CASE REPORT

Scimitar syndrome with tetralogy of fallot and pulmonary atresia

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Abstract Scimitar syndrome is a rare variant of partial anomalous pulmonary venous connection. The association of Scimitar syndrome with another cardiac congenital anomaly such as tetralogy of Fallot with pulmonary atresia is extremely rare; we are reporting a successful combined treatment using transcatheter closure of major aorto-pulmonary collateral and a single-stage surgical correction in eighteen month old boy diagnosed as Scimitar syndrome with tetralogy of Fallot and pulmonary atresia.

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1. Case report

An eighteen month old boy was referred to our center due to cyanosis and shortness of breath. He was a product of full term normal vaginal delivery and the first child in his family. His birth weight was 3.5 kg. On admission his weight was 14 kg (95% centile) and height 83 cm (95% centile), oxygen saturation on room air 75–80%. The chest radiography showed mesocardia and curvilinear shadow in the right chest (Scimitar sign). Based on the clinical and chest radiography findings Scimitar syndrome was suspected. 2D echocardiogram revealed large sub aortic ventricular septal defect with outlet extension, overriding aorta, atretic pulmonary valve with confluent pulmonary artery branches, moderate size patent ductus arteriosus, bilateral superior vena cava and partial anomalous pulmonary venous drainage (PAPVD). The right pulmonary veins are draining to the inferior vena cava–right atrium (IVC–RA) junction (Fig. 1). In order to confirm the diagnosis Cardiac Computed Tomography and Angiography (CT angiogram) was performed, it confirmed that the right pulmonary veins are draining to the IVC–RA junction above the diaphragm, with two major aorto-pulmonary collaterals toward the lower lobe of the right lung (Fig. 2). Trans-catheter coil embolization of these major systemic collaterals was done successfully, under general anesthesia; the patient was prepped and draped as per protocol. The left femoral vein was accessed percutaneously using a Vygon 20 sheath. The right femoral artery was accessed using the modified Seldinger technique and a 4 French sheath was inserted over a guide wire. Angiography of the descending Aorta was performed which showed a patent left sided Aortic arch, confluent pulmonary arteries, and two major Aorto-pulmonary collaterals supplying the lower lobe of the right lung. A 6 mm Amplatzer vascular plug II was deployed in the lower collateral, and a 5 x 5 coil
embolized also in the lower collateral. A 3 × 3 coil was embolized in the upper collateral (Fig. 3). Then PA/VSD and PAP-VD were repaired surgically.

After longitudinal sternotomy; systemic heparinization and cardiopulmonary bypass were established. The cardioplegia solution was infused through the needle inserted at the aortic root and the heart was arrested. Wide right atriotomy was performed downward till the lateral wall of the IVC, autologous pericardial patch was inserted rerouting the pulmonary vein flow toward the atrial septal defect (ASD), keeping the IVC inflow free from any residual stenosis. Through the same atriotomy the ventricular septal defect (VSD) was closed by a Gortex patch. Longitudinal ventriculotomy was performed, right ventricular outflow tract (RVOT) muscle bands were resected and right ventricle to pulmonary artery conterga conduit size 16 mm was inserted. The lowest temperature was 30 °C. The patient was weaned uneventfully from cardiopulmonary bypass. Routine trans-esophageal Echocardiography (TEE) showed free IVC inflow and no obstruction at the level of the pulmonary veins. The child recovered well after surgery with normal saturation and was discharged home with satisfactory condition.

2. Discussion

Scimitar syndrome or congenital pulmonary venolobar syndrome is a rare entity but a well described constellation of cardiopulmonary anomalies. The term “Scimitar” was first used by Neill2,9 to describe the syndrome. This anomaly consists of partial or total anomalous venous connection of the right lung draining into the inferior vena cava or the azygos system, concomitant with lung hypoplasia and anomalous systemic arterial blood supply to the right lung rising from the abdominal aorta or its branches.1 These anomalous vessels are often visible on chest X-ray as a curvilinear shadow just above the right diaphragm resembling a Turkish sword or scimitar.10 The incidence of Scimitar syndrome was reported as 0.5–1% of CHD. The incidence of associated congenital cardiovascular abnormalities is about 36% in the pediatric age group and 75% among neonates. These abnormalities include atrial septal defect, ventricular septal defect, and coarctation of the aorta. Other congenital anomalies involving the aortic arch and abnormal relationship of the pulmonary arteries and lung abnormalities such as hypoplasia and horseshoe lung should be considered in the work out of Scimitar syndrome patients.3,7–9,11 Several literatures describe other rare associations of Scimitar syndrome with other cardiac abnormalities: Hakim et al. described two cases of Scimitar syndrome, one case was scimitar syndrome with Sub-Aortic membrane, and another where the scimitar vein showed tortuous and

Figure 1  Echo picture – sub-costal view showing sub-aortic ventricular septal defect (VSD) with aortic overriding. LA: Left atrium; LV: Left ventricle; RV: Right ventricle.

Figure 2  Angio CT scan showing anomalous pulmonary venous drainage of the right lung to the IVC–RA junction (red arrow).

Figure 3  Coil occlusion of the 2 abnormal collaterals supplying the right lung.
abnormal course of the pulmonary veins with normal drainage into the left atrium. Takeda reported two cases: in one case the scimitar vein entered both the inferior vena cava and the left atrium without any intracardiac shunt; in the other case the Scimitar vein showed a meandering course and then drained into the left atrium, so surgical intervention was not required. Bilateral scimitar syndrome with abnormal venous drainage to inferior vena cava has also been described.

Isolated partial anomalous pulmonary venous return to the inferior caval vein is also called incomplete scimitar syndrome. The typical scimitar sign was present in 57 of the 67 cases reviewed by Kiely et al. Association of pulmonary atresia, ventricular septal defect and Scimitar syndrome is a very rare combination. Our case had the classical characteristics of Scimitar syndrome with pulmonary atresia and ventricular septal defect. Pre-operative diagnostic cardiac catheterization and CT angiography have central role in the diagnosis of such variation especially when the pattern of pulmonary venous drainage is not clearly identified by echocardiography. After successful combined trans-catheter and surgical repair, the patient had smooth outcome and was discharged home with full recovery. One month later an echocardiography follow-up was performed showing no residual cardiac lesions and pulmonary vein flow pattern was normal. As the risk of baffle obstruction at follow-up is well reported we keep our patients on low dose aspirin therapy for prophylaxis.

We can conclude from this case study that CT angiogram and cardiac angio catheterization are the most appropriate diagnostic modalities to confirm the anatomy of this unusual variant of Scimitar syndrome. Combination of therapeutic angiography and surgery was feasible, and contributed to the success of this case management. Elimination of collateral flow, correcting the venous drainage and correcting intracardiac abnormality help in preventing the development of pulmonary hypertension and normalize the blood flow to the lung which make the hybrid approach ideal for short and long term results.

Conflict of interest

I disclose that there were no conflict of interest and no funding for this study and it was approved by the hospital ethics committee.

References