Acta Cardiol Sin 2018;34:440–442
Letter to the Editor
doi: 10.6515/ACS.201809_34(5).20160611B

Scimitar Syndrome with Partial Left Pulmonary Artery Sling in a Neonate

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Key Words: Bronchial stenosis • Infancy • Pulmonary artery sling • Scimitar syndrome

INTRODUCTION

Scimitar syndrome is a rare condition including right lung hypoplasia, anomalous pulmonary venous drainage, dextroposition of the heart, and hypoplastic right pulmonary artery (RPA). The infantile form is sometimes associated with pulmonary hypertension, cardiac failure, and a poorer prognosis. We present a 1-day-old girl with a rare combination of partial left pulmonary artery (LPA) sling and scimitar syndrome, which was suspected in prenatal sonography and confirmed by echocardiography, computed tomography and cardiac catheterization. The patient received re-implantation of the aberrant left lower pulmonary artery and patch augmentation of the main-left upper pulmonary arteries at 4 months of age. Another operation was performed at 7 months of age with patch augmentation for stenosis of the left lower pulmonary artery and left pulmonary vein elevation to relieve its clenching of the left lower bronchus, with improvement of respiratory distress.

CASE REPORT

A 1-day-old female full-term neonate had been suspected of having scimitar syndrome at the gestational age of 23 weeks by prenatal sonography with findings of hypoplastic right lung and dextroposition of the heart. The baby was delivered by cesarean section due to prior cesarean section, with a neonatologist on standby. Her birth weight was 3680 gm, with Apgar scores of 9 and 10 at one and five minutes, respectively. She presented with severe respiratory distress one day after birth, and chest radiograph (CXR) showed dextroposition of the heart and lateral hemivertebra at the 12th thoracic vertebra (T12) level (Figure 1A). Echocardiography showed dextroposition with situs solitus, and some collateral arteries arising from the abdominal aorta. The right pulmonary veins could not be clearly traced. There was a normal bifurcation of the main pulmonary artery (MPA). However, the proximal RPA was seen turning leftward (Figure 1B). Computed tomography (CT) at three-days-old showed that MPA bifurcated into a dilated proximal RPA and a mildly hypoplastic LPA. After a short normal course of proximal RPA, it turned leftward and curved behind the trachea, becoming a left sided RPA (left-RPA). The left-RPA (aberrant left lower pulmonary artery, LLPA) and the original LPA (left upper pulmonary artery, LUPA) clenched the trachea to form a vascular ring (Figure 1C). With levophase, the right pulmonary veins confluenced into a scimitar vein, which subsequently drained to the inferior vena cava (IVC) (Figure 1D). Fiberoptic bronchoscopy demonstrated mild narrowing of the left lower bronchus. The pattern of bronchial branching was normal, and B-type natriuretic peptide level was 204 pg/ml.

Cardiac catheterization when the patient was 5-days-old showed an extremely hypoplastic distal RPA supplying the hypoplastic right lung. The LPA supplied the left upper lobe, and the left-RPA irrigated the left inferior lobe and part of the left upper lobe (Figure 2A). Anomalous drainage of the right lower pulmonary veins

Received: October 2, 2015 Accepted: June 11, 2016
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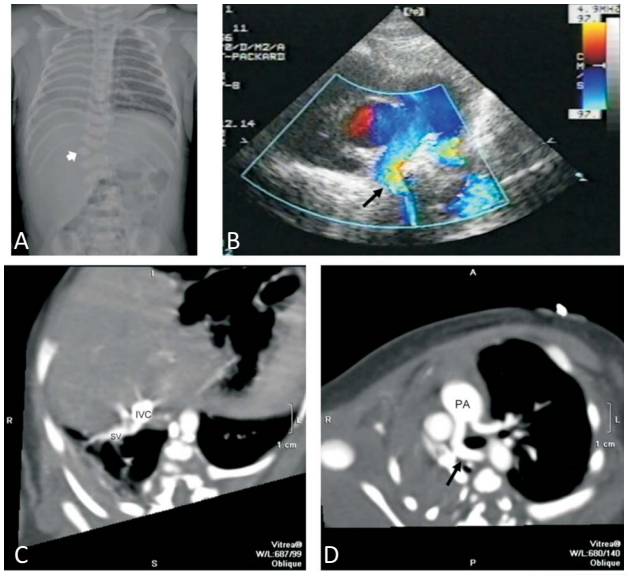

Figure 1. *(A) Chest radiograph. (arrow-head: hemivertebra). (B) Echocardiography in parasternal short axis view. (arrows: partial left pulmonary artery sling). (C) Computed tomography: SV (scimitar vein), IVC (inferior vena cava). (D) Computed tomography showed partial left pulmonary artery sling (arrow).*

to the IVC was confirmed by cine angiography. Contrast injection to the descending aorta revealed several collateral arteries supplying the right lower lobe (Figure 2B). Hemodynamic data showed a left-to-right shunt with a Qp/Qs value of 1.36, and elevated MPA pressure (78/39 mmHg, mean 53 mmHg).

