

# Radiological features of azygos and hemiazygos continuation of inferior vena cava

## A case report

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### Abstract

**Rationale:** Azygos and hemiazygos continuation of the inferior vena cava (IVC) is uncommon. It is rare especially when it is not associated with congenital heart disease or deep venous thrombosis.

**Patient concerns:** We report an interesting case of an interstitial lung disease with an interrupted IVC with azygos and hemiazygos continuation. A 67-year-old man suffered from cough and shortness of breath.

**Diagnoses:** Computed tomography revealed absence of the hepatic segment of the IVC with azygos and hemiazygos continuation. The patient did not have congenital anomalies of the remaining thoracoabdominal vasculature and viscera. The diagnosis of azygos and hemiazygos continuation of IVC was made by inferior venacavography.

**Interventions:** There was no significant association between interstitial lung disease and expanded azygos and hemiazygos veins. The patient was treated with corticosteroids.

**Outcomes:** After 6 months of follow-up, the patient was asymptomatic.

**Lessons:** It is important to recognize the enlarged azygos vein at the confluence with the superior vena cava and in the retrocrural space to avoid misdiagnosis as a right-sided paratracheal mass or retrocrural adenopathy.

**Abbreviations:** DVT = deep venous thrombosis, IVC = inferior vena cava, SVC = superior vena cava.

**Keywords:** azygos and hemiazygos continuation, inferior vena cava, inferior venacavography

## 1. Introduction

Azygos and hemiazygos continuation of the inferior vena cava (IVC) has also been termed infrahepatic interruption of IVC with azygos and hemiazygos continuation. The absence of the intrahepatic segment of IVC with azygos and hemiazygos continuation occurs in 0.6% of patients investigated for congenital heart disease and frequently coincides with situs inversus, asplenia, or polysplenism, persistent left superior vena cava (SVC), and congenital pulmonary venolobar syndrome.<sup>[1,2]</sup> Azygos and hemiazygos continuation of the IVC is rare especially when it is not associated with congenital heart disease.<sup>[3]</sup> Here we report a case of an interstitial lung disease with an interrupted IVC with azygos and hemiazygos continuation, which have not

been previously reported. The angiographic features and clinical importance of this condition are discussed. The study was approved by the ethics committee of Nanjing Drum Tower Hospital. Written informed consent was obtained from the patient for the publication of this manuscript and accompanying images.

## 2. Case report

A 67-year-old man was referred to our hospital in May 2017 with complaints of cough and shortness of breath. He had no prior history of coronary artery disease, congestive heart failure, hypertension, or diabetes. He had a history of colon cancer resection. No history of deep venous thrombosis (DVT) was reported. Clinical examination found a regular pulse rate of 86 beats/min of normal volume and character, and blood pressure 128/61 mm Hg and oxygen saturation on room air, 95%. A physical examination revealed fine crackles in both lower lung fields. The remainder of his physical examination was unremarkable.

Routine laboratory findings were normal. Brain natriuretic peptide and D-dimer level were within the normal range. A chest CT scan revealed peripheral, subpleural peribronchovascular opacities, and consolidations without honeycomb changes in bilateral lungs (Fig. 1A). Initial imaging showed dilatation of the azygos veins and hemiazygos veins azygos (Fig. 1B–D). Color Doppler ultrasound showed the inferior vena cava from the left renal vein to second hepatic portal was occlusive. Color Doppler echocardiography suggested normal function of both ventricles, the pulmonary artery pressure was presumed to be about 38 mm Hg. We then performed pulmonary function tests and found a forced vital capacity (FVC) of 2.19 L (66.7% predicted), forced

