Simultaneous transcatheter closure of intralobar pulmonary sequestration and patent ductus arteriosus in a patient with infantile Scimitar syndrome

Scimitar sendromlu bir süt çocuğunda intralober pulmoner sekestrasyon arteri ve patent duktus arteriyozusun eş zamanlı kateter yoluyla kapatılması

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Summary– Scimitar syndrome is a rare disease associated with a right lung sequestration vascularised by arteries arising from the abdominal aorta and abnormal venous drainage into the inferior vena cava. The infantile form is generally presented with severe heart failure, pulmonary hypertension and respiratory distress. It may be associated with various intracardiac defects, including atrial septal defects, ventricular septal defects, patent ductus arteriosus or more complicated structural congenital heart defects. Here, we present a 2-month-old girl with Scimitar syndrome whose pulmonary arterial pressure decreased after transcatheter patent ductus arteriosus closure and embolization of the anomalous systemic arterial supply.

Özet– Scimitar sendromu, sağ akciğer sekestrasyonunun abdominal aortadan çıkan arterler tarafından kanlandırıldığı ve inferiyor vena kavaya anormal venöz drenaj ile ilişkili nadir bir hastalıktır. İnfantil formu genellikle ağır kalp yetersizliği, pulmoner hipertansiyon ve solunum güçlüğü ile karşımıza çıkar. Atriyal septal defekt, ventriküler septal defekt, patent duktus arteriyozus ya da karmaşık yapısal kalp hastalıkları gibi çeşitli kalp-içi patolojilerle birliktelik gösterebilmektedir. Bu yazıda, kateter yoluyla patent duktus arteriyozus kapatılması ve anormal sistemik arteryel kaynağın embolizasyonu ile pulmoner arter basıncında azalma görülen Scimitar sendromlu iki aylık bir kız hastayı sunduk.

Scimitar syndrome (SS) is a rare congenital disease characterized by hypoplasia of the right lung and right pulmonary artery, sequestration arteries from the aorta, and anomalous drainage of partial or total right side pulmonary veins into the inferior vena cava (IVC).[1] Only rarely are left side pulmonary veins involved in SS. The infantile form presents early in life with symptoms of tachypnea, chest infection, and heart failure due to pulmonary hypertension (PH) and other associated defects. Nearly half of cases have various intracardiac defects, including atrial septal defect (ASD), patent ductus arteriosus (PDA), ventricular septal defect (VSD), or more complicated structural congenital heart defects.[1,2]

Abbreviations:

ADO-II	*Amplatzer Duct Occluder-II*
ASD	*Atrial septal defect*
AVP-II	*Amplatzer Vascular Plug-II*
IVC	*Inferior vena cava*
PDA	*Patent ductus arteriosus*
PH	*Pulmonary hypertension*
SS	*Scimitar syndrome*
VSD	*Ventricular septal defect*

Most SS cases suffer from PH due to one or more of the following reasons: A large shunt from anomalous systemic arteries supplying the lower part of the right lung (vascular sequestration), an intracardiac shunt, or pulmonary vein stenosis. Embolization of anomalous systemic arteries by transcatheter

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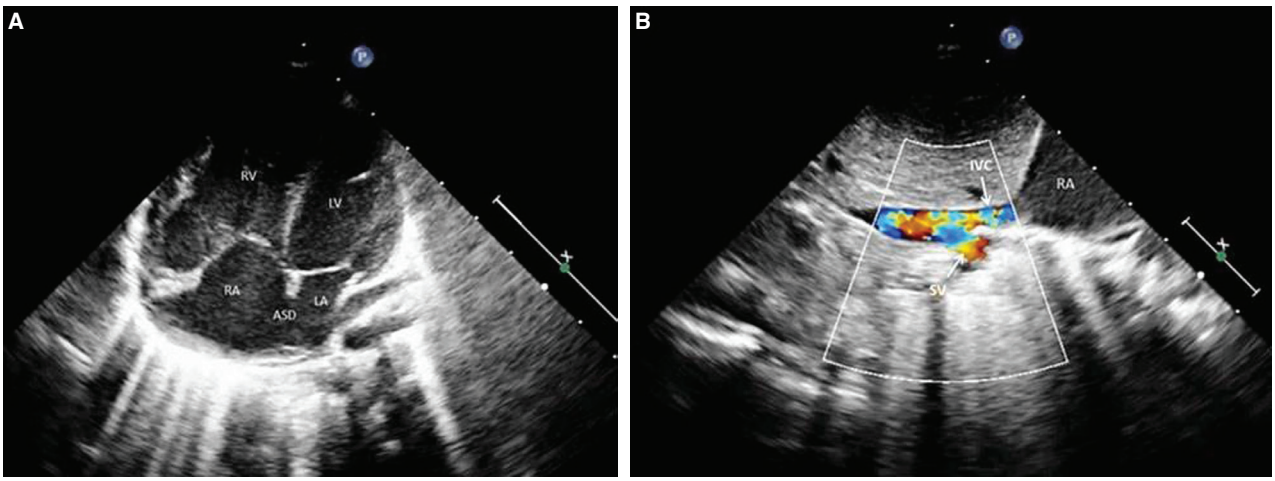

