

Incidental Finding of Scimitar Syndrome in an Adult Woman Presenting with Intermittent Dysphonia and Dyspnea: A Case Report

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Abstract

Case Report

Scimitar syndrome is a rare congenital pulmonary venolobar anomaly characterized by partial anomalous pulmonary venous return of the right lung into the inferior vena cava, frequently associated with right lung hypoplasia and cardiac malposition. We report the case of a 39-year-old woman presenting with intermittent dysphonia and dyspnea. Laryngeal dysplasia was detected following endoscopy with biopsy. Cervicothoracic CT and contrast-enhanced CT angiography demonstrated imaging features consistent with scimitar syndrome, including anomalous right pulmonary venous drainage into the inferior vena cava, right lung hypoplasia, and dextroposition of the heart. This case highlights an adult presentation of a typically pediatric congenital anomaly and underscores the central role of CT angiography in diagnosis.

Keywords: Scimitar syndrome, partial anomalous pulmonary venous return, CT angiography, lung hypoplasia, congenital vascular anomaly.

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INTRODUCTION

Scimitar syndrome is a rare congenital anomaly belonging to the spectrum of partial anomalous pulmonary venous return, in which venous drainage from the right lung drains partially or totally into the inferior vena cava. It is usually diagnosed in childhood, although adult presentations are increasingly reported in the literature. Typical imaging features include right lung hypoplasia, partial or entire anomalous pulmonary venous return, and cardiac dextroposition.

CASE REPORT

A 39-year-old woman presented with intermittent dysphonia and dyspnea without dysphagia. Nasofibroscopy and biopsy revealed findings described as supraglottic dysplasia, prompting cervicothoracic computed tomography (CT) evaluation.

In the cervical region, no laryngeal mass syndrome was found.

Thoracic contrast-enhanced CT and CT angiography demonstrated:

- Right lung hypoplasia (Fig 1)
- Dextroposition of the heart (Fig 1)
- Anomalous pulmonary venous return from the right lung draining into the inferior vena cava (scimitar vein) (Fig 2)
- Dilatation of the inferior vena cava (Fig 2)
- Enlargement of the pulmonary arterial trunk and branches, suggestive of pulmonary hypertension
- No mediastinal lymphadenopathy
- No pleural or pericardial effusion
- Furthermore, no arterial abnormalities were detected, in particular no signs suggestive of pulmonary sequestration
- These findings were consistent with scimitar syndrome.

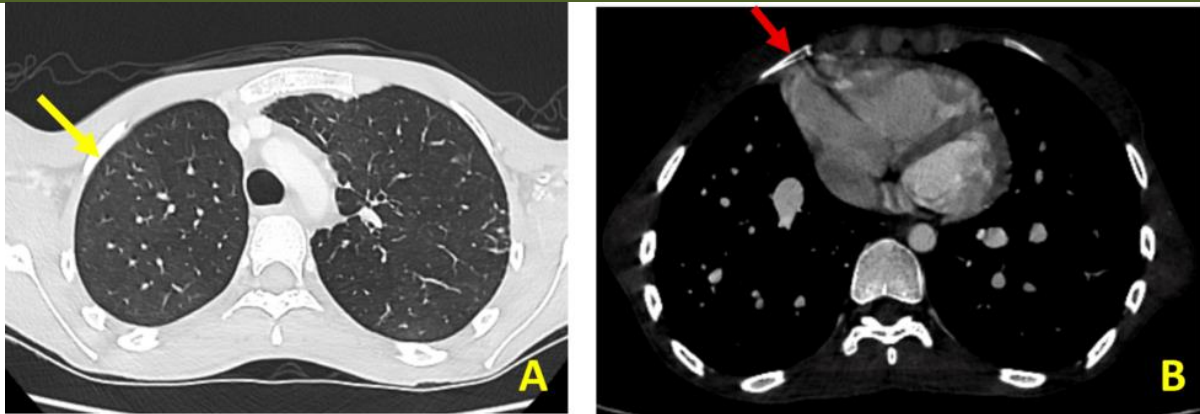


Figure 1: Axial contrast-enhanced CT images in parenchymal (A) an mediastinal (B) window showing right lung hypoplasia (yellow arrow) and mediastinal shift with dextroposition of the heart (red arrow)

