



Time-Saving and Successful Palliative Treatment for Scimitar Syndrome

Scimitar syndrome is a rare congenital cardiopulmonary anomaly characterized by the abnormal drainage of pulmonary veins into the inferior vena cava. It is often associated with additional anomalies, including an abnormal arterial supply from the descending aorta to the underdeveloped right lung and a rightward shift of the heart. In the infantile form of the disease, patients are at risk of early mortality due to respiratory distress, recurrent pneumonia, and heart failure. We present the effective interventional palliation of a patient with infantile-type Scimitar syndrome who had heart failure, was cyanotic, and required intubation due to respiratory distress.

Case - The patient, born with a weight of 3100 g at 38 weeks of gestation, was hospitalized in the neonatal intensive care unit due to neonatal pneumonia. The patient was transported to our clinic after eight days in the hospital since her medical condition showed no signs of improvement. Echocardiography of the patient showed a 5.6 mm right-left dominant atrial septal defect (ASD) with bidirectional shunt and a small secundum ASD in the high venosum region in the interatrial septum. The interventricular septum was intact. The right ventricle was extremely enlarged and hypertrophic. Deviation of the interventricular septum to the left during systole was observed (D-shaped left ventricle). Left ventricular functions were within normal limits. Outflow tracts were observed to be open. Grade 3 tricuspid valve regurgitation flow with a flow velocity of 3.8 m/sec was observed, and right ventricular systolic pressure was calculated as 65 mmHg due to tricuspid regurgitation. The vena cava inferior was wide, the right pulmonary veins merged posterior to the left atrium and opened into the inferior vena cava as a common venous structure, and a flow velocity of 1.7 m/sec was measured distal to the region. The left inferior and superior pulmonary veins drained into the left atrium. No venous structure or vertical vein opening to the left innominate vein was observed. The pulmonary artery was observed to be wide. Partial anomalous pulmonary venous return, severe pulmonary hypertension, and secundum ASD diagnoses were made. Computed tomography angiography (CTA) was performed to clarify the diagnosis and define the pathology more clearly with 3D modeling. CT angiography showed that the right pulmonary veins merged posterior to the left atrium and opened into the inferior vena cava via a common venous structure (Scimitar vein). The Scimitar vein measured 4.4 mm in diameter at the level where it merged with the IVC and was evaluated as stenotic. An arterial structure with a diameter of approximately 3 mm arises from 2 mm above the celiac trunk and supplies the lower lobe of the right lung (Figure 1). The findings of the CTA are compatible with hypoplastic lung syndrome-Scimitar syndrome. The patient, who had been intubated and was receiving inotropic support in the intensive care unit, had an approximately 70% oxygen saturation level. The patient's right heart border seems to be blurred on the chest radiograph, and an abnormal draining vein is seen as a tubular structure parallel to the right heart border that resembles the tip of a Turkish sword. The patient's angiography revealed a sequestration artery that was 2 mm above the celiac artery and supplied blood to the lower lobe of the right lung. It was observed that there were large tortoise dysmorphic vascular structures in the lower lobe of the right lung, and the venous return phase was towards the inferior vena cava. A decision was made to occlude the sequestration artery using the Amplatzer Piccolo Occluder® (5x6 mm), and the procedure was carried out successfully. Six days after the closure of the sequestration artery, the patient, no longer in need of intensive care, was moved to the pediatric department. Following a one-month observation period there, it was determined

Akif Kavgacı¹

Sultan Göncü²

Berna Uçan³

Fatma Zehra Öztekin Çelebi⁴

Utku Arman Örün¹

¹Department of Pediatric Cardiology, Ankara Etilik City Hospital, Ankara, Türkiye

²Department of Pediatric Intensive Care Unit, Ankara Etilik City Hospital, Ankara, Türkiye

³Department of Pediatric Radiology, Ankara Etilik City Hospital, Ankara, Türkiye

⁴Department of Pediatrics, Ankara Etilik City Hospital, Ankara, Türkiye

Corresponding author:

Akif Kavgacı

✉ akifkavgaci@gmail.com

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Figure 1. Sequestered arteries supplying the right lung in the CTA.

that outpatient follow-up was necessary to assess the need for surgical intervention, and the patient was discharged in a healed condition. Informed consent was obtained from the patient's parents.

Scimitar syndrome is a rare congenital cardiopulmonary anomaly with an incidence of approximately 1-3/100,000 live births (1). This syndrome is characterized by abnormal pulmonary venous drainage to the inferior vena cava and is frequently accompanied by anomalies, including abnormal arterial supply from the descending aorta to the right hypoplastic lung and dextroposition of the heart (2). The hypoplastic right lung receives its blood supply from systemic arteries, particularly those originating from the thoracic or abdominal aorta. This systemic blood supply is often directed to the lower lobes of the lung (3). Scimitar syndrome can be categorized into three main types. The first is the adult form, which typically presents as asymptomatic during infancy. The second is the infantile form, which manifests with symptoms and pulmonary hypertension. The third form is associated with congenital cardiac defects (4). Patients with the infantile form of Scimitar syndrome typically exhibit severe and life-threatening symptoms, including pulmonary hypertension and indications of congestive heart failure. These patients often demand an immediate and coordinated multidisciplinary approach to medical care.

The treatment for Scimitar syndrome primarily involves the closure of the anomalous systemic arteries that supply the right lung with a device in the catheterization room. Additionally, surgical correction is required to address the abnormal pulmonary venous drainage, either through tunneling or by direct re-implantation into the left atrium (5). Catheterization is a valuable diagnostic procedure that offers precise assessments of various aspects related to Scimitar syndrome. These assessments include the evaluation of abnor-

mal pulmonary venous drainage, measurement of pulmonary artery pressure, determination of the volume load on the pulmonary circulation, identification of potential obstructions in the pulmonary veins, examination of the hemodynamics of abnormal systemic arteries, and the detection of any associated structural cardiac abnormalities. In patients with Scimitar syndrome, catheterization is also used for interventional procedures, which may involve embolizing abnormal systemic arteries using coils or devices, as well as the application of balloons or stents to address any stenosis. Embolization of the large abnormal systemic arteries is a beneficial procedure in cases of Scimitar syndrome, as these arteries can otherwise contribute to early congestive heart failure and recurrent lung infections. This intervention helps alleviate symptoms by decreasing pulmonary artery pressure. In cases of Scimitar syndrome where it is accompanied by other cardiac malformations, preoperative interventional embolization of the abnormal systemic arteries can be instrumental in preventing "perfusion lung" (5).

Scimitar syndrome, especially in its infantile form, may manifest with symptoms such as cyanosis, respiratory distress, tachypnea, recurrent pneumonia, and heart failure. Timely recognition of the disease and the interventional closure of systemic arteries supplying the affected lung lobe, when combined with appropriate medical management, can substantially enhance patient survival until complete surgical correction is achieved, thereby assisting this patient group in reaching adulthood.

Informed Consent: Written informed consent was obtained from the patients parents for the publication of the case report and the accompanying images.

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