ESC European Society of Cardiology

European Heart Journal - Case Reports (2019) **3**, 1–4
doi:10.1093/ehjcr/ytz050

CASE REPORT
Congenital heart disease

Scimitar sign in the absence of anomalous pulmonary venous drainage: a case report

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Received 28 September 2018; accepted 2 April 2019; online publish-ahead-of-print 23 April 2019

Background Scimitar syndrome consists of anomalous pulmonary vein drainage to the inferior vena cava. Its name derives from the image this anomalous pulmonary vein creates on a chest radiograph. We describe a case of normal venous pulmonary vein drainage that also presented the scimitar sign due to an aorto-collateral vessel.

Case summary A 15-month-old girl presented with mild dyspnoea and fever. Control chest X-ray showed an image of cardiac dextroposition, hypoplastic right lung, and the 'scimitar sign'. Although the transthoracic echocardiogram confirmed the initial suspicion of anomalous pulmonary venous drainage, the computed tomography (CT) scan showed normal right pulmonary veins connected to the left atrium and revealed that an aorto-collateral vessel caused the scimitar sign.

Discussion Although the patient had several typical alterations of the scimitar syndrome, the pulmonary venous connection was normal, and the scimitar sign was due to an aorto-collateral vessel. It might be difficult to describe venous pulmonary connections on the basis of echocardiography, so an angio CT scan proved to be a valuable tool in this scenario.

Keywords Anomalous pulmonary venous drainage • Scimitar syndrome • Case report • Congenital heart disease • Collateral artery

Learning points

- The radiologic 'scimitar sign' may be present in patients with normal right pulmonary veins connection. This sign may be caused by an aorto-collateral vessel.
- Echocardiography may fail to describe the venous pulmonary connections, so multimodality evaluation is frequently needed to obtain a broad evaluation of complex cardiac anatomy in patients with congenital heart disease.

Introduction

Scimitar syndrome is a rare disease (with incidence between 1 and 3 in 100 000 live births)[1] and consists of partial or total anomalous right pulmonary venous drainage to the inferior vena cava (IVC). This syndrome is commonly associated with hypoplasia of the right lung, pulmonary sequestration, and dextroposition of the heart,[1] and appears as a 'scimitar sign' at frontal radiography.[2] We describe an unusual case of scimitar sign in the absence of anomalous pulmonary venous drainage.

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Handling Editor: Riccardo Liga
Peer-reviewers: Elena Surkova and George Giannakoulas
Compliance Editor: Christian Fielder Camm
Supplementary Material Editor: Peysh A. Patel

Timeline

Day 1	Mild dyspnoea and fever; a pneumonia diagnosis was given and antibiotics started
Day 3	Chest X-ray showed a hypoplasic right lung, cardiac dextroposition, and a 'scimitar sign'
Day 4	Scimitar syndrome confirmed by echocardiogram
Day 6	Angio computed tomography scan showed normal pulmonary venous connection and an aorto-collateral vessel that was responsible for the scimitar sign
Day 15	Cardiac catheterization showed normal pulmonary artery pressure and confirmed that the scimitar sign was due the collateral vessel
Day 16	Patient in good clinical status, been followed-up at outpatient clinic

Case presentation

A previously asymptomatic 15-month-old girl presented with mild dyspnoea and fever (up to 38.5°C for the last 4 days). She did not present with cough or feeding difficulties; there was no previous history of breathlessness or recurrent infections, either. On initial presentation, the patient was euvolaemic, the heartbeat was regular with a heart rate of 105 b.p.m., and heart sounds were normal without murmurs. Arterial blood pressure was 90 × 50 mmHg, and oxygen saturation in room air was 96%. The respiratory rate was 26 b.p.m. Pneumonia was diagnosed, and antibiotics were started (amoxicillin-clavulanate—90 mg/kg/day PO divided in two doses for 14 days). The clinical examination remained abnormal after antibiotic treatment, and low-intensity vesicular breath sounds were present in the right thorax. A chest X-ray (*Figure 1*) revealed cardiac dextroposition, a small right lung, and a vascular shadow behind the heart border, which raised suspicion of scimitar syndrome. A transthoracic echocardiogram showed a 4-mm ostium secundum atrial septal defect (ASD) in the fossa ovalis region, with left-to-right shunt, and right inferior pulmonary vein connected with the IVC, thus supporting the diagnosis of scimitar syndrome (*Figure 2*).

