Case Report

Not all right-sided hearts are the same—the importance of identifying the correct diagnosis

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Abstract: Scimitar syndrome is characterized by an anomalous venous return with the characteristic chest roentgenogram (CxR) appearance of the anomalous vein draining into the inferior vena cava (IVC). This appears as a curvilinear opacity paralleling the right border of the heart resembling a curved sword or Scimitar. A 27-year-old white woman with a reported history of dextrocardia was admitted after a drug overdose. Examination demonstrated an obtunded woman with tachycardia and right sided heart sounds. Her CxR revealed a right sided heart image with two curvilinear opacities in the retrocardiac area. Chest computed tomography (CT) demonstrated that these opacities join to represent an anomalous vein draining into IVC. Furthermore, an anomalous systemic artery arising from the abdominal aorta was seen to supply the right lower lobe. The patient was eventually diagnosed with Scimitar syndrome. This syndrome affects 1–3 in 100,000 live births while nearly half of the patients remain asymptomatic with some initially being misdiagnosed as dextrocardia, such as in our case. Correctly diagnosing these patients is of paramount importance as some can develop severe pulmonary hypertension and right ventricular failure. In turn, close ongoing echocardiographic monitoring can help identify those that may benefit from surgical interventions to prevent them from developing these complications.

Keywords: Partial anomalous venous return; congenital pulmonary venolobar syndrome (CPVS); Scimitar syndrome; dextrocardia

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Case presentation

A 27-year-old white woman with a reported history of "dextrocardia" was admitted after a drug overdose. On admission she was obtunded and tachycardic, but hemodynamically stable. Heart sounds were more prominent over the right chest. Lung auscultation was normal and the rest of the exam was unremarkable. She was intubated due to decreased level of consciousness, but was successfully extubated the next day. She was eventually transferred to the psychiatry ward for further treatment.

Urine toxicology screen revealed tricyclic antidepressants. Head computed tomography (CT) scan was normal. An electrocardiogram revealed sinus tachycardia. A chest roentgenogram (CxR) confirmed heart malpositioning (*Figure 1*). Additionally, she underwent chest CT angiography (*Figures 2-5*) and echocardiography (*Figure 6*).

At this point the differential diagnosis included Scimitar syndrome, Sawyer James syndrome, Kartagener syndrome and dextrocardia of embryonic arrest. However, closer inspection of the chest imaging revealed dextropositioning of the heart rather than dextrocardia, which essentially ruled out Kartagener syndrome and dextrocardia of embryonic arrest. Additionally, in the absence of recurrent childhood infections and presence of abnormal arterial and venous blood supply, the diagnosis of Sawyer James syndrome was not considered to be likely.

The final diagnosis was Scimitar syndrome. The CxR revealed two Scimitar veins, with chest CT angiography confirming these veins joining into a common vessel draining to the supra-hepatic inferior vena cava (IVC). Additionally, there was an aberrant systemic arterial supply to the right lower lobe, arising directly from the abdominal aorta. Other findings included a hypoplastic right pulmonary artery, hypoplasia of the right lung, and

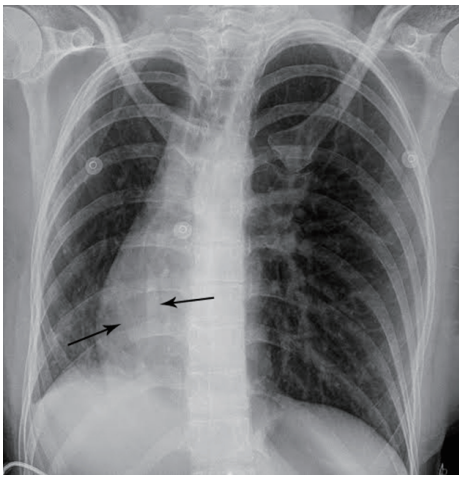

Figure 1 Frontal chest radiograph demonstrates characteristic features of Scimitar syndrome, including an asymmetrically smaller right lung with reduced pulmonary vascularity relative to the left lung, a right-sided cardiac shadow, and vertically oriented tubular densities overlying the cardiac shadow representing the scimitar veins in this case.

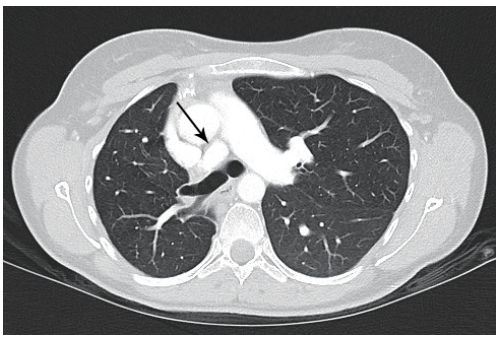

Figure 2 Axial CT image demonstrating a hypoplastic right lung and hypoplastic right pulmonary artery (arrow). CT, computed tomography.

