Case report - Congenital

Atypical infantile form of scimitar syndrome with bronchomalacia

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Abstract

A male infant presenting with severe heart failure and respiratory distress was diagnosed with a hypoplastic right lung, scimitar syndrome with pulmonary sequestration and multiple anomalous systemic arteries, left bronchomalacia, a large atrial septal defect and coarctation of the aorta. The infant underwent a successful combined treatment of surgical and transcatheter intervention, including coil embolization and endobronchial stenting.

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The infantile form of scimitar syndrome generally presents with severe heart failure, pulmonary hypertension and respiratory distress. In these infants, complex associated anomalies, including hypoplastic right pulmonary artery and lung, still prompt a poor prognosis despite aggressive management. We report a successful combined-therapy treatment of surgical and transcatheter intervention in an infant with scimitar syndrome and bronchomalacia.

1. Case report

A male infant was presented with severe heart failure and respiratory distress the day he was born and was diagnosed with a right diaphragmatic hernia, an extremely hypoplastic right pulmonary artery and right lung, scimitar syndrome with pulmonary sequestration, a large atrial septal defect and coarctation of the aorta. Cardiac catheterization demonstrated severe pulmonary hypertension with a large left-to-right shunt (Qp/Qs = 4.1). Anomalous drainage of the right lower pulmonary veins to the inferior vena cava (IVC) was confirmed by cine angiography. An aortogram revealed multiple anomalous systemic arteries from the descending aorta supplying the right lower lobe (Fig. 1A). An extremely large systemic artery originating from the abdominal aorta also had communication with the portal vein, which caused portal hypertension. On the 14th day after birth, transcatheter coil embolization of the anomalous systemic arteries was performed, resulting in a dramatic lessening of patient’s symptoms and left-to-right shunt (Qp/Qs = 1.9) (Fig. 1B).

At 35 days, the infant underwent cardiopulmonary bypass and profound deep hypothermia for primary repair of the coarctation and the partial anomalous pulmonary venous return (PAPVR). Aortic cannulation through an expanded polytetrafluoroethylene graft sutured on the innominate artery allowed us to clamp the proximal arch. Following the removal of the ductal tissue and narrowed portion of the isthmus, we performed extended end-to-end anastomosis between the aortic arch and the descending aorta. The IVC cannula was removed, and repair of the PAPVR was accomplished by baffling the anomalous vein from its entry into the IVC through the right atrium to the left atrium by way of a large atrial septal defect using an autologous pericardium. The patient had been weaned easily from bypass, but during the weaning from the respiratory support, the patient experienced severe dyspnea. Post-operative echocardiography did not show any significant re-coarctation or any stenosis of either the anomalous vein or the IVC. A postoperative computed tomographic scan and fiberoptic bronchoscopy demonstrated the obstruction of the left main bronchus. Aortography suggested that the bronchial stenosis was being compressed by the reconstructed...
aortic arch. Two months later, after the primary repair, an exploratory thoracotomy was performed on extracorporeal membranous oxygenation. Although the compression by the aortic arch did not seem severe, we freed the stenotic bronchus from the surrounding tissue. The respiratory distress improved temporarily but gradually became severe due to the left bronchomalacia. Therefore, at 5 months of age and weighing 6 kg, the patient underwent endobronchial stenting with a Cordis Palmaz stent, P 2007E (7.0 mm × 17.8 mm, Johnson & Johnson) (Fig. 2). Although stent replacement had improved the dyspnea dramatically, there was a gradual re-stenosis of the left bronchus due to the formation of granulation. Following the balloon dilatation for relief of the stenosis, corticosteroid and epinephrine were administered into the left bronchus several times. As the granuloma gradually disappeared, the infant was weaned from respiratory support and was discharged from our institute.

2. Comment

Infants presenting with scimitar syndrome have severe symptoms and pulmonary hypertension [1,2]. Anomalous systemic arterial supply, significant intra-cardiac shunt and pulmonary vein stenosis are significant causes of this deterioration. Other cardiovascular malformations such as coarctation or reduction of the pulmonary vascular bed in associated diaphragmatic hernia [3], as observed in our case, are also main causes of pulmonary hypertension [4]. Ligation or embolization of the anomalous artery produces dramatic improvement from cardiac failure in infants without significant associated cardiovascular anomalies or pulmonary vein stenosis [5]. Embolization of the anomalous systemic arteries have been performed using detachable balloons, coils or tissue adhesive [2]. Coil occlusion of anomalous systemic arteries can improve symptoms of heart failure and pulmonary hypertension in infants and may bring about good surgical results for this disease. Gao et al. [2] reported a case of scimitar syndrome with Shone’ syndrome, pulmonary sequestration and right diaphragmatic hernia. In such a case, reduction of the pulmonary vascular bed or anomalous systemic arteries causes pulmonary hypertension. One of the main causes of the congestive heart failure in our case was an excessive left-to-right shunt through these abnormal arteries, which supplied the right lower sequestration area and right upper lobe of the liver. Embolization of these arteries was very effective in the short term, with no presentation of pulmonary infarction. Late evaluation after intra-cardiac repair showed persistent pulmonary hypertension. It seemed that reduction of the pulmonary vascular beds causes pulmonary hypertension.
We considered performing a correction of the liver herniation for right lung development, however, this procedure has been known to cause secondary kinking of the IVC or hepatic vein. Although occlusion of the systemic arterial collateral might be part of the surgical strategy, the significant associated cardiac anomaly should be repaired or corrected.

Postoperative pulmonary venous obstruction is prevalent, especially in small infants [6]. A long baffle may shrink, causing occlusion of the intra-atrial rout, or the orifice of the anomalous pulmonary vein itself may be stenotic. Therefore, the translocation of the anomalous vein to the right atrial lateral wall is an alternative method [1,7]. As the orifice of the anomalous vein was closed to the atrial septal defect in our patient, a long baffle was not necessary. Although repair of the anomalous venous return and ligation of collaterals is generally recommended, right pneumonectomy (either as a primary treatment or if a previous repair has failed) has had similar early and late results in spite of its potential long-term risks in infants [1].

Although left bronchomalacia is thought to be a secondary condition caused by inadequate aortic arch reconstruction, in our patient, the aortic arch was found not to be involved in the left bronchial compression, suggesting that it was not iatrogenic. Hsieh et al. [8] reported a case of scimitar syndrome where stenosis of the main stem of the left bronchus was also considered non-iatrogenic and which, along with an emphysematous change of the left lung, shifted the heart to the right side and compressed the right lung. The surgical management of bronchomalacia is challenging, although aortopexy and bronchopexy have been effective in select patients. Recently, expandable balloon stents (Palmaz) have been inserted into infants to relieve major airway obstructions [9,10]. Stenting in select infants has been effective in relieving bronchomalacia, however, granulation tissue has often developed over the stents, which are often removed by scraping or balloon compression [8]. The repeated administration of corticosteroid and/or epinephrine into the granuloma was also effective in our case.

We have herein reported a successful combined-therapy treatment of surgical and transcatheter intervention in an infant with scimitar syndrome and bronchomalacia.

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References