

## Azygos Continuation of the Inferior Vena Cava: Potential for Misdiagnosis as Lung Neoplasm

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### ABSTRACT

**Background:** The azygos venous system is an accessory venous pathway supplying an important collateral circulation between the superior and inferior vena cava (IVC).

**Case summary:** We report a case of complicated community-acquired pneumonia with interruption of the IVC with azygos continuation that was misdiagnosed as lung neoplasm.

**Discussion:** The aim of this case report is to emphasize the importance of recognizing an enlarged azygos vein at the confluence with the superior vena cava and in the retrocrural space to avoid misdiagnosis as a right-sided para-tracheal mass. The angiographic features and clinical importance of this condition are discussed.

### LEARNING POINTS

- An enlarged azygos vein at the confluence with the superior vena cava and in the retrocrural space can be misdiagnosed as a right-sided para-tracheal mass/neoplasm or retrocrural adenopathy.
- Biopsy is the most important tool to confirm the diagnosis of malignancy but could be catastrophic in this situation.
- Re-imaging after treatment for infection and pneumonia could potentially avoid this error.

### KEYWORDS

Azygos continuation, inferior vena cava, lung neoplasm, pneumonia

### CASE DESCRIPTION

A 57-year-old man presented with a 5-day history of right-sided pleuritic chest pain and breathlessness on exertion associated with fever and rigors. He was a smoker with a 20 pack-year history and no prior history of ischaemic heart disease, congestive heart failure, hypertension or diabetes. Physical examination revealed decreased air entry and crepitations at the right lung base. The remainder of the physical examination was unremarkable.

### Investigation

Routine laboratory findings revealed C-reactive protein (CRP) of 236 mg/l, an elevated white cell count of  $13.2 \times 10^9/l$  with neutrophilia (neutrophil count  $10 \times 10^9/l$ ), and a raised D-dimer level. Arterial blood gas (ABG) analysis showed hypoxia with normal pH, low  $\text{PaO}_2$  of 8.0 kPa, and  $\text{PaCO}_2$  of 5.1 kPa. The electrocardiogram (ECG) demonstrated sinus tachycardia with no ST-T changes.

Chest x-ray (Fig. 1) confirmed an area of consolidation at the right base with blunting of the right costophrenic angle due to pleural effusion.

The patient was commenced on treatment for pneumonia with antibiotics and prophylactic anticoagulation. However, despite antibiotic treatment, there was no change in his oxygen requirement and symptoms, and so computed tomography pulmonary angiography (CTPA) was performed, which demonstrated right segmental pulmonary emboli. CT imaging raised concern of possible neoplastic infiltration at the level of the inferior right hilum para-tracheal region with coalesced lymph nodes with aorto-pulmonary and sub-carinal lymphadenopathy and pleural infiltration with pleural effusion and right base opacity (Fig. 2). CT of the abdomen-pelvis and CT of the brain excluded any evidence of metastatic spread or significant other lymphadenopathy.



Figure 1. Chest x-ray showing right lower lobe opacity with pleural effusion

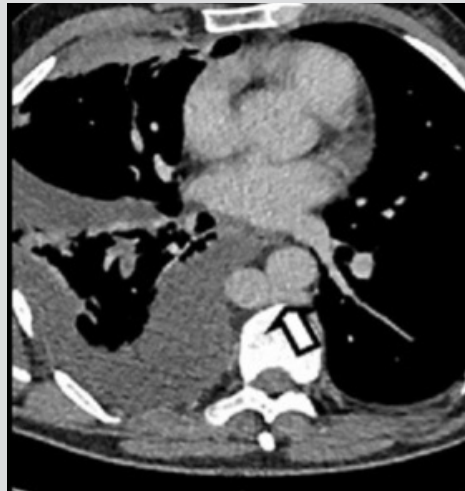


Figure 2. Axial contrast-enhanced CT of the abdomen showing a complex right-sided pleural effusion and incidental hemiazygos vein emptying into the azygous continuation of the inferior vena cava (open arrow) in the retroaortic space

### Treatment

The patient was treated for community-acquired pneumonia with para-pneumonic effusion and pulmonary embolism with antibiotics according to national guidelines. This treatment consisted of ceftriaxone 2 g IV once daily and doxycycline 100 mg orally twice daily for 10 days, therapeutic anticoagulation (innohep 175 IU/kg subcutaneously once daily), supplemental oxygen and chest physiotherapy. The patient improved clinically on treatment and was referred to a rapid access lung cancer clinic (RALC) for follow-up. He was reviewed at RALC and the initial impression was of likely metastatic lung cancer with a poor prognosis based on x-ray and CT findings. The possible diagnosis of malignancy was conveyed to the patient at the clinic. A positron emission tomography (PET) CT scan and trans-bronchial biopsy was planned to obtain a tissue sample. The diagnosis of possible malignancy had a significant impact on the patient's mood.

The case was discussed at an oncology multidisciplinary meeting (MDM) at RALC. It was felt that the soft tissue mass on the right para-tracheal region believed to be a neoplasm was an azygos continuation of the inferior vena cava (IVC), a normal vascular variant. The presentation was complicated with pneumonia, effusion and segmental pulmonary embolism. The biopsy and PET scan were cancelled and a follow-up CT of the thorax was recommended in 12 weeks' time for further evaluation. The patient was very relieved by the new developments.

### Follow-up

CT of the thorax showed resolution of pleural effusions, a reduction in the size of the aorto-pulmonary and sub-carinal lymphadenopathy, and azygos continuation of the IVC (Figs. 3–5). The patient is currently doing well, has stopped smoking and will be followed up in the outpatient clinic with a repeat scan in a year's time.