A computed tomography angiography scan evidenced small ostium secundum ASD (5 mm), hypoplasia of the right lung, agenesis of the right pulmonary artery, normally connected pulmonary veins (*Figure 3*), and a collateral artery arising from the abdominal aorta supplying the hypoplastic right lung (*Figure 4*). Surprisingly, the collateral artery accounted for the scimitar sign on the chest X-ray, and there was no anomalous vein drainage to IVC as shown by echo. The chest computed tomography (CT) scan also showed altered bronchial segmentation with a blind-ended right bronchus to the intermediate lobe.

Cardiac catheterization confirmed the diagnosis and showed systolic left pulmonary artery pressure of 34 mmHg (slightly increased), systolic aortic pressure of 74 mmHg, and systolic pressure of 22 mmHg at the distal collateral vessel due to stenosis at its origin. The mean pressure values at left and right atria were respectively

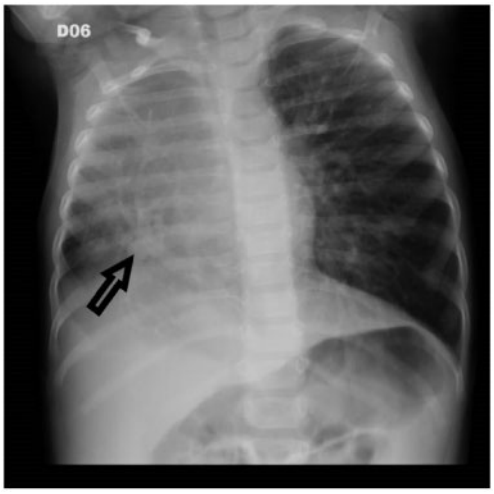

Figure 1 Chest X-ray showing the scimitar sign (arrow), hypoplastic right lung, and dextrocardia.

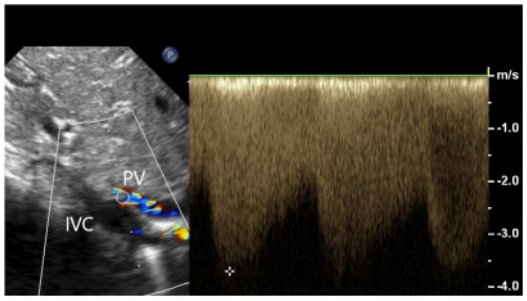

Figure 2 Echocardiogram, subcostal view: the aorto-collateral vessel was mistaken for an anomalous pulmonary vein draining to the inferior vena cava. IVC, inferior vena cava; PV, pulmonary vein.

Figure 3 Three-dimensional volume-rendered computed tomography reconstruction shows normal drainage of the right pulmonary veins to the left atrium (white arrows). LA, left atrium.

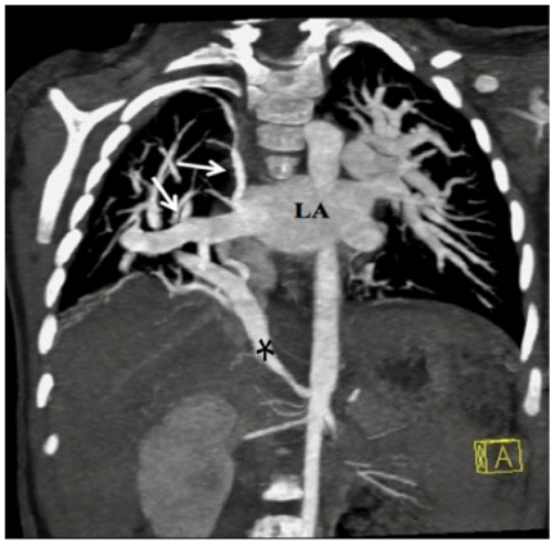

Figure 4 Maximum intensity projection image in a coronal plane of the computed tomography scan demonstrating the right pulmonary veins and their connection to the left atrium (white arrows). The aortopulmonary collateral arising from the abdominal aorta to the right lung is also seen (*). LA, left atrium.

