Congenital and acquired heart disease converge: ischaemic heart disease with a background of scimitar syndrome

Ioannis Dimarakis¹, Vaikom S. Mahadevan², and J. Andreas Hoschitzky¹*

¹Department of Cardiothoracic Surgery, Manchester Royal Infirmary, Oxford Road, Manchester, M13 9WL, UK; and ²Department of Cardiology, Manchester Royal Infirmary, Manchester, UK

* Corresponding author. Tel: +44 161 901 0122, Fax: +44 161 276 8522, Email: andreas.hoschitzky@cmft.nhs.uk

A 66-year-old woman presented with NYHA and CCS class III symptoms. On questioning, her breathlessness was found to trace back to young adulthood worsening over time, and her angina was found to be present only in the preceding 5 months. Medical history included chronic obstructive pulmonary disease, cerebrovascular accident, peptic ulcer disease, and hypothyroidism. Chest roentgenogram demonstrated an increased cardiothoracic ratio and the classic ‘scimitar’ sign, a tubular opacity resembling a scimitar created by the curvilinear course of the anomalous pulmonary vein towards the right cardiophrenic angle (Panel A). Computed tomography confirmed drainage of the entire right-sided venous return via the scimitar vein into the inferior vena cava at the level of the diaphragm with enlarged pulmonary vessels suggesting pulmonary hypertension (Panels B and C). She underwent transoesophageal echocardiography and right/left cardiac catheterization, which demonstrated dilatation of the right heart chambers with scimitar vein drainage into the inferior vena cava (Panel D; see Supplementary material online, Video S1), a mean pulmonary-to-systemic flow ratio (Qp:Qs) of 1.9:1, and a 70% proximal lesion in a large dominant right coronary artery (see Supplementary material online, Video S2). In view of the aforementioned findings, we proceeded to concomitant correction of the anomalous pulmonary venous drainage and coronary artery bypass graft surgery. The anomalous pulmonary venous drainage was baffled with a native pericardial patch to the left atrium via a surgically created secundum atrial septal defect. A reverse saphenous vein graft from the ascending aorta was anastomosed to the distal right coronary artery. The postoperative recovery was uneventful and 6 months after the surgery, the patient remains well.

In adult presentations of scimitar syndrome, contemporaneous acquired pathology must be excluded prior to surgical correction.

Supplementary material is available at European Heart Journal online.