Scimitar sign with normal pulmonary venous drainage and anomalous inferior vena cava

B HERER, F JAUBERT, C DELAISEMENTS, G HUCHON, J CHRETIEN
From the Clinique de Pneumo-Phtisiologie, Université René Descartes, Hôpital Laennec, Paris, France

ABSTRACT A case of the scimitar sign due to an anomaly of the right sided pulmonary vein with normal drainage into the left atrium was associated with an azygos continuation of the inferior vena cava. Digital subtraction angiography allows the identification of these rare congenital vascular malformations.

The “scimitar syndrome” is a combination of a hypoplastic right lung, dextrocardia, and anomalous right sided pulmonary venous drainage into the inferior vena cava. It produces a curved vascular shadow along the right heart border on the chest radiograph, the scimitar sign.1 We report a case in which the scimitar sign was due to an anomaly of the right pulmonary vein with normal drainage into the left atrium. This malformation was associated with an anomalous systemic venous drainage without evident cardiac abnormality. A similar syndrome has not been described previously.

Case report

A 48 year old non-smoking woman was referred for investigation of an abnormal chest radiograph and a history of mild dyspnoea of recent onset with an occasional unproductive cough. She had no past history of cardiac or pulmonary disease and no known personal or family history of congenital malformations. She had five healthy children and was herself the product of a normal pregnancy.

Physical examination showed a healthy looking woman with no cyanosis or finger clubbing. The thorax was symmetrical, the heart sounds were normal, and the breath sounds were vesicular. The rest of the clinical examination showed nothing abnormal. The electrocardiograph and results of lung function tests were normal. Arterial blood gas values were: an oxygen tension (P0₂) of 12-4 kPa, a carbon dioxide tension (PCO₂) 5-2 kPa, and pH 7-36. A chest radiograph disclosed a scimitar sign as a curvilinear vascular shadow running parallel to the right heart border (fig 1). Pulmonary scintigraphy showed a substantial decrease in perfusion of the right lung. Echocardiography showed nothing abnormal.

A conventional angiogram via the right femoral vein disclosed an infrahepatic interruption of the inferior vena cava with an azygos vein continuation. The supradiaphragmatic examination was non-contributory owing to incom-

Fig 1 Original chest radiograph of the patient showing the scimitar sign.

Fig 2 Digital subtraction angiogram showing a large pulmonary vein descending to the diaphragm and making a sharp turn to drain into the left atrium.

Address for reprint requests: Dr B Herer, Hôpital Laennec, 42 rue de Sévres, F-75040 Paris, France.

Accepted 15 April 1988
Phragn and then made a sharp turn upwards to drain completely into the left atrium (fig 2). No pulmonary arteriovenous fistula was detected. The second part of the examination confirmed the infrahepatic interruption of the inferior vena cava with an azygos continuation.

Discussion

The presence of a scimitar sign is attributable here to an abnormally large pulmonary vein draining normally into the left atrium. The scimitar sign is usually seen as part of the scimitar syndrome. The syndrome is characterised by anomalous right sided pulmonary venous drainage into the inferior vena cava in association with other anomalies, mainly right lung hypoplasia, dextrocardia, and abnormal bronchopulmonary segmentation or other abnormal vascular connections.1 The scimitar sign may, however, be seen in many pathological conditions—for example, anomalous right pulmonary venous drainage to the superior vena cava2 or the left atrium and inferior vena cava,3 or in exceptional cases, as in our patient, exclusively to the left atrium.4 A pulmonary vessel sequestration by a right subphrenic artery has also been described.5 Our case shows an abnormality similar to the one described by Morgan et al,4 but information from digital subtraction angiograms was sufficient to eliminate the need for conventional angiography.

In addition, in our patient the azygos vein was found to continue into the inferior vena cava. The association of these two unusual vascular malformations in the same patient has, to our knowledge, been described. Although it could be a coincidental occurrence, our observation suggests that a single event during fetal development was the origin of this dual vascular malformation.6 Digital subtraction angiography could be useful in the search of associated systemic venous abnormalities in cases of anomalous pulmonary venous drainage.

References

Scimitar sign with normal pulmonary venous drainage and anomalous inferior vena cava.
B Herer, F Jaubert, C Delaisements, G Huchon and J Chretien

Thorax 1988 43: 651-652
doi: 10.1136/thx.43.8.651

Updated information and services can be found at:
http://thorax.bmj.com/content/43/8/651

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/