The patient was discharged at 1-month-old with support of oxygen and nasal continuous positive airway pressure ventilation at home. As progressive dyspnea developed, she was admitted to the pediatric intensive care unit at 4 months of age; at that time, she was unable to be weaned from the ventilator. Subsequent cardiac CT and echocardiography imaging showed junctional stenosis at LUPA to pulmonary trunk connection. The patient underwent surgery for relief of the partial pulmonary artery sling at 4 months of age, with re-implantation of the aberrant LLPA, and MPA-LUPA patch augmentation. Unfortunately, the patient suffered from post-surgical recurrent respiratory tract infections and dyspnea, and remained difficult to wean from a ventilator. After follow-up echocardiography showed severe LLPA stenosis, the patient underwent further surgery for pulmonary artery plasty with patch augmentation and left pulmonary vein elevation (for relieving the clenching of the left lower bronchus by left pulmonary vein

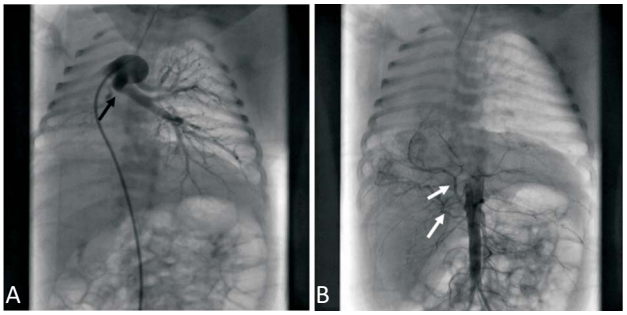

Figure 2. *(A) Pulmonary artery angiogram showed partial left pulmonary artery sling (arrow). (B) Aortogram demonstrated several collateral arteries (white-arrows) supplying the right lower lobe of the lung.*

and LLPA) at age of 7 months. However, after surgery it was observed that this patient showed some improvement of her respiratory distress.

DISCUSSION

Scimitar syndrome is an extremely rare disease with an incidence of 0.057% among the population presenting with congenital heart in an Asian country,[1] and predominantly affects females.[2] As noted in previous reports, 19-31% of patients with scimitar syndrome have associated cardiac anomalies.[3] The most common associated cardiac anomalies noted in previous literature are atrial septal defect (80%), followed by patent ductus arteriosus (75%), ventricular septal defect (30%), and pulmonary vein stenosis (20%).[2] In our patient, she had a unique combination of scimitar syndrome, left partial pulmonary sling, left bronchial stenosis and hemivertebra.

The classic scimitar syndrome on CXR is a curvilinear shadow adjacent to the right heart border, resembling a curved Turkish sword. However, in our case, CXR did not show such a typical appearance. This may be attributable to the patient's neonatal age, extreme dextroposition of the heart, and vascular congestion of the right lung, which can mask the venous drainage of the right lung in CXR.

Infants presenting with scimitar syndrome often exhibit severe heart failure and pulmonary hypertension, and have a large left-to-right shunt and poor prognosis.[3] Infantile scimitar syndrome may exist concurrent with a high incidence of pulmonary vein obstruction. In such cases, pulmonary vein surgical intervention is required. Because this patient developed severe dyspnea 1 day af-

ter birth, emergency cardiac catheterization was performed for evaluation of congestive breast failure due to left-to-right shunt (Qp/Qs), pulmonary hypertension and aortopulmonary collaterals. Infantile scimitar syndrome often presents with systemic collateral arteries to supply the RPA.[4-6] If the patient has pulmonary vein obstruction, the collateral arteries will aggravate pulmonary hypertension. However, embolization with a coil or Amplatzer vascular plug of the collateral artery can relieve pulmonary hypertension.[5-7] The indications for repair (reimplantation of the pulmonary venous drainage to the left side of the heart with/without coil occlusion of anomalous systemic arteries) are pulmonary hypertension owing to a significant left-to-right shunt (Qp/Qs > 2:1) as determined by cardiac catheterization and recurrent pulmonary infections.[4-6] Our patient had a small left-to-right shunt. Ultimately, there was no pulmonary vein stenosis found, and coil occlusion was not performed during cardiac catheterization.

Partial LPA sling is also an extremely rare condition and can be a cause of airway deterioration. To correct the airway problems, the patient received a redirecting operation of the partial pulmonary artery sling at 4 months of age, with re-implantation of LLPA and MPA-LUPA augmentation. At 7 months, pulmonary artery plasty with patch augmentation for severe LLPA stenosis and left pulmonary vein elevation to relieve the clenching of the left lower bronchus by the left pulmonary vein and LLPA were performed. The respiratory distress of this patient improved after surgery.

In conclusion, we have herein presented a unique case of a rare combination of partial left pulmonary sling and scimitar syndrome, which was not previously reported. Moreover, the case presented here emphasizes that pulmonary artery anomalies can coexist with scimitar syndrome and should be carefully surveyed using CT and cardiac catheterization.

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