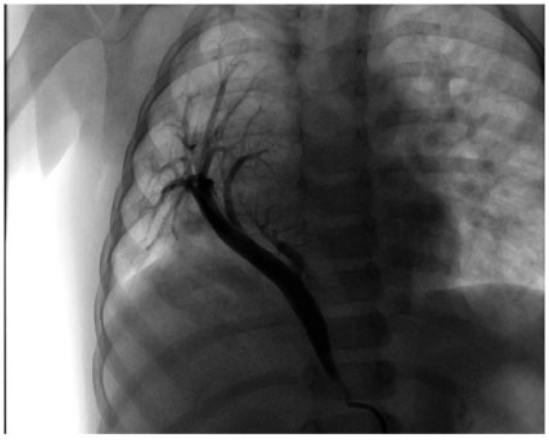

Figure 5 Angiogram of the aorto-collateral vessel mimicking the 'scimitar sign'.

13 and 10 mmHg, pointing to a small, restrictive ASD. Catheterization also confirmed that the scimitar sign was due to the collateral vessel (*Figure 5*).

The patient recovered well from pneumonia. Because she had no anomalous venous connection and presented very good clinical evolution, the clinical group decided to re-evaluate the ASD in the near future. Eighteen months after the initial diagnosis, the patient remains asymptomatic, and the transthoracic echo did not show any alterations compared with the initial one.

Discussion

Scimitar syndrome consists of anomalous drainage of the right pulmonary vein to the IVC and is usually associated with ASD, right lung hypoplasia, and dextrocardia.[3] Its name derives from the typical scimitar appearance the syndrome creates on a chest radiograph. Agenesia of the right pulmonary artery and systemic blood supply to the right lung by aorto-collateral vessels are also usually described.[4]

Transthoracic echocardiography is usually the first-line imaging diagnostic test for patients with suspected pulmonary venous anomalies. However, echocardiography is constrained by acoustic windows and has limited ability to display the complex three-dimensional relationships in some patients with pulmonary venous anomalies. The diagnostic accuracy also depends on the operator's skills and working experience. Although direct visualization of anomalous venoatrial connections is desirable, it is not always possible, so imaging modalities such as CT can be useful. In the case described here, the operator mistook an aortopulmonary collateral vessel for anomalous pulmonary vein drainage to the IVC. The transthoracic echocardiography pointed to an initial diagnosis of scimitar syndrome, but the CT provided more comprehensive and accurate imaging evaluation of the pulmonary vein connection. Cardiac magnetic resonance imaging and transoesophageal echo may not always be an option due to the lengthy examination time, need for sedation although the procedure does not involve exposure to radiation.[5]

This case highlights that multimodality evaluation is frequently needed to obtain a broad evaluation of complex cardiac anatomy in patients with congenital heart disease.[5]

While most authors claim that the scimitar syndrome must have anomalous pulmonary venous drainage, others have described variants of the syndrome with pulmonary venous drainage to the left atrium.[6]

The scimitar sign has already been described in cases with normal pulmonary venous drainage.[7] The sign may present with the radiologic signal due to a large abnormal pulmonary vein—'meandering vein'—with normal drainage to the left atrium.[8]

The treatment of patients with scimitar syndrome must be individualized, and the medical group must consider the age of presentation and the presence of heart failure and associated lesions before defining clinical or surgical treatment. Even though right lung lobectomy may be indicated in patients due to recurrent right lung infections, this patient has had just one episode so far. Lobectomy or pneumonectomy is only performed in patients with recurrent infections, diffuse bronchiectasia, or haemoptysis. Since her right bronchus is blind-ended, close surveillance in future episodes of pulmonary infections is warranted.[4,9]

This is a unique case. The scimitar sign due to a collateral vessel has never been described. The patient presented not only the radiologic sign, but also all the main features of the scimitar syndrome except anomalous pulmonary vein drainage. Since anomalous pulmonary vein drainage is absent, this case may not be classified as scimitar syndrome or scimitar variant either, because pulmonary vein drainage is normal.

Conclusion

Scimitar syndrome is a rare, complex congenital heart disease with a broad range of presentations. The radiologic 'scimitar sign' may be present in patients with normal right pulmonary vein connection. The sign may be due to an aorto-collateral vessel. Hence, a multimodality evaluation is needed to improve diagnosis accuracy.

Lead author biography

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Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Acknowledgements

The authors thank to Cynthia Manso for her kind English revision.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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