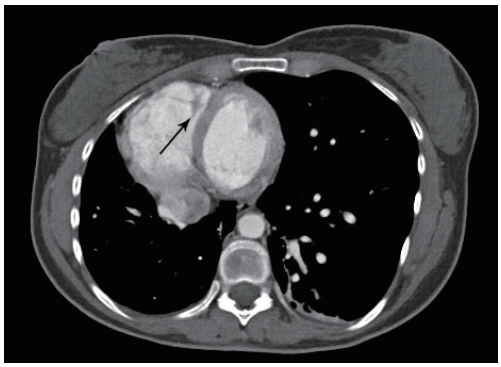

Figure 3 Axial CT image demonstrating right-sided positioning of the morphologic right ventricle as defined by the moderator band (arrow), indicating this is dextropositioning of the heart consequent to right lung hypoplasia, rather than dextrocardia. CT, computed tomography.

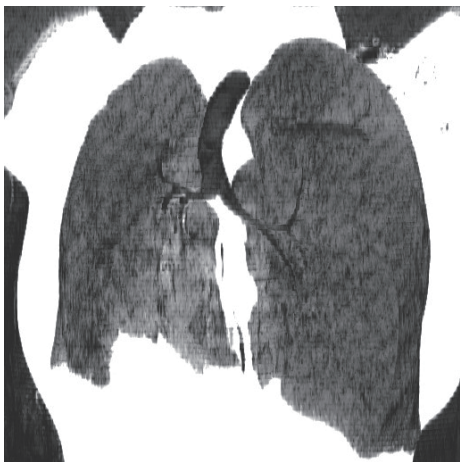

Figure 4 Coronal minimum-intensity projection demonstrating right lung hypoplasia and anomalous right lung bronchial anatomy mirroring left bronchial anatomy (left bronchial isomerism).

abnormal right lung lobation with a hyparterial right bronchus (left sided bronchial isomerism). These features are illustrated in *Figures 2-5*.

This patient was discharged home from the psychiatry ward. She was counseled regarding her diagnosis and a repeat echocardiogram and follow up visit with her primary care physician were suggested.

Discussion

Although the syndrome was first described by Chassinat and Cooper (1,2) nearly 300 years ago, the exact anatomical abnormalities were only delineated in the mid-twentieth century (3) when the term "Scimitar syndrome" was first used (4). Since then, several reports have described the corresponding anatomical abnormalities, though confusion persists regarding the classification (5,6) and management of this condition. Some include this as one of the congenital pulmonary venolobar syndromes (CPVS) (7), but many regard it as a form of partial anomalous pulmonary venous return (PAPVR) (8).

The syndrome is constantly characterized by PAPVR

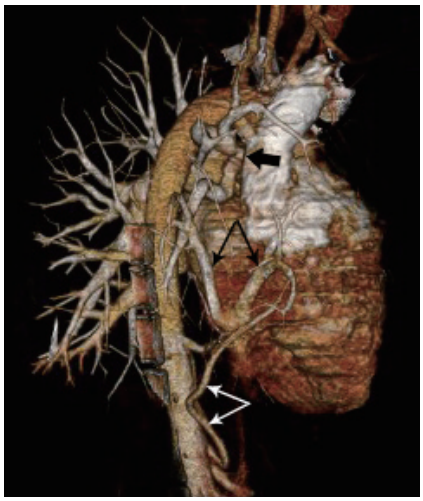

Figure 5 Volume rendered right posterior oblique view demonstrating hypoplastic right pulmonary artery branches (thick black arrow), the anomalous right pulmonary (Scimitar) veins draining to the suprahepatic IVC (thin black arrows), and a systemic artery supplying the right lower lobe arising from the abdominal aorta (thin white arrows). IVC, inferior vena cava.

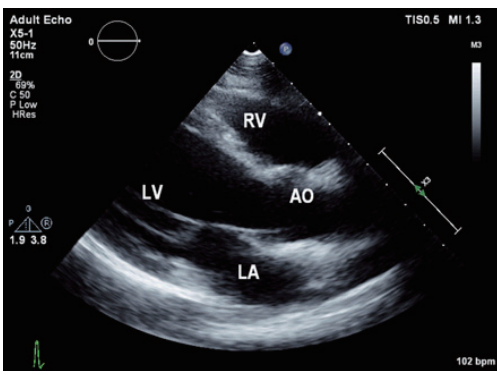

Figure 6 In this case the patient has dextroposition of the heart in which there was normal alignment of the major axis of the heart. The echocardiogram views were done with the echo probe on the right side of the sternum.

with oxygenated blood from the right lung draining directly into the IVC. This anomalous vein appears as a curvilinear opacity parallel to the right heart border, forming the characteristic CxR appearance of a curved sword, the "Scimitar sign". The Scimitar syndrome is clearly differentiated from other forms of CPVS or PAPVR by this characteristic "Scimitar" shaped vein which is not present when it drains to other areas such as the superior vena cava (SVC). Associated anomalies include hypoplasia and abnormal lobation of the right lung, pulmonary sequestration, right pulmonary artery hypoplasia, heart dextroposition, anomalous systemic right lower lung arterial supply, atrial septal defect, and right diaphragmatic hernia (9).

