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Brief Report

Scimitar syndrome and anomalous origin of the circumflex artery from the main pulmonary artery in infancy: a case report

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Abstract We present a neonate with an antenatal diagnosis of Scimitar syndrome and aortic arch hypoplasia. After delivery, computerised tomography scan additionally revealed an anomalous origin of the circumflex coronary artery from the main pulmonary artery. The management of this rare combination is discussed.

Keywords: Scimitar syndrome; anomalous coronary; anomalous pulmonary venous return

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A FEMALE INFANT WAS BORN AT TERM AFTER AN antenatal diagnosis of Scimitar syndrome and aortic arch hypoplasia. On examination she was fully saturated and had normal pulses. There were reduced breath sounds over the right lung and normal heart sounds with no murmurs. In view of a possible coarctation, prostaglandin therapy was commenced and she was transferred to our institution. The electrocardiogram on admission was normal and the echocardiogram confirmed the diagnosis of Scimitar syndrome with all right-sided pulmonary veins draining into the inferior caval vein, a hypoplastic right pulmonary artery, and aortopulmonary collaterals arising from the descending aorta. There was only mild arch hypoplasia with no coarctation. The origins of the coronary arteries were seen but not the bifurcation of the left main stem.

A computerised tomography angiogram was performed to further delineate the anatomy. In addition to the above findings, it showed the left circumflex artery arising from the main pulmonary artery at the level of the origin of the right pulmonary artery. There were two aortopulmonary collaterals arising from the descending aorta and supplying a sequestered basal segment of the right lung (Figs 1 and 2). Cardiac catheterisation with coil embolisation of the aortopulmonary collateral to the right lung was performed. The anomalous circumflex artery was confirmed and the pulmonary pressures were suprasystemic.

The infant was discharged with ambulatory follow-up. At 7 weeks of age, both the electrocardiogram and echocardiogram showed evidence of ischaemia in the region of the circumflex artery with posterior wall dyskinesia, and she was therefore referred for surgery.

Intra-operatively, the circumflex artery was seen arising from the origin and undersurface of the hypoplastic right pulmonary artery and was supplying a large area of the left ventricle. The main pulmonary artery was divided and the ostium of the circumflex artery was detached with a button of the right pulmonary artery wall and mobilised by a few millimetres. There was significant collateral backflow through this artery. The right pulmonary artery was repaired with a patch of autologous pericardium. The circumflex artery was re-implanted into the ascending aorta through a trapdoor incision just above

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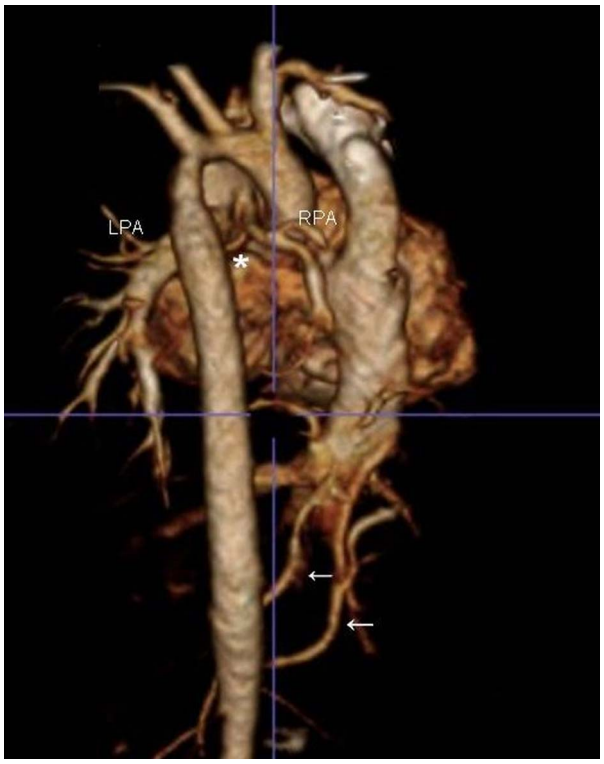

Figure 1.
A computerised tomography angiogram reconstruction – volume rendering, posterior view – showing the origin of the circumflex artery (*) from the inferior aspect of the hypoplastic right pulmonary artery. The arrows indicate the two aortopulmonary collaterals. LPA = left pulmonary artery; RPA = right pulmonary artery.

