hemiazygos*, and left superior intercostal veins form a H-shaped pattern, indicating the *azygos* venous system [2,3]. IVC hypoplasia, characterized by an *azygos/hemiazygos* system and collateral circle compensation, is an uncommon vascular defect with numerous variations [4-8]. The anomaly is primarily caused by the abnormal regression or persistence of embryological veins (anterocardinal, postcardinal, subcardinal, supracardinal, and vitelline) that form the five embryological segments of the final structure of the IVC, namely, iliac, subrenal, renal, suprarenal, and hepatic, including suprahepatic and retrohepatic [9]. In this case, the patient had a very small IVC in the right iliac fossa, with larger *azygos/hemiazygos* veins, indicating an expanded *hemiazygos* system as the primary drainage system.

Imaging plays a critical role in the detection of this type of vascular abnormality. Herein, CT helped differentiate all vessels, discover variants, and increase the *azygos* system. The imaging options for studying the vascular abnormality include echocardiographic techniques and color Doppler.

Imaging, CT angiography, and IVC angiography can detect hypoplastic/interrupted IVC, identify abnormal vessels, and assess *azygos* system dilatation due to increased flow. Angiography is useful for determining the precise anatomy of vessel drainage for surgical purposes. Detecting venous abnormalities is crucial because they can interfere with right heart catheterization, cardiopulmonary bypass surgery, and pacemaker insertion. IVC hypoplasia, or interrupted IVC in extreme cases, also known as *azygos–hemiazygos* continuation, is a benign disorder that does not require treatment owing to adequate vascularization [10-11].

However, patient knowledge is critical in the event of surgical intervention [9-14]. A misdiagnosis mayoccurr because of possible mediastinal shadow enlargement on chest X-ray images or dilated azygos or hemiazygos vein adjacent to the descending aorta on transesophageal echocardiography mimicking aortic pathology [5,10,12].

Interrupted IVC is often linked to other congenital defects, particularly in the cardiac region, prompting a search for related conditions. The presence of other pathologies must be evaluated. Excluding portosystemic shunting is crucial for management because chronic congenital portosystemic shunts can lead to serious complications. The presented case did not correlate with other congenital defects or the patient's malignancy. Both situations can be regarded as independent.

CONCLUSION

This report presents an uncommon venous abnormality in the chest and abdomen of an asymptomatic adult with hypoplastic IVC accompanied by *azygos/hemiazygos* hypertropia and the presence of numerous collateral venous circles. This case highlights the importance of imaging in the detection of complex vascular abnormalities. Physicians should carefully examine this unique vascular abnormality to prevent misdiagnosis and improve surgical outcomes.

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