Figure 1. Transthoracic echocardiography showing enlarged right cardiac chambers **(A)**, and scimitar vein drainage to the inferior vena cava vein-right atrium junction **(B)**. ASD: Atrial septal defect; IVC: Inferior vena cava; LA: Left atrium; LV: Left ventricle; RA: Right atrium; RV: Right ventricle; SV: Scimitar vein.

interventions improves symptoms of cardiac failure and PH in symptomatic infants.[3]

Herein, we report an infant with SS whose clinical condition improved and pulmonary arterial pressure decreased after transcatheter embolization of anomalous systemic arterial supply and PDA closure.

CASE REPORT

A 2-month-old, 3.8 kg female infant was referred to our hospital for evaluation of a heart murmur. Physical examination revealed tachypnea with 65 breaths per minute, and a 3/6 grade systolic murmur at the left sternal border consistent with a pulmonic outflow murmur. Her transcutaneous oxygen saturation was 91% and laboratory values were within normal limits. A chest X-ray revealed mildly cardiomegaly. Transthoracic echocardiography showed significantly enlarged right cardiac chambers (Fig 1a), a 12-mm secundum ASD, small apical trabecular VSD, PDA, and partial abnormal venous drainage of the right pulmonary veins to the IVC (Fig 1b). An estimated systolic pulmonary artery pressure from tricuspid regurgitation was 60 mmHg. Computerized cardiac tomography angiography showed partial abnormal venous drainage to the IVC, systemic arterial supply, and a smaller right pulmonary artery compared to left pulmonary artery (Fig 2 a-c). Cardiac catheterization

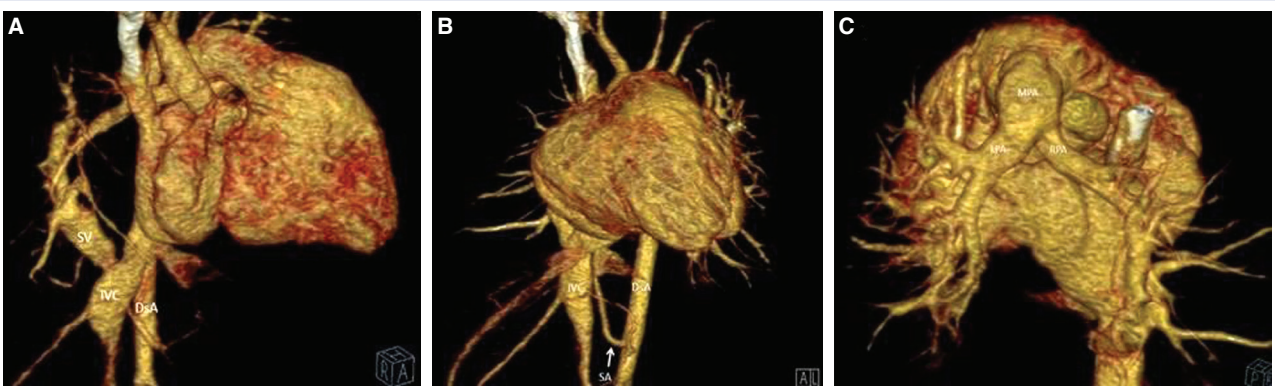

Figure 2. Computerized tomography angiography scan showing partial abnormal venous drainage of upper and lower right lung to the inferior vena cava vein-right atrium junction **(A)**, systemic arterial supply from the abdominal aorta to the right lower lung **(B)**, and small right pulmonary artery compared to left pulmonary artery **(C)**. DsA: Descending aorta; IVC: Inferior vena cava; LPA: Left pulmonary aorta; MPA: Mean pulmonary aorta; RPA: Right pulmonary aorta; SA: Sequestration artery; SV: Scimitar vein.

