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Adult Scimitar Syndrome in a Woman Followed for Asthma: CT Angiography Findings and Four-Year Imaging Stability

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Abstract **Case Report**

Scimitar syndrome is a rare congenital cardiopulmonary anomaly. Part or all of the right lung drains through an anomalous vein into the inferior vena cava (IVC), usually with right lung hypoplasia and cardiac dextroposition. We report an asymptomatic 47-year-old woman with well-controlled asthma, whose scimitar syndrome had been diagnosed on CT in 2022. Transthoracic echocardiography had not identified any anomalous pulmonary venous return. CT angiography was performed for routine follow-up. It showed an anomalous vein draining the anterior basal segment of the right lower lobe. The vein coursed along the right hemidiaphragm and joined the IVC just below the cavoatrial junction. CT also showed right lung hypoplasia, cardiac dextroposition and elevation of the right hemidiaphragm. No anomalous systemic arterial supply, CT sign of pulmonary hypertension or right ventricular enlargement was identified. The findings were unchanged from a CT performed four years earlier. This case illustrates the mild adult form of the syndrome and the limits of echocardiography in adults. It also shows how CT angiography provides the anatomical information needed to choose between conservative follow-up and surgical repair.

Keywords: Scimitar Syndrome, Partial Anomalous Pulmonary Venous Return, Congenital Pulmonary Venolobar Syndrome, CT Angiography, Pulmonary Hypoplasia, Adult Congenital Heart Disease.

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INTRODUCTION

Scimitar syndrome is a partial or, less often, total anomalous pulmonary venous connection of the right lung to the IVC. It is usually associated with right lung hypoplasia, cardiac dextroposition, hypoplasia of the right pulmonary artery and, in some patients, anomalous systemic arterial supply to the right lower lobe [1, 2]. It belongs to the spectrum of the congenital pulmonary venolobar syndrome, also called hypogenetic lung syndrome, which reflects an early developmental disorder of the whole right lung bud [1, 2].

The syndrome has a bimodal presentation [2–4]. The infantile form is often associated with other congenital heart defects and pulmonary hypertension, and its mortality is high. The childhood and adult form is usually mild and is often discovered incidentally. In the largest multicentre European registry (485 patients), survival at 30 years was 88% and was lower in patients with associated congenital heart disease or pulmonary hypertension [3]. The best management of the asymptomatic adult remains debated, because surgical repair carries a substantial risk of postoperative stenosis of the scimitar drainage [3-6].

Cross-sectional imaging is central to this decision. We report a case of adult scimitar syndrome in a woman followed for asthma, in whom echocardiography had not shown the anomalous venous return. We describe the CT angiography findings, their stability over four years and the information the radiologist should provide to the clinical team.

CASE REPORT

A 47-year-old woman with bronchial asthma since 2010, well controlled on treatment, was followed in the cardiovascular surgery department for scimitar syndrome diagnosed on chest CT in 2022. She was asymptomatic, and physical examination was unremarkable. A previous transthoracic echocardiogram had not identified any anomalous pulmonary venous return; its full report was not available. No cardiac MRI or cardiac catheterisation had been performed. Chest CT angiography was performed as part of routine follow-up.

CT Technique

The examination was performed on a SOMATOM Definition AS+ scanner (Siemens Healthineers). It included an unenhanced acquisition followed by three contrast-enhanced acquisitions at 100 kVp, timed for the pulmonary arterial, systemic arterial and delayed phases. Images were reconstructed with a 1-mm slice thickness. Multiplanar reformations and maximum-intensity projections (MIP) were generated. A total of 100 mL of iodinated contrast medium was injected at 4 mL/s.

CT Findings

An anomalous vein drained the anterior basal segment of the right lower lobe. It coursed inferomedially along the superior surface of the right hemidiaphragm and joined the IVC just below the cavoatrial junction, above the diaphragm (Fig. 1). Its maximal diameter was 10 mm, with no stenosis at its caval termination. The other right pulmonary veins joined a common trunk that drained normally into the left atrium, so the anomalous drainage was partial.

