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Mainstem bronchial atresia: a lethal anomaly amenable to fetal surgical treatment

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ABSTRACT

Purpose: The purpose of this study was to review the unique imaging characteristics, prenatal course, and outcomes for fetuses with mainstem bronchial atresia (MBA).
Methods: The records of all patients referred for a fetal lung malformation from 2001 to 2012 and the medical literature were reviewed to identify cases of MBA.
Results: Of 129 fetuses evaluated, 3 were diagnosed prenatally with right-sided MBA. The first had a CCAM-volume ratio (CVR) of 9, hydrops, mirror syndrome, and preterm delivery of a nonviable fetus. The second (CVR 2.6) had ascites, preterm delivery at 34-weeks, and neonatal demise. The third fetus (CVR 5.7) presented with hydrops at 21-weeks, prompting fetal pneumonectomy. Postoperatively, hydrops resolved, and the contralateral lung grew dramatically, but preterm delivery occurred 3 weeks later. Ventilation could not be sustained, and the infant died. Four similar cases of MBA were in the literature, all right-sided. Two fetuses with hydrops delivered at 25-weeks and died immediately. One pregnancy was terminated. One fetus underwent pneumonectomy at 24-weeks but died intraoperatively.
Conclusion: MBA is a rare and lethal lesion that must be distinguished from other right-sided lung masses. Fetal pneumonectomy can be performed with resolution of hydrops and compensatory contralateral lung growth, but remains limited by complications of preterm birth.

Mainstem bronchial atresia (MBA) is a rare and likely under-recognized, congenital malformation of the pulmonary architecture that remains incompatible with life. Unlike lobar or segmental bronchial atresia, MBA has a dramatic perinatal course characterized by marked overdistention and malformation of the right lung, mediastinal compression with contralateral shift, hydrops, mirror syndrome, and ultimately fetal or neonatal demise [1,2]. The lethal outcomes in this condition have prompted consideration of fetal surgical intervention as a potential means of fetal salvage, but to date, no successful outcomes are reported. The purpose of this study is to report 3 cases of prenatally diagnosed MBA, including one that was treated with open fetal pneumonectomy, and to discuss the keys to diagnosis and treatment of this uncommon entity.

1. Methods

With approval from the Institutional Review Board of Baylor College of Medicine (H-29695), the records of all patients referred to our comprehensive fetal treatment center for a fetal lung malformation from 2001 to 2012 were reviewed. MEDLINE®/PubMed® database searches were conducted in an iterative manner during January–March 2013 to retrieve articles related to the prenatal diagnoses of fetal lung masses with an emphasis on mainstem bronchial atresia over a 30 year period from January 1982 to January 2012. Search terms were restricted to the English language and included "congenital lung malformation", "congenital cystic adenomatoid malformation", "CCAM", "bronchial atresia," "mainstem bronchial atresia", "prenatal diagnosis", "fetal surgery", and "fetal intervention." No specific key words were required as inclusion criteria since a relatively small number of studies existed on the topic, so a "bottom-up" search strategy was required.

2. Results

Of 129 fetuses evaluated for a fetal lung malformation at our center, three (2.3%) were diagnosed prenatally with MBA. All cases were male and showed large, right-sided masses with dilated right mainstem bronchi on fetal MRI. Mortality in these cases was 100%.

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2.1. Case reports

2.1.1. Patient 1

A 32-year primigravida presented to the Texas Children's Fetal Center for evaluation of a right-sided lung mass detected on routine ultrasound at 19 weeks' gestation. The pregnancy was conceived without assistance and had been progressing uneventfully until that point, and several first trimester ultrasounds had been normal. Evaluation with fetal MRI at 22 3/7 weeks showed an extremely large, right-sided mass with marked contralateral mediastinal shift, eversion of the right hemidiaphragm, and a very small left lung. A cystic lesion was noted centrally, suggestive