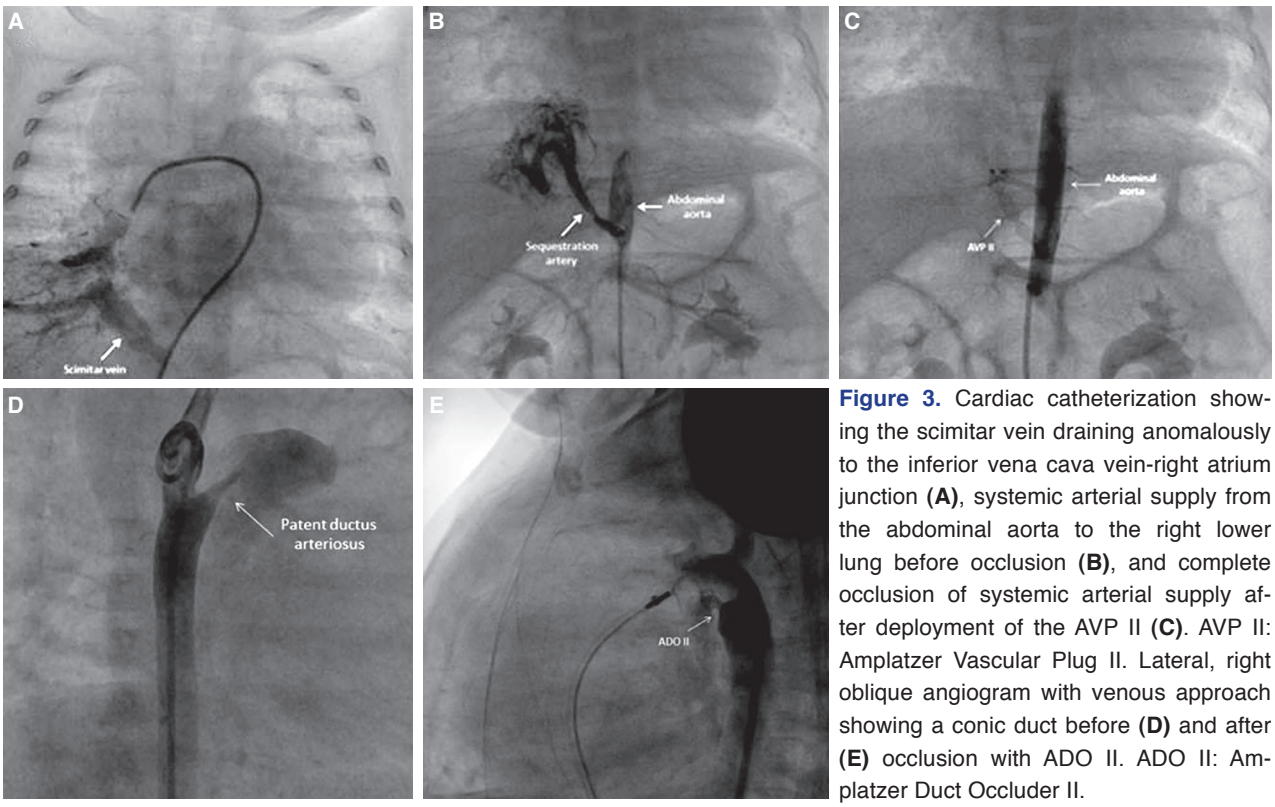

Figure 3. Cardiac catheterization showing the scimitar vein draining anomalously to the inferior vena cava vein-right atrium junction **(A)**, systemic arterial supply from the abdominal aorta to the right lower lung before occlusion **(B)**, and complete occlusion of systemic arterial supply after deployment of the AVP II **(C)**. AVP II: Amplatzer Vascular Plug II. Lateral, right oblique angiogram with venous approach showing a conic duct before **(D)** and after **(E)** occlusion with ADO II. ADO II: Amplatzer Duct Occluder II.