The right hemithorax was reduced in volume, with right lung hypoplasia, a rightward shift of the heart (dextroposition) and mild elevation of the right hemidiaphragm (Fig. 2). Linear atelectasis was present in the anterior basal segments of both lower lobes. There was no pulmonary nodule, mediastinal lymphadenopathy, pleural effusion or pericardial effusion.

The main, right and left pulmonary arteries were normal in calibre, without endoluminal thrombus. The heart was not enlarged. No anomalous systemic artery arising from the descending thoracic or upper abdominal aorta and supplying the right lower lobe was identified (Fig. 4). The main pulmonary artery measured 24 mm, and the ratio of its diameter to that of the ascending aorta was about 0.9. The right-to-left ventricular diameter ratio was 0.89.

Comparison

Compared with a CT angiography performed in July 2022, the course, calibre and termination of the scimitar vein, as well as the associated parenchymal findings, were unchanged (Fig. 3).

Management

The patient was asymptomatic, had no CT sign of pulmonary hypertension or right ventricular enlargement, and her findings were stable. The multidisciplinary team therefore chose conservative clinical and imaging follow-up.

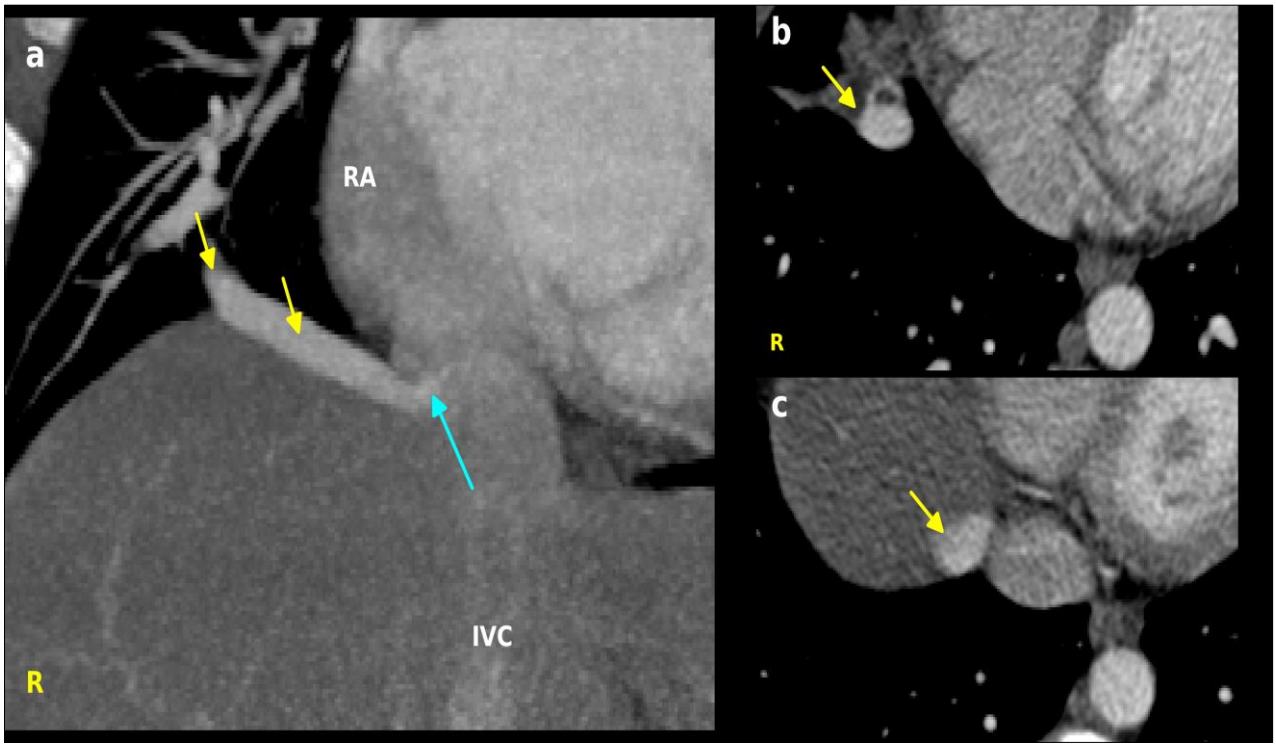


Figure 1: Contrast-enhanced chest CT angiography in a 47-year-old woman with scimitar syndrome. (a) Coronal 10-mm maximum-intensity projection (MIP) shows an anomalous vein (scimitar vein, yellow arrows). It drains the anterior basal segment of the right lower lobe, courses inferomedially along the right hemidiaphragm and joins the inferior vena cava (IVC) just below the cavoatrial junction (cyan arrow). RA, right atrium. (b, c) Axial images at the level of the right lung base (b) and the diaphragmatic dome (c) show the anomalous vein in cross-section (arrows) as it courses medially towards the IVC. R, patient's right side

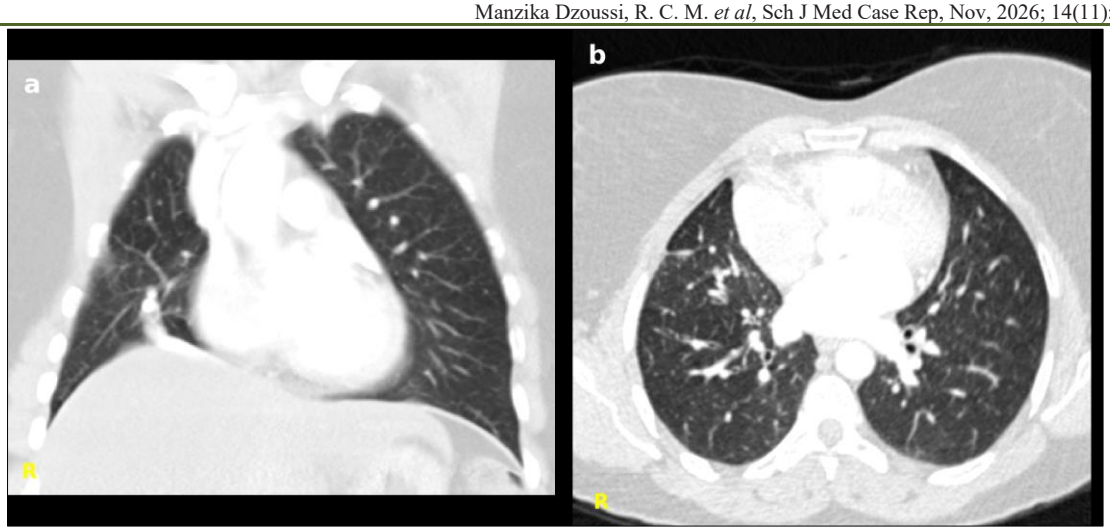


Figure 2: Coronal (a) and axial (b) CT images in lung window. The right hemithorax is reduced in volume, with right lung hypoplasia, rightward shift of the heart (dextroposition) and elevation of the right hemidiaphragm. R, patient's right side