This condition is usually diagnosed in older children during evaluation for unexplained dyspnea, but the diagnosis is seldom entertained in adults due to its rarity and lack of recognition. The incidence is reported as 1–3 in 100,000 live births (10), although the true incidence might be higher, as nearly half of the patients remain asymptomatic.

There is a correlation between presenting age and disease severity, with the disease accordingly being divided into three groups. Group 1 is the adult form with no evidence of pulmonary artery hypertension (PAH), as in our patient. Group 3 is the infantile form with severe PAH, conveying poor prognosis. The remaining patients fall into group 2 and have variable degrees of PAH and symptoms (11).

The most common radiographic findings include heart dextroposition and right sided mediastinal shift with compensatory left lung hyperinflation. However, the most pathognomonic radiographic feature remains the Scimitar sign, corresponding to the anomalous vein draining into the IVC. Therefore, the "Scimitar syndrome" term should be reserved only for cases of PAPVR draining to the IVC, despite pathophysiological commonalities with those with alternate drainage, such as to the SVC. Finally, one must understand that this finding may be occasionally obscured, due to the degree of dextroposition. In these cases, echocardiography and chest CT angiography should be performed to establish the correct diagnosis.

Echocardiography allows not only detecting the Scimitar vein behind the right atrium (12), but also identifying other commonly associated cardiac anomalies. Currently, there is increased recognition of this syndrome in-utero using echocardiography which aids in planning care for some of the complicated cases.

CT scans have been used to describe PAPVR since the 1980s. Currently, advanced technology with 3-D reconstructed images, aside from visualizing thoracic vasculature (13), provides information on the degree of left-to-right shunting (14). Furthermore, CT scans detect lung sequestration which, albeit an elusive finding, is an important one as it poses a risk for recurrent lung infections. Finally, despite all the information gathered from non-invasive studies, cardiac catheterization is still sometimes

required to confirm pulmonary hypertension and/or to better define the degree of pulmonary hypertension.

Management of the Scimitar syndrome focuses on risk prevention, such as avoiding recurrent infections, with identification of sequestered lung and resection when necessary. Additionally, repair of the anomalous pulmonary venous return in symptomatic patients with moderate to severe left to right shunting and associated right ventricular strain and pulmonary hypertension is required (15). Interestingly, there is a reported case of congenitally stenotic scimitar vessel in which the scimitar vessel was stenotic at the junction of the IVC allowing for the majority of the oxygenated blood from the lung to be drained into the left atrium and hence not requiring repair of the anomalous pulmonary venous return (16). This case exemplifies the need for detailed and objective assessment of the degree of shunting, in order to better guide therapy. Experts suggest using Doppler echocardiography (17) to determine pulmonary and systemic flow ratios (Qp:Qs) estimating left to right heart shunt. A ratio of 1:1 indicates equal volumes of blood travel through the lungs (Qp) and the systemic circulation (Qs) (18). In other congenital cardiac conditions the shunt ratio helps prognosticate the need for surgery (8,19). A similar approach could be used in the Scimitar syndrome, suggesting that ratios ≥1.5 indicate consideration for corrective surgery.

Fortunately, most patients with Scimitar syndrome remain asymptomatic, leading normal lives. However, to avoid potential complications and evolving cardio-pulmonary pathology, we propose that, aside from identifying and correcting lung sequestration when needed (with recurrent lung infections), these patients should have yearly (sooner when symptomatic) echocardiography to assess for the presence and/or degree of left to right shunt (20).

Additionally, management of aberrant arterial blood supply of the sequestered lung segment is worth mentioning as patients with Scimitar syndrome often present with a sequestered lung segment. There has been a case report that details fatal complications as a result of disruption of systemic arterial supply of the intra-lobar sequestered lung segment (21). Fortunately, our patient did not present with this complication. Detailed explanation of this condition to the patient and the family can help in earlier recognition and management of this potentially fatal complication.

In conclusion, we present a case illustrating the importance of distinction between cardiac dextroposition and true dextrocardia as adequate recognition of the pathology in patients with a right sided heart allows physicians to provide better care and avoid potential complications of the condition, particularly if the diagnosis is "Scimitar syndrome".

Learning points

(I) Not all right sided hearts represent dextrocardia;
(II) Patients with heart malpositioning must have careful review of chest imaging;
(III) Further confirmation of Scimitar syndrome can be made through echocardiography and heart catheterization;
(IV) Patients with Scimitar syndrome should be monitored closely for developing pulmonary hypertension and consideration for potential surgery in order to avert right heart failure;
(V) Sequestered lung is an important consideration when assessing for recurrent lung infections.

Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Informed Consent: Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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