### DISCUSSION

The azygos, hemiazygos and accessory hemiazygos veins originate from the last portion of the posterior cardinal veins at the junction of the right ascending lumbar and subcostal veins. The azygos system is a paired paravertebral venous pathway in the posterior thorax. It ascends along the anterolateral surface of the thoracic vertebrae and arches ventrally to the right main bronchus at T5–T6, draining into the superior vena cava (SVC). More rarely, the azygos vein drains into the right brachiocephalic vein, right subclavian vein, intra-pericardial SVC or right atrium<sup>[1]</sup>. The prevalence of azygos continuation is about 0.6%<sup>[2]</sup>. During embryogenesis, the normal IVC is made up of four segments including the hepatic, prerenal, renal and post-renal segments. The IVC collects the blood from the lower limbs, pelvic area and abdomen<sup>[3,4]</sup>. Failure of the union between the hepatic and prerenal segments during embryological development results in the so-called 'infrahepatic interruption of the inferior vena cava (IVC) with azygos continuation' (Fig. 4).

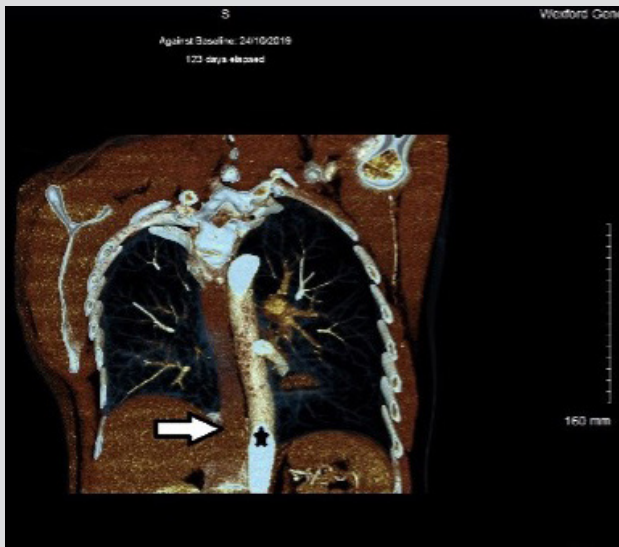


Figure 3. Coronal oblique thick section (100 mm slab) volumetric rendered technique (VRT) thin section reconstruction showing contrast opacifying the descending thoracic aorta (asterisk) and poor contrast enhancement of the azygos continuation of the inferior vena cava (open arrow)



Figure 4. Axial arterial phase contrast-enhanced CT of the thorax showing un-opacified venous blood flowing into the superior vena cava from the azygos continuation of the inferior vena cava (open arrow)

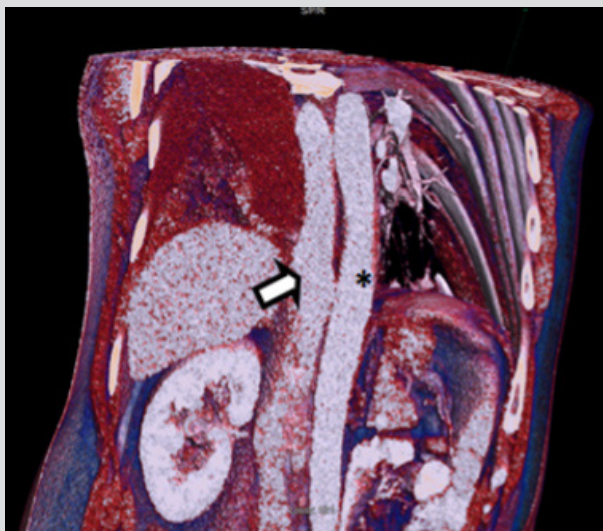


Figure 5. Coronal oblique thick section (100 mm slab) volumetric rendered technique (VRT) thin section reconstruction showing contrast opacifying the descending thoracic aorta (asterisk) and azygos continuation of the inferior vena cava (open arrow)

IVC interruption occurs in 0.3% of otherwise healthy individuals and in 0.6% of patients with other cardiovascular defects [5, 6]. Azygos continuation can be observed in association with severe congenital heart disease, asplenia or polysplenia syndrome [6]. The patients are usually asymptomatic and the anomaly is detected incidentally. The most frequent congenital abnormalities include an azygos fissure, absence of the azygos vein, aortic nipple, azygos continuation of the IVC, and partial venous return [7]. The most common cause of such azygos vein enlargement is azygos continuation of the interrupted IVC [8].

Patients with interruption of the IVC may present with a spectrum of clinical signs and symptoms, such as leg swelling, leg pain, varices of lower extremities, abdominal pain, and rarely haematochezia [9, 10]. Diagnosis of this vascular anomaly is also important before cardiac catheterisation, especially in case of interventional procedures such as balloon dilatation, stent implantation or umbrella placement, and certain surgical procedures such as ligation of the azygos vein during thoracotomy or portocaval decompression surgery [11, 12].

## Diagnosis

On chest x-ray, azygos continuation can be suspected in the presence of a focal enlargement of the right paratracheal stripe above the right main stem bronchus<sup>[6]</sup>. Non-invasive 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>[13]</sup>. A CT scan confirms the mild enlargement of the azygos vein and azygos arch secondary to increased flow. Moreover, in azygos vein continuation the infrahepatic portion of the IVC is absent<sup>[6]</sup>. Although vascular structures can usually be readily identified on contrast-enhanced CT scans, some cases require IVC angiography for a complete description of the IVC system<sup>[14]</sup>.

Our patient did not have congenital anomalies of the remaining thoracic-abdominal vasculature and viscera, and was asymptomatic with the anomaly. There was no significant association between pneumonia and expanded azygos and hemiazygos veins. After antibiotic treatment consolidation, effusion and lymphadenopathy in the right lung resolved.

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