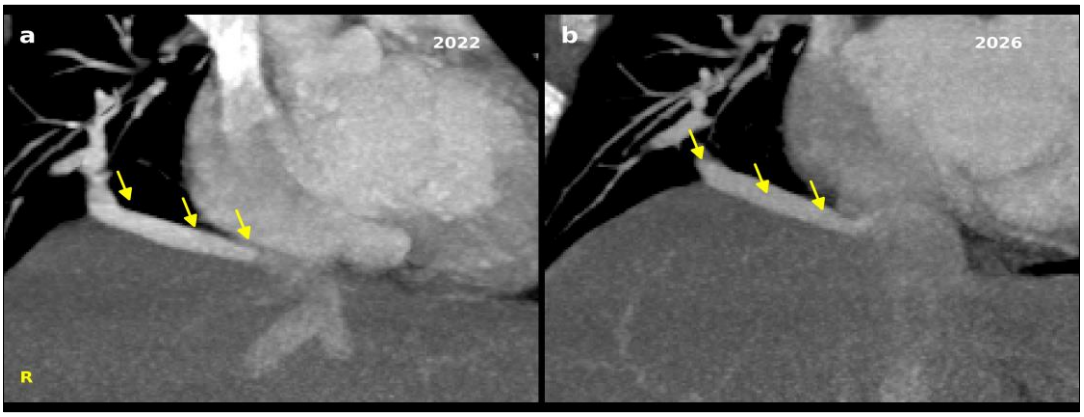


Figure 3: Coronal MIP images from July 2022 (a) and October 2026 (b), reconstructed in matched planes. The course, calibre and caval termination of the scimitar vein (arrows) have not changed over the four-year interval

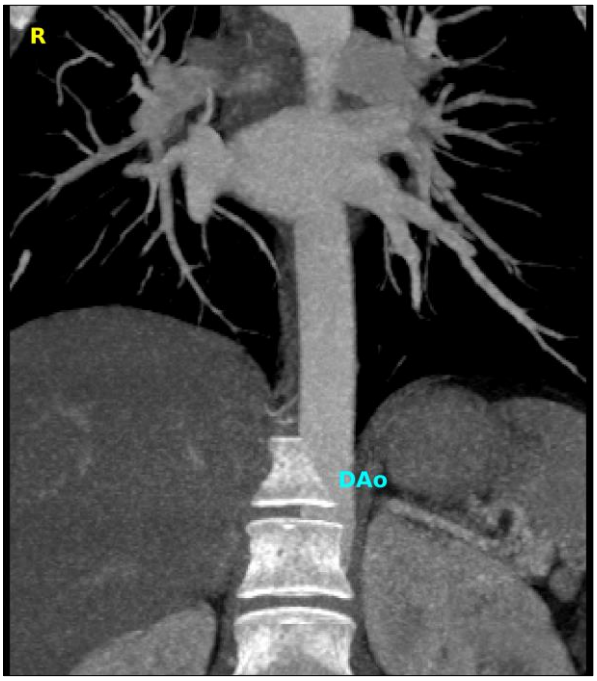


Figure 4: Posterior coronal MIP image acquired in the systemic arterial phase. The descending thoracic and upper abdominal aorta (DAo) has no anomalous systemic arterial branch supplying the right lower lobe. R, patient's right side.

DISCUSSION

Scimitar syndrome is defined by anomalous drainage of part or all of the right lung into the IVC, just above or below the diaphragm [2-4]. The name comes from the curved tubular opacity along the right heart border on frontal chest radiographs, which resembles a Turkish sword. However, this sign is seen in only about half of patients and in a minority of infants [7]. A normal radiograph therefore does not exclude the diagnosis.

Spectrum and Associated Anomalies

Our patient had the classic triad of anomalous venous drainage to the IVC, right lung hypoplasia and dextroposition. Other components should be looked for systematically, because they change both prognosis and treatment. Associated cardiac and extracardiac anomalies are reported in up to three-quarters of patients [5]. In a recent CT-based series, right pulmonary artery hypoplasia was found in 45% of patients, aortopulmonary collaterals in 27%, horseshoe lung in 36% and bronchopulmonary sequestration in 36% [8]. Anomalous systemic arterial supply to the right lower lobe matters for two reasons. It can cause haemoptysis and contribute to pulmonary hypertension, and it can be treated by transcatheter embolisation [9–11]. In our patient, no such artery was found on CT angiography that covered the upper abdominal aorta.

Adult versus Infantile Form

The infantile form typically presents in the first months of life with heart failure and pulmonary hypertension. It is often associated with congenital heart defects, right pulmonary artery hypoplasia and stenosis of the scimitar vein, and its mortality is high [9, 10]. The adult form, as in our case, is usually mild. In an Italian multicentre series of 44 patients whose anomalous drainage was left uncorrected, most of whom were diagnosed in childhood, 39 of 42 survivors (93%) were asymptomatic after a median follow-up of 6.4 years [12]. In a Mayo Clinic cohort of 80 adults with unrepaired partial anomalous pulmonary venous return and an intact atrial septum, clinical status and right heart indices did not deteriorate at 3 and 5 years [13]. Most of these patients, however, had drainage to the superior vena cava or the innominate vein, and only one had drainage to the IVC, which limits how far these results apply to scimitar syndrome. Adult cases have also been reported from Morocco [14]. The stability of our patient over four years is consistent with these data.

