A 35-year-old Asian male cab driver presented with a 1-week history of increasing swelling of the left lower limb. Findings on examination of the left leg were suggestive of proximal deep vein thrombosis. Ultrasonography of the deep veins of the leg and computed tomographic (CT) scan of the lower abdomen revealed the presence of thrombosis in the left common femoral vein and in the left internal and external iliac veins. The patient was administered intravenous heparin for anticoagulation followed by oral warfarin.

A chest radiograph showed a well-defined rounded right paratracheal mass (Fig 1). A CT scan of the chest at the level of the aortic arch (Fig 2) revealed a massively dilated azygos arch. A CT scan at the level of the diaphragmatic crura (Fig 3) showed two retrocrural densities on either side of the descending aorta accounted for by a dilated azygos vein on the right and a more dilated hemiazygos vein on the left. No definable inferior vena cava was seen above the renal veins. Otherwise, both the chest and abdominal CT scans were unremarkable. An echocardiogram was within normal limits.
Diagnosis: Infrahepatic interruption of the inferior vena cava with azygos continuation

Infrahepatic interruption of the inferior vena cava with azygos continuation is a rare congenital anomaly seen in 0.6 to 2% of patients with congenital heart disease and in less than 0.3% of otherwise normal individuals. This developmental anomaly results in termination of the inferior vena cava below the hepatic vein, and systemic venous flow beyond this point is accommodated by the dilated azygos and hemiazygos veins, which eventually empty into the superior vena cava via a dilated azygos arch. This dilated azygos arch accounts for the right paratracheal mass seen on the chest radiography in Figure 1. Other causes of a dilated azygos venous system include right heart failure, superior vena caval obstruction, inferior vena caval obstruction, pregnancy, and portal hypertension.

The diagnosis of azygos continuation of the inferior vena cava can be made by the demonstration of a constellation of findings on a chest CT scan. They include enlargement of the azygos arch, enlargement of the paraspinal portions of the hemiazygos and azygos veins, confluence of the hemiazygos and azygos veins in the thorax, and the enlargement of the retrocrural portion of the azygos and hemiazygos veins in the absence of a definable inferior vena cava.

Embryogenesis of the inferior vena cava, azygos, and hemiazygos veins involves the subcardinal and supracardinal venous system in the retroperitoneum. Developmentally, the inferior vena cava is made up of the hepatic, prerenal, renal, and postrenal segments, which by vessel fusion, regression and midline anastomosis forms the adult inferior vena cava. The hepatic segment is formed from the hepatic vein and hepatic sinusoids. The prerenal segment is formed from the right subcardinal vein, the renal segment from the anastomosis of the subcardinal and supracardinal veins, and the post renal segment from the subcardinal vein. The azygos and hemiazygos veins are derived for the anterior portion of the supracardinal veins and receives blood from the ascending lumbar veins. Failure of fusion of the hepatic and prerenal segments is the most common developmental anomaly of the inferior vena cava and results in caval interruption below the hepatic vein. The hepatic segment of the inferior vena cava then drains directly into the right atrium while the inferior vena cava below the level of the renal veins remains unaffected.

Known associated cardiac anomalies include atrial septal defect, pulmonic stenosis or atresia, double outflow right ventricle, atioventricular canal, corbilocular, and anomalous pulmonary venous drainage. Also documented is the presence of mirror-image dextrocardia, dextroversion, situs inversus, partial inversion of the abdominal viscera, bilateral superior vena cava, and asplenia or polysplenia.

When this anomaly of the inferior vena cava occurs as an isolated finding, it can lead to misdiagnosis. On chest radiograph, the dilated azygos arch may be misinterpreted as a right paratracheal noplasm, while the dilated azygos and hemiazygos veins in the paravertebral location may be mistaken for a posterior mediastinal neoplasm. On CT scanning, these dilated veins in the retrocrural position mimic retrocrural adenopathy.

In right-heart catheterization via the femoral vein, the presence of this anomaly can lead to confusion as the catheter follows a reverse “U” course through the azygos vein and superior vena cava into the right atrium. The use of a venous filter in the treatment of complicated thromboembolic disease and the surgical creation of portosystemic shunts in the treatment of portal hypertension may not be possible in the presence of this anomaly. Accidental ligation of the azygos vein leading to a lethal outcome has been reported.

The recognition of this congenital anomaly is therefore important, and contrast CT scanning is the preferred method in making the diagnosis.

REFERENCES