confirmed the partial anomalous pulmonary venous return of right-sided pulmonary veins into the IVC with no dual connection, sequestration artery, PDA, and PH (Fig 3a, b, d). Also, cardiac catheterization was performed to close the PDA and sequestration artery (of diameter 4.2 mm). Aortic and pulmonary artery pressures were measured as 80/42 (60) mmHg and 56/18 (35) mmHg respectively.

A 4-Fr Cook long sheath (Cook, Bloomington,IN) was introduced into the sequestration artery. Then, a 6-mm Amplatzer Vascular Plug-II (AVP-II) (AGA Medical, MN) was implanted (Fig 3c). Transcatheter closure of the PDA was performed by the antegrade approach, using a 4-Fr Cook long sheath. Initially, we deployed 3 mm x 6 mm Amplatzer Duct Occluder-II (ADO-II) (AGA Medical, MN) as the PDA measured 2.2 mm at the pulmonary side. However, the aortic disc protruded into the descending aorta, so we had to retrieve this device and deploy a 3 mm x 4 mm ADO-II (Fig 3e). Repeat angiography after the subsequent implantation showed complete occlusion of the sequestration and ductal artery. Pulmonary artery pressure decreased to 35/16 (22) mmHg. Ten days after the procedure, the patient's oxygen saturation was 98% with 32 breaths per minute. Right ventricle size and mean pulmonary artery pressure were within normal limits.

DISCUSSION

Scimitar syndrome is a rare congenital anomaly and presents in various forms. Due to anomalous systemic arterial supply, significant intra-cardiac shunt and pulmonary vein stenosis, infants generally present with severe symptoms and PH. Scimitar syndrome requires individualized treatment based on symptomatology and associated anatomy.[1]

Complete correction by surgery remains the gold standard therapy. However, a two-step procedure with a transcatheter approach to scimitar patients may be an alternative in selected cases. Embolization of the aberrant vessels as the first-stage intervention has shown clinical improvement and reduction of left-to-right shunt and pulmonary artery pressure.[3] Various kinds of devices, such as ductal coils,[4] AVP II,[5] AVP IV[6] etc., may be chosen for embolization of anomalous systemic arteries. We chose a 6-mm AVP II to

close the anomalous arterial supply to the sequestered segments.

Amplatzer Vascular Plugs are effective, safe and straightforward devices. The AVPs family offers different sizes to fit a wide range of vascular diameters. For paediatric applications, the major advantage is the reduced profile of the device, allowing small introducer sheath sizes. Amplatzer Vascular Plug II has been used in various low- and high-flow lesions including venous collaterals, aorto-pulmonary collaterals, modified Blalock Taussig shunts, porto-systemic connections.[7] In small infants, pulselessness is a common finding following introduction of large sheaths into the arteries. In our case, we used the AVP II through a small guide catheter (4Fr), which did not cause any arterial complication.

In scimitar patients, structural cardiac defects can contribute to PH, so occlusion of these defects can improve symptoms.[8] In our patient, we decided to close the PDA as it was one of the main reasons for the PH. Percutaneous closure of a PDA in infants can be efficiently done using the detachable coils for small ducts and ADO or AVPs for larger ducts. However, with the use of these devices, problems have occurred, including procedural failure, high incidence of residual shunts, aortic protrusion, left pulmonary artery stenosis, device embolization, hemolysis or arrhythmia.[9] In the presented patient, we had to extract the 3 mm x 6 mm ADO II due to aortic disc protrusion into the descending aorta, and deploy 3 mm x 4 mm ADO II, which is of shorter length. This device did not extend into the descending aorta.

In conclusion, in infantile SS, symptoms depend on the degree of shunting and severity of associated anomalies. Early intervention is necessary to prevent infections and progressive cardio-respiratory symptoms. Transcatheter closure of the anomalous systemic artery and associated anomalies simultaneously may improve symptoms of heart failure and PH in infants.

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