Our patient had well-controlled asthma and was asymptomatic. In symptomatic adults, the presenting features of scimitar syndrome include exertional dyspnoea and recurrent respiratory infections [2-5]. When such symptoms are attributed to a common airway disease, the underlying vascular anomaly may go unrecognised. To our knowledge, the coexistence of asthma and scimitar syndrome has not been specifically studied, and no causal relationship can be inferred from a single case.

Role of Imaging

Transthoracic echocardiography is the usual first-line test. It assesses right heart size, pulmonary pressures and the atrial septum. In adults, however, the IVC confluence and the course of an anomalous vein are often poorly seen, and diagnostic errors in anomalous pulmonary venous connections are documented with echocardiography but rarely with CT or MRI [15]. Our case illustrates this limitation: echocardiography had not identified the anomalous return, whereas CT showed it clearly.

CT angiography provides a complete anatomical assessment in a single acquisition [1-17]. The radiology report should state:

1. Which segments drain anomalously, and whether the drainage is partial or total;
2. The course and calibre of the scimitar vein, and the level of its termination relative to the diaphragm, the cavoatrialjunction and the hepatic veins;
3. Any stenosis of the vein;
4. The presence, origin and calibre of any anomalous systemic artery;
5. The calibre of the pulmonary arteries, including right pulmonary artery hypoplasia;
6. Indirect signs of pulmonary hypertension and right heart overload;
7. The atrial septum;
8. The bronchial and parenchymal anomalies of the venolobar spectrum.

Wide coverage down to the upper abdomen is needed, so that an anomalous artery arising from the abdominal aorta or the coeliac trunk is not missed. Cardiac MRI complements CT by quantifying the shunt (Qp/Qs) with phase-contrast imaging, without radiation. 4D flow imaging can show pulmonary venous flow before and after repair [16-18].

The main differential diagnosis is the pseudo-scimitar or meandering pulmonary vein. In this variant, a tortuous vein mimics a scimitar vein but drains normally into the left atrium, so there is no left-to-right shunt [19, 20]. Following the vein to its termination on multiplanar reformations distinguishes the two entities.

Management

Intervention is usually considered when there is a significant left-to-right shunt (Qp/Qs > 1.5) with right ventricular volume overload, pulmonary hypertension, heart failure, recurrent pulmonary infections, scimitar vein stenosis or an associated cardiac defect [2-21]. Surgical options include an intra-atrial baffle, direct reimplantation of the vein into the left atrium and, more recently, in situ pericardial rerouting [6-18]. Reported overall operative mortality ranges from 4.8% to 5.9%

[5]. Post-repair obstruction of the scimitar drainage remains the main complication: it occurred in 25% of operated patients in the European registry and was more frequent when surgery was performed early in life [3-5]. In asymptomatic adults without haemodynamic compromise, conservative follow-up is therefore reasonable [3-13], as in our patient.

Learning Points

- A normal chest radiograph or echocardiogram does not exclude scimitar syndrome in adults; CT angiography is the reference test for anatomical assessment.
- The CT report should describe the level of caval termination, any stenosis of the vein, any anomalous systemic artery and signs of pulmonary hypertension, because these findings determine management.
- Asymptomatic adults with stable anatomy and no haemodynamic compromise can be followed conservatively.

CONCLUSION

Scimitar syndrome may remain unrecognised until adulthood, particularly when respiratory symptoms are attributed to another condition and echocardiography is inconclusive. CT angiography confirms the diagnosis and maps the full venolobar spectrum. It also provides the elements that guide management: the level of caval termination, any venous stenosis, any anomalous systemic arterial supply and signs of pulmonary hypertension. In asymptomatic adults with stable anatomy, conservative follow-up is a reasonable option.

Patient Consent

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

DECLARATIONS

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