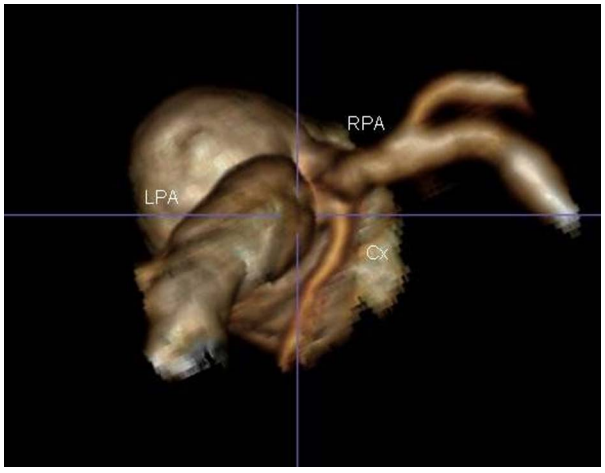

Figure 2.
A computerised tomography angiogram reconstruction – volume rendering, superior view. Cx = circumflex artery; LPA = left pulmonary artery; RPA = right pulmonary artery.

the natural origin of the left anterior descending branch. The post-operative course was uneventful.

On the follow-up ambulatory assessment, she was clinically well. The electrocardiogram and the left ventricular systolic function on echocardiogram were normal with resolution of the posterior wall dyskinesia. The estimated right ventricular systolic pressure from tricuspid regurgitation was normal.

Discussion

Scimitar syndrome is a rare congenital heart disease characterised by an anomalous connection of all or some of the pulmonary veins from the right lung to the inferior caval vein.[1] The characteristic appearance of the pulmonary veins on chest radiograms gives the condition its name.

The anomalous pulmonary venous anatomy, rather than being the sole determinant, is part of a spectrum of malformations including hypoplasia of the right lung and right pulmonary artery, bronchial anomalies, cardiac dextroposition, aortopulmonary collaterals, and pulmonary sequestration.[1] Clinical manifestations depend on the degree of aortopulmonary blood supply, pulmonary venous stenosis, and pulmonary hypertension, which is often secondary to these factors.[1]

Classic Scimitar syndrome is usually not associated with anomalies other than atrial septal defects but has been described less commonly with ventricular septal defects, patent arterial duct, tetralogy of Fallot, coarctation of the aorta, hypoplastic left heart syndrome, total anomalous pulmonary venous connection, cor triatriatum and sub-aortic stenosis.[2] Association with anomalous origins of the coronary arteries from the main pulmonary artery is very rare and has been described in two patients.[3,4] Boning et al described a case of Scimitar syndrome in a child associated with an anomalous origin of the left coronary artery from the main pulmonary artery. The only reported case of Scimitar syndrome and anomalous origin of the circumflex artery from the pulmonary artery was in an adult, a 27-year-old woman, with single coronary origin of the right coronary artery and the left anterior descending artery and the circumflex artery originating from the main pulmonary artery.[4] In this report, the patient underwent combined surgery for correction of the Scimitar vein and anomalous coronary but developed late pulmonary hypertension from failure of the coronary graft. Our patient is the only reported case of this association presenting during infancy.

Anomalous origin of the circumflex artery from the main pulmonary artery is itself a rare entity. Echocardiographic diagnosis is often difficult and symptoms may be masked by other conditions causing pulmonary hypertension, which maintain coronary perfusion and prevent "steal". For this reason, the diagnosis of anomalous coronary artery

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in our patient who had no evidence of ventricular ischaemia was made only using computerised tomography; angiography was requested later for further delineation of the anatomy.

The presence of a complex cardiac defect should not distract one from making a clear demonstration of the coronary anatomy. Surgical repair was challenging because the origin of the circumflex artery was just at the origin of the hypoplastic right pulmonary artery. The strategy of occluding the aortopulmonary collateral, waiting for the pulmonary hypertension to improve and then re-implanting the anomalous coronary artery contributed to a good outcome in this patient, as the presence of pulmonary hypertension would have increased the peri-operative risk. We elected not to perform the surgical correction of the Scimitar vein at the same time because there is a higher risk and rate of complications, including residual pulmonary vein stenosis, in patients operated during infancy. Furthermore, our patient was asymptomatic and did not have evidence of increased left-to-right shunt, which is the recommendation for surgical correction of Scimitar syndrome in our service and will guide further interventions in our patient.

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Supplementary material

For supplementary material referred to in this article, please visit http://dx.doi.org/10.1017/S1047951113000243

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