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Brief Report

Transcatheter correction of Scimitar syndrome: occlusion of abnormal pulmonary venous drainage and vascular supply in an infant

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Abstract Treatment of Scimitar syndrome is usually surgical; however, if there is "dual drainage" – that is, one to the inferior caval vein and the other to the left atrium – it is possible to successfully treat this anomaly via a less-invasive transcatheter approach. We report a case of Scimitar syndrome in a 21-month-old, male infant successfully treated with transcatheter embolisation.

Keywords: Scimitar syndrome; transcatheter correction; partial anomalous pulmonary venous connection

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SCIMITAR SYNDROME INCLUDES THE PRESENCE OF A partial anomalous pulmonary venous connection, and is associated with anomalous systemic arterial supply from the abdominal aorta to a part of the lungs.[1] Treatment is usually surgical with re-routing of abnormal pulmonary veins to the left atrium. If there is dual venous drainage, it may be possible to treat this anomaly via a transcatheter approach. We present a case of Scimitar syndrome where the presence of dual drainage was successfully treated using the transcatheter approach with vascular plugs.

Case report

A 21-month-old, male infant was admitted to a local hospital with fever and cough. Transthoracic echocardiography showed mildly enlarged right cardiac chambers and an abnormal flow pattern in the inferior caval vein. Cardiac CT revealed partially abnormal venous drainage to the inferior caval vein and an anomalous arterial blood supply originating from the aorta. The patient was then referred to our centre for cardiac catheterisation.

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Cardiac catheterisation demonstrated normal pulmonary artery pressures with a QP/QS ratio of 1.5. Angiographic levophase of the right pulmonary artery showed partial anomalous venous drainage via a vertical vein into the inferior caval vein together with unobstructed drainage to the left atrium (Fig 1a). Selective injection of the abnormal artery showed a feeding aortopulmonary artery arising from the descending aorta (Fig 1b). According to the angiographic findings, a decision was made to correct the anomaly through a transcatheter approach.

The procedure was performed under general anaesthesia: two venous accesses and one arterial access were achieved via the femoral route. The anomalous pulmonary vein was identified and cannulated. An exchange guide wire and a 12-mm Tyshack balloon were positioned into the anomalous (scimitar) vein, which was totally obstructed by the inflated balloon. Contrast was injected into the right pulmonary artery; angiography confirmed the non-obstructed decompressing vein to the left atrium (Fig 1c). Pulmonary arterial pressure and arterial saturation were monitored for 10 minutes, and they remained unchanged following vein occlusion. An 8-French, multipurpose guiding catheter was exchanged over an extra-stiff exchange wire, and a 14-mm Amplatzer™ vascular plug II (St. Jude Medical, Austin, TX, United States of America) was

