Right mainstem bronchial atresia successfully corrected with slide tracheobronchoplasty

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Abstract

Mainstem bronchial atresia (MBA) is a rare and fatal entity with no survivors reported to date. We describe a neonate born at 36 weeks gestational age (GA) with right MBA who underwent successful slide tracheobronchoplasty at 32 days of life. It is theorized that during fetal life a transient fistulous connection developed, allowing right lung decompression and left lung development.

Introduction

Bronchial atresia is a rare condition, typically affecting the subsegmental bronchi[1], and often noted on prenatal ultrasounds as a hyperexpanded fluid filled lung segment. The more proximal the atresia, the more severe and life-threatening the presentation. Only ten cases of mainstem bronchial atresia (MBA) have been reported to date, with universally poor outcomes[2-6]. Unlike peripheral bronchial atresia, MBA presents a unique challenge with expansion of the involved lung, mediastinal compression and contralateral shift, hydrops and fetal or infant death. This represents a form of unilateral Congenital High Airway Obstructive Syndrome (CHAOS), with laryngeal or tracheal atresia causing bilateral lung hyperexpansion and diaphragmatic eversion[7].

Case Report

A 26-year-old female was diagnosed at 22 weeks gestational age (GA) with possible fetal congenital high airway obstruction syndrome (CHAOS). Fetal MRI at 25 weeks GA revealed overexpansion of the right lung and suggested obstruction of the right mainstem bronchus (MSB) and ascites with polyhydramnios (Figure 1A). Fetal echocardiogram was normal. Fetal MRI at 33 weeks GA found decompression of the lung and resolution of hydrops (Figure 1B).

Due to fetal distress at 36+1 weeks GA, a cesarean section was performed. The male infant (1940 grams)had APGAR scores of 7/7 and required intubation on placental support. He demonstrated asymmetric chest wall movement and had absent breath sounds on the right. Chest x-ray showed complete opacification of the right lung (shown in Figure 2A). Echocardiogram revealed suprasystemic right ventricular pressure (RVP) with normal biventricular function. The infant was stabilized with inhaled epoprostenol, inhaled nitric oxide, intravenous epinephrine and intravenous phenylephrine.

At day of life 13, bronchoscopy confirmed complete stenosis of the right MSB just distal to the carina with normal anatomy otherwise including the left MSB (Figure 3A and 3B). Following regression of the ductus arteriosus, the infant had a long-segment left pulmonary artery (LPA) stenosis. Angiographically, the distal left lung vasculature appeared normal but the right side appeared dilated and tortuous. After angioplasty,

the proximal LPA peak gradient improved (37 mmHg to 23 mmHg). Inotropic support was weaned off in days and the inhaled pulmonary vasodilatory agents were transitioned to enteral sildenafil.

MRI of the chest (Figure 1C) at 19 days of life revealed a normal bronchial branching pattern distal to the bronchial atresia and a substantial amount of right lung with normal appearing vasculature. Phase contrast illustrated a flow differential of 10% and 90% effective flow to the right and left lung, respectively.

The infant was extubated to non-invasive ventilatory support at 22 days of life (Figure 2B). Serial echocardiography illustrated worsening RVP due to LPA stenosis from progressive rightward mediastinal shift. Although the infant appeared clinically stable, his condition was thought to be tenuous and operative intervention was pursued. At 32 days of life (2,410 grams) the infant underwent a right slide tracheobronchoplasty on Extracorporeal Membrane Oxygenation (ECMO) support.

Through a median sternotomy and wide opening of the pleural spaces and pericardium in the midline,suspension of the rightward pericardium allowed direct right atrial and aortic cannulation. A 5 mm fibrous stalk was present connecting a short stub of the proximal MSB with the more distal MSB. This was transected distally and mucus was cleared from the right lung. Proximally, a wedge was removed from the trachea at the stump of the right MSB, and the incision carried superiorly creating a V shape opening. Ensuring similar circumferences, the distal MSB was then sutured to the trachea using a double armed 6-0 PDS, posteriorly to anteriorly. A leak test was performed and a fibrin sealant was applied around the anastomosis.

Postoperative chest x-ray showed right lung expansion (Figure 2C). On postoperative echocardiogram the infant had mild LPA stenosis with a peak gradient of ˜24 mmHg and normalization of RVP estimates. Sildenafil was discontinued. He was extubated on post-operative day five to continuous positive airway pressure (CPAP). Late post-operative bronchoscopic evaluation was performed 2 months post slide tracheobronchoplasty, and showed a well healed widely patent anastomosis (Figure 4). At the time of this report, the infant is discharged without respiratory support and thriving at six months of age.

Discussion

As reported in the literature, all cases but one have been right sided. Treatments have included antenatal steroids, fetal pneumonectomy, fetal thoracoamniotic shunt placement, observation and elective termination[2-6]. Only three of the ten reported cases survived to delivery but none survived past day one of life. One had undergone fetal thoracoamniotic shunt at 23 weeks GA, was delivered at 25 weeks GA[3]. Another had a pneumonectomy at 21 weeks GA and delivered at 24 weeks GA[2]. The third underwent fetal pneumonectomy at 26 weeks GA, and was delivered at 32 weeks GA[5].

In all cases, the atretic MSB caused hyperexpansion of the ipsilateral lung, and compression of the contralateral lung in utero. Although fetal pneumonectomy theoretically allows the uninvolved lung time to grow and compensate, this did not occur likely secondary to prematurity. Here, the right lung distal to the atretic MSB was hyperexpanded but later collapsed (Figure 1A and 1B), with evidence of retained secretions and a normal bronchial branching pattern, allowing the left lung to function, albeit with significant mediastinal shift and consequent hemodynamic changes. We hypothesize that atresia at the mainstem level initially caused significant hyperexpansion on the right side, with mediastinal shift to the left, and a consequent developmental abnormality of the (short) LPA. At some point, the elevated pressure of lung fluid in the right bronchial tree presumably resulted in the formation of a fistulous connection, allowing decompression of the hyperexpanded right lung. The small fistula would have then collapsed under lower pressure and closed later in the 3rd trimester when the fetal lungs are not producing high volumes of fluid, leaving behind a 5 mm long fibrous attachment between the trachea and MSB. Postnatally, ventilation and expansion of the left lung further collapsed the right lung with worsening mediastinal shift. This exacerbated the stretching of the developmentally short LPA, with consequent narrowing of the stretched vessel and elevation of RVP. Slide tracheobronchoplasty allowed re-expansion of the right lung with mediastinal normalization, lessened the stretch and narrowing of the LPA, lowered the vascular resistance in the newly aerated lungs, with normalization of the RVP.

Slide tracheobronchoplasty for isolated bronchial pathology is a rare operation. The absence of bronchial cartilage causing a ball-valve effect and obstruction of the right MSB is the usual indication. This is a rare case of MBA successfully treated with slide tracheobronchoplasty following spontaneous resolution of fetal hydrops. Our management and surgical approach preserved his entire right lung and maximized the possibility of a long-term favorable outcome without the complications associated with pneumonectomy.

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