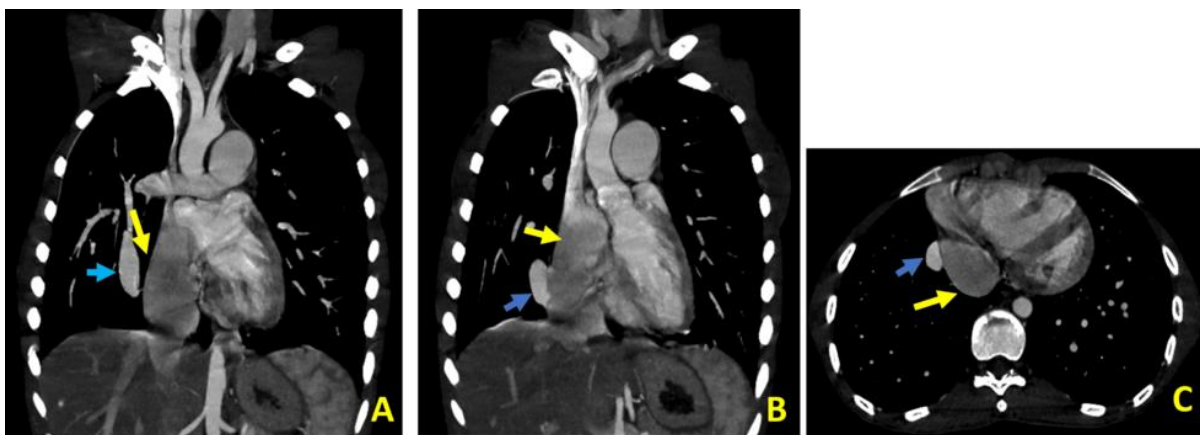


Figure 2: Coronal reconstruction CT (with 5 mm MIP) (A, B) and axial reconstruction (C) demonstrating the venous collector (Scimeter vein, blue arrow) descending along the right cardiac border towards the inferior vena which is dilated (yellow arrow)

In addition, a butterfly vertebra was noted. (Fig 3)

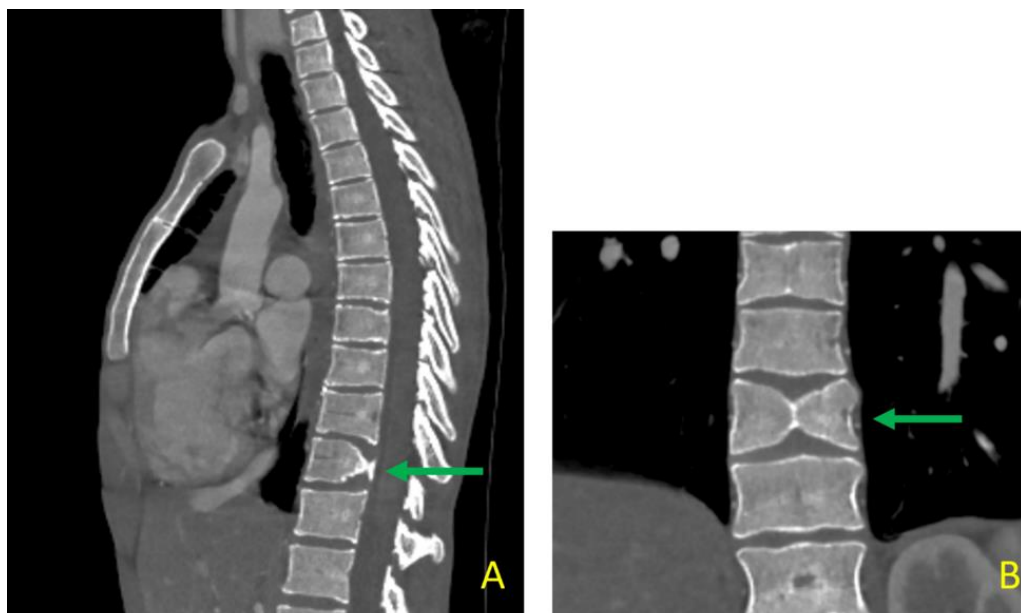


Figure 3: Sagittal (A) and coronal (B) bone reconstruction CT showing associated D11 butterfly vertebra (Green arrow)
Furthermore, signs of a left-sided lung infection, likely caused by inhalation, were found

DISCUSSION

Scimitar syndrome is a rare form of partial anomalous pulmonary venous return, most commonly involving the right lung venous drainage into the inferior vena cava. It is frequently associated with right lung hypoplasia, and cardiac dextroposition. In some cases, systemic arterial supply to the right lung may be present, resulting in associated pulmonary sequestration. [1]

Other associations include congenital heart disease (atrial septal defect, tetralogy of Fallot ...), horseshoe lung, diaphragmatic, vertebral or genitourinary anomalies... [1]

The pediatric form, which is usually severe, presents as heart failure; these cases are generally diagnosed within the first few months of life and tend to be associated with other congenital anomalies. [2]

When diagnosed in adulthood, the condition is typically asymptomatic, although recurrent respiratory infections (predominantly on the right side) or exertional dyspnea may occur. [2]

In this case, the presence of intermittent dyspnea and dysphonia is mostly due to the laryngeal dysplasia, scimitar syndrome is more likely an incidental finding.

Diagnosis can be made by transthoracic or transesophageal echocardiography, angiography, or by CT or MR angiography. [1], [3]

A standard chest X-ray can help identify the diagnosis (hypoplastic right lung, the shadow of the descending pulmonary vein along the right cardiac border shaped like a scimitar (turkish sabre), dextrocardia and mediastinal shift...)

The main differential diagnosis is pulmonary sequestration.

Treatment involves medical management in cases of heart failure, and surgery in cases of significant left-to-right shunt or pulmonary arterial hypertension.

CONCLUSION

Scimitar syndrome, although typically diagnosed in infancy, may present in adulthood with nonspecific respiratory symptoms. CT angiography plays a central role in establishing the diagnosis and assessing associated thoracic anomalies. This case emphasizes the importance of considering congenital vascular anomalies in adult patients with or without unexplained respiratory symptoms.

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Data availability: All relevant data are included in the article. Additional data are available from the corresponding author upon reasonable request. All patient data were anonymised.

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