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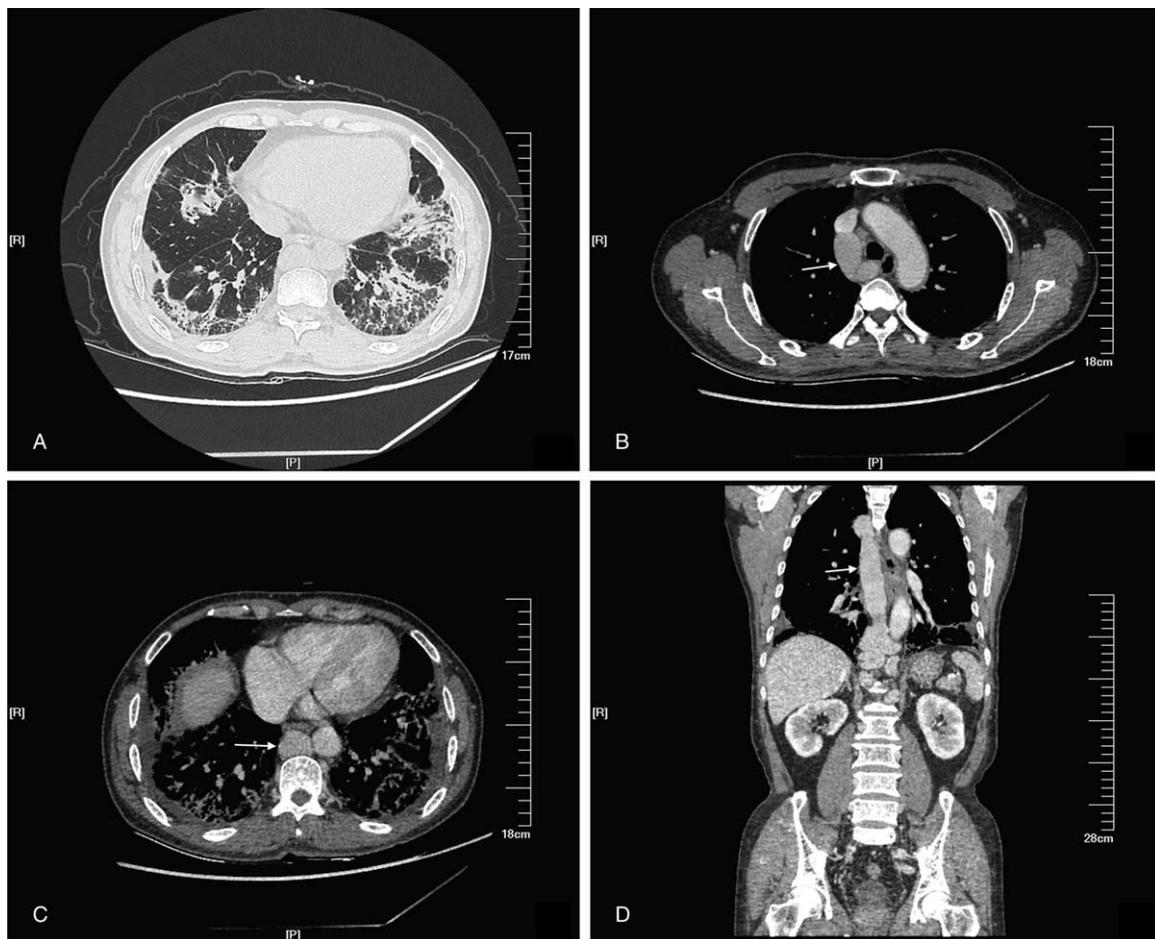
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**Figure 1.** The high-resolution computed tomography. (A) Chest CT showed peripheral, subpleural reticular opacities in bilateral lungs. (B)–(D) CT angiography images showed a dilated azygos vein accompanied by continuation of interrupted IVC (arrows). IVC=inferior vena cava.

expiratory volume in 1 seconds (FEV1) of 1.59 L (61.7% predicted), and an FEV1/FVC ratio of 65.8%. A venous duplex scan of both legs did not reveal deep venous thrombosis, deep, or superficial venous incompetence. A CT scan of chest without contrast showed a possible right-sided paratracheal mass or retrocrural adenopathy. CT angiography of the thorax and abdomen was performed for further evaluation of congenital vascular defects. The IVC was found obstructed with multiple collateral circulation formation from the level of the left renal vein to second hepatic portal.

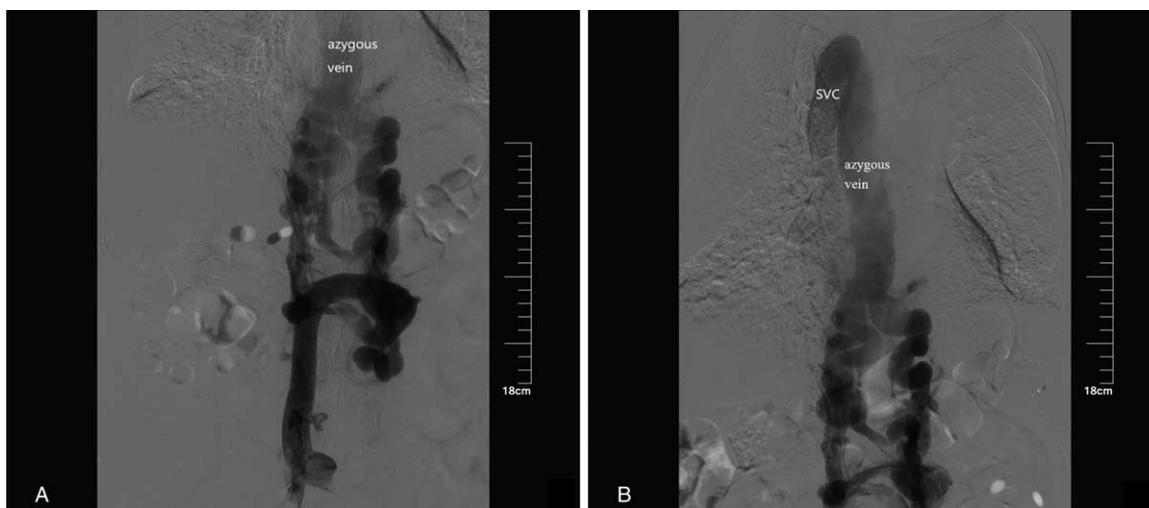
IVC angiography revealed complete absence of IVC with azygos and hemiazygos continuation and collateral circulation between vertebral venous plexus and azygos system (Fig. 2A and B). Since the collateral circulation was well constructed and the edema of lower extremities didn't occur, he didn't need special treatments for occluded IVC. Because there was no significant association between interstitial lung disease and expanded azygos veins and hemiazygos veins, he was given corticosteroids therapy.

