

Anomalous pulmonary venous drainage: chest radiography and cardiac imaging

INTRODUCTION

Scimitar syndrome (or pulmonary vena-lobar syndrome) is a rare congenital cardiac condition characterised by abnormal pulmonary venous return from the right lung directly into the inferior vena cava. It can be associated with hypoplastic right lung, pulmonary artery hypoplasia and dextrocardia, among others.^{1–3} There are complete and incomplete variations. In the complete form, classically, the right lung is hypoplastic and is supplied by a systemic arterial branch from the aorta with hypoplasia of the right pulmonary artery, resulting in an abnormal right heart border on imaging.

Most cases are diagnosed incidentally and are asymptomatic. Surgical treatment is indicated if there is a large left to right shunt (>50%) leading to pulmonary hypertension. Other indications also include lung sequestration, recurrent right-sided chest infections, or recurrent haemoptysis sometimes secondary to systemic arterial pressure supplying the vessels of the lung.^{1–4}

We present a case of incomplete form of Scimitar syndrome picked up by review of chest radiography and include images from other complementary cardiac imaging modalities.

CASE PRESENTATION

A 22-year-old woman, previously well, presented to our general medical unit with a cough and was investigated for a suspected chest infection. On admission, a chest radiograph was performed. This, somewhat surprisingly demonstrated radiographic

features consistent with either a right-sided pleural effusion or volume loss (figure 1A, thick arrow).

To investigate further, contrast CT of the chest was performed. No pulmonary thromboembolism was demonstrated during the arterial phase of the study; however, a clear abnormality was seen in the venous phase. The presence of a right-sided anomalous (Scimitar) vein draining into the intrahepatic inferior vena cava (IVC) (figure 1B, arrow) was clearly visible, but no pulmonary sequestration was seen. In retrospect, this anomaly could be noted in the chest radiograph as well (figure 1A, thin arrow). The CT confirmed that there was demonstrable left-sided pulmonary venous drainage, but total absence of any direct right-sided pulmonary venous drainage into the left atrium (figure 1C, arrow).

We undertook transthoracic and transoesophageal echocardiography to exclude associated cardiac anomalies and to confirm the absence of any direct right-sided pulmonary venous drainage into the left atrium. A right pulmonary vein (RPV) was draining into the intrahepatic portion of the IVC (figure 1D) was seen, and no right-sided pulmonary venous drainage into the left atrium was seen.

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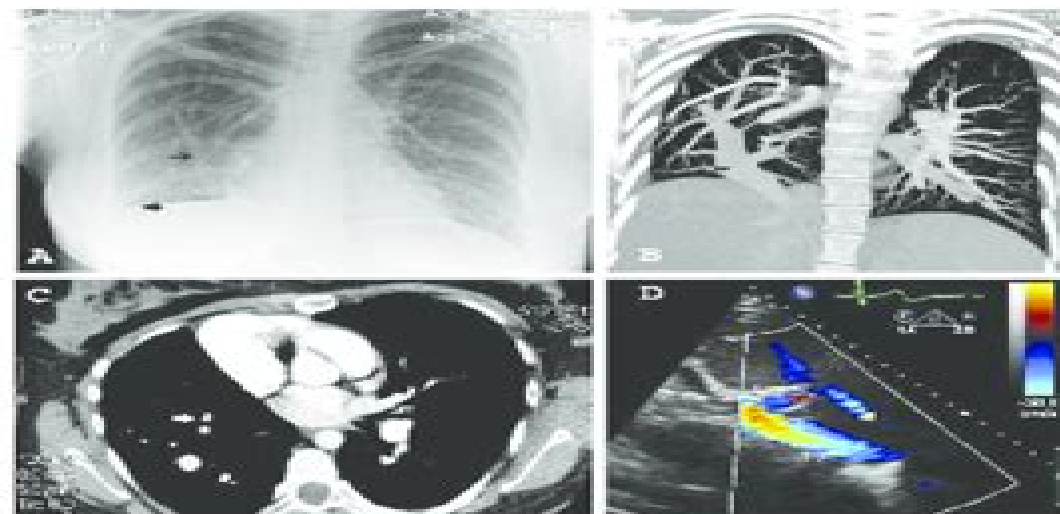
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Acknowledgements: Department of Cardiology, Ninewells Hospital and Medical School, Dundee, UK.

Contributors: I have collected images and written the manuscript, initial review and restructuring with Dr Koch. 2nd Review and additions by Dr Mordi and Tziemon.

Figure 1 Multi-modality radiographic (chest radiography (A), computed tomography of chest with contrast (B, C) and trans-thoracic echocardiographic (D, subcostal view) images illustrating partial anomalous pulmonary venous drainage. This figure is only reproduced in colour in the online version.



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