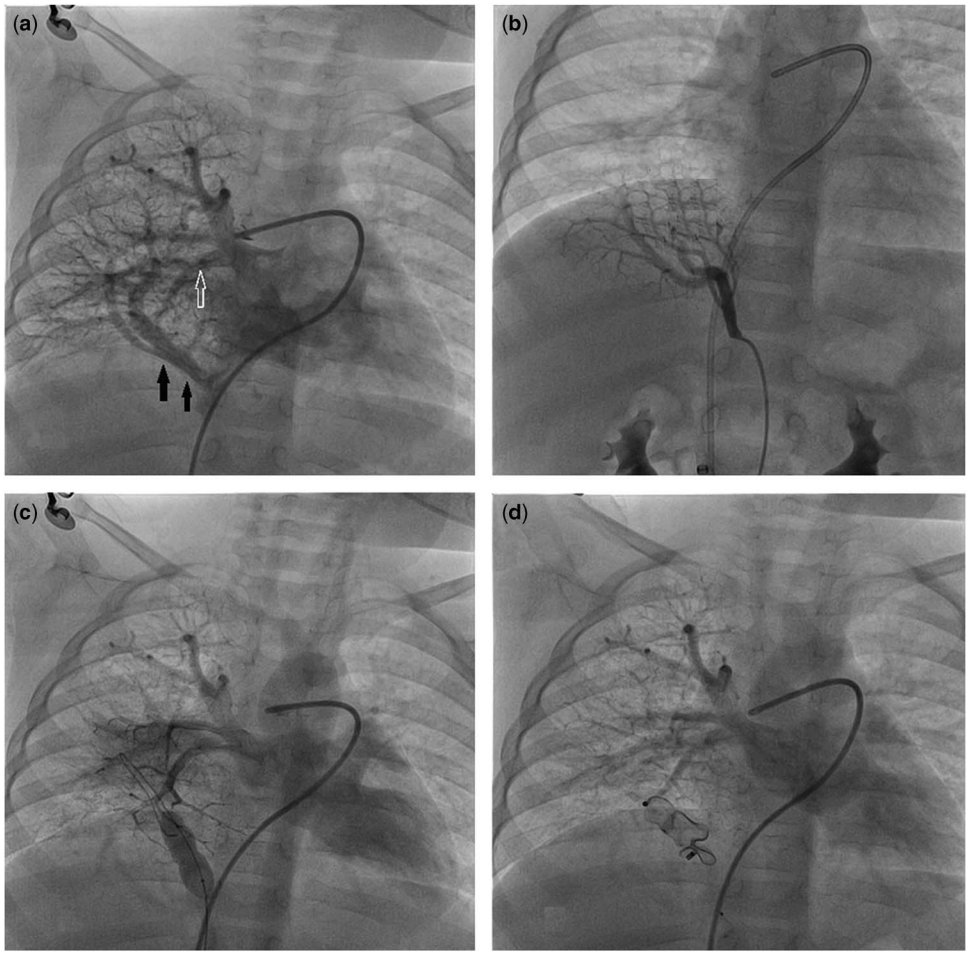

Figure 1.
*(**a**) Levophase of the right pulmonary artery following contrast injection showing a dual system of the pulmonary venous drainage of the right lung (white arrow shows normal return to the left atrium; black arrows show abnormal return via the Scimitar vein to the inferior caval vein). (**b**) Systemic arterial supply from the abdominal aorta to the right lower lung before occlusion. (**c**) Selective angiography of the right pulmonary artery, in the levophase, showing complete occlusion of the Scimitar vein by the balloon and normal pulmonary venous drainage of the right lung. (**d**) In the levophase of the right pulmonary artery, contrast injection showing complete occlusion of the Scimitar vein by plug and normal pulmonary venous drainage of the right lung to the left atrium.*

deployed at the stenotic segment producing good results. Repeated right pulmonary artery and venous contrast injections and imaging confirmed the appropriate position of the vascular plug and confirmed the patency of the decompressing veins to the left atrium. The device was released, and control pulmonary artery angiography showed an unobstructed pulmonary venous return to the left atrium (Fig 1d).

As a staged procedure, the aortopulmonary collateral was catheterised via the femoral artery with 5-F JR4 catheter. Following the diagnostic infusions and measurements, a 6-mm Amplatzer™ vascular plug 4 (St. Jude Medical) was placed in the distal segment of the feeding vessel. The device position was confirmed and then released. A control injection demonstrated almost complete vascular occlusion. The patient was discharged from the hospital on the next day, without any complications. No antiplatelet or anticoagulant therapies have been used thus far. The infant was asymptomatic at the 22-month follow-up.

Discussion

Partial anomalous pulmonary venous connection is one of the major components of Scimitar syndrome.[1] As in isolated partial anomalous pulmonary venous connection, there is only one abnormal pulmonary venous drainage in Scimitar syndrome; however, occasionally, the existence of a dual drainage system has been reported – one to an abnormal cardiac structure and one to the left atrium.[2–4] Similarly, dual drainage can also occur in Scimitar syndrome, which has been described in other situations of partial

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anomalous pulmonary venous connection.[5,6] The presence of dual drainage is a phenomenon that may lead to a less-invasive transcatheter treatment as an alternative to surgery as in our case.

The first step in evaluating appropriate patients for transcatheter occlusion is detecting the presence of alternative venous pathways to the left atrium. A detailed assessment of pulmonary venous drainage is mandatory to define the presence of dual drainage. An important consideration in transcatheter occlusion, when the Scimitar vein is fully occluded, is whether the remaining dual vein causes pulmonary vein stenosis or not. In 1998, Forbess et al[3] reported the first experience of assessment of the dual drainage via balloon occlusion of the Scimitar vein and the junction of the inferior caval vein. Sarquella-Brugada et al[5] and Wilson et al[6] suggested that monitoring pulmonary venous pressure during balloon occlusion of the Scimitar vein was a marker for unobstructed flow to the left atrium via the dual connections. Mas et al,[7] however, described occlusion of the Scimitar vein drainage, without balloon test occlusion, but advised checking the angiographic images before releasing the device. In our patient, instead of pulmonary venous pressure measurement, we only monitored the pulmonary arterial pressure and oxygen saturation during the balloon occlusion. More importantly, after observing the pattern of non-obstructive pulmonary venous flow to the left atrium, while the abnormal vein was totally occluded, we continued the procedure.

Singh et al[8] first reported the use of a vascular plug in obstruction of the Scimitar vein and inferior caval vein junction. Amplatzer™ vascular plugs, as used in this case, are well suited for occluding these anomalous veins. We recommend the use of an oversized device, by 30–50% of the diameter measured at catheterisation, as also recommended by Wilson et al.[6]

Following occlusion of the Scimitar vein, the occlusion of the feeding artery is simpler and can be performed using coils or plugs. For more than 20 years, transcatheter occlusion of the anomalous systemic arterial supply has been reported as an alternative to surgical ligation in symptomatic infants before cardiac surgery.[9] Furthermore, a benefit of transcatheter embolisation of aberrant vessels to the sequestered segments has been reported to include a reduction in recurrent respiratory infection.[10]

In conclusion, we report a case of Scimitar syndrome in a 21-month-old, male infant successfully corrected with transcatheter occlusion. As this report has demonstrated, the less-invasive approach of transcatheter occlusion may be appropriate if cardiac catheterisation demonstrates dual venous drainage.

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Conflicts of Interest

None.

Ethical Standards

The authors assert that this study complies with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008. This study was approved by the institutional committee of Cerrahpasa Medical School.

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