### 3. Discussion

In normal anatomy, the azygos vein drains blood from the oesophageal, mediastinal, intercostal, pericardial, and bronchial veins.<sup>[2]</sup> A normal azygos vein is so small that it cannot be

detected by chest radiography. Collateral circulation or fistula formation or increased right atrial pressure will result in enlarged azygos vein by increased venous blood flow.<sup>[4]</sup> The most common cause of such azygos vein enlargement is azygos continuation of the interrupted IVC.<sup>[4,5]</sup>

During embryogenesis, the normal IVC is made up of 4 segments including the hepatic, pre renal, renal, and post renal segments. The IVC collects the blood from the lower limbs, pelvic area, and abdomen.<sup>[6,7]</sup> Various anomalies of the IVC can be seen depending on abnormal regression or abnormal persistence of embryonic veins. Infrahepatic interruption of the IVC with azygos and hemiazygos continuation is a rare finding especially when it is not associated with congenital heart disease. IVC interruption occurs in 0.3% of otherwise healthy individuals and on 0.6% of patients with other cardiovascular defects.<sup>[8,9]</sup> IVC Interruption most likely stems from agenesis of a segment of the IVC, or a failure of fusion between the suprarenal and hepatic segment of the IVC.<sup>[3]</sup> The embryonic event is theorized to be failure to form the right subcardinal-hepatic anastomosis, with resulting atrophy of the right subcardinal vein. This development anomaly results in termination of the IVC below the hepatic vein. Consequently, systemic venous flow beyond this point is accommodated by the enlarged azygos and hemiazygos veins. Eventually, it will empty into the SVC through a dilated azygos arch.<sup>[3,10]</sup>



**Figure 2.** IVC angiography. (A) and (B) IVC angiography confirmed the total interruption of IVC below the hepatic vein with azygos vein continuation, draining into the SVC. IVC=inferior vena cava, SVC=superior vena cava.

IVC abnormalities have been associated with congenital heart anomalies, polysplenia syndrome, and situs anomalies associated with asplenia.<sup>[9,10]</sup> There is a slight preponderance in males.<sup>[11,12]</sup> This anomaly is usually associated with recurrent DVT of the lower limbs or with sick sinus syndrome and it is typically detected in the early to middle ages of life.<sup>[11,13]</sup> Patients with interruption of the IVC may be present with a spectrum of clinical signs and symptoms, such as leg swelling, leg pain, varices of lower extremities, abdominal pain, and rarely hemochezia.<sup>[8,14]</sup> If well-developed azygos and hemiazygos continuation is present, this patient presumably will be asymptomatic.

Noninvasive imaging modalities such as contrast-enhanced CT and magnetic resonance imaging are the most reliable methods for identification of these anomalies in an asymptomatic patient.<sup>[15]</sup> Although vascular structures can usually be readily identified on contrast-enhanced CT scans, identification of unusual venous arrangements may be difficult. In such patients, IVC angiography allows for a complete description of these abnormalities and displays a complete picture of the IVC system.

The patient did not have congenital anomalies of the remaining thoracoabdominal vasculature and viscera. There was no significant association between interstitial lung disease and expanded azygos and hemiazygos veins. No evidence confirms the correlations between the 2 conditions. They are 2 independent diseases and do not correlate with each other. After corticosteroids treatment, opacities and consolidations in bilateral lungs were diminished. The patient was continued follow-up at present.

The major limitations of our study include its retrospective nature, only one case, and the short follow-up period. But rarity of the interruption or stenosis of the IVC makes it very hard to conduct studies with a larger patient population.

In conclusion, it is important to recognize the enlarged azygos vein at the confluence with the SVC and in the retrocrural space to avoid misdiagnosis as a right-sided paratracheal mass or retrocrural adenopathy. Contrast-enhanced CT scan and venography can be helpful. The recognition of this venous anomaly is important for the pulmonologist, especially for

conditions such as a paracardiac or mediastinal mass on chest radiography.

#### Author contributions

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**Writing – review & editing:** Yin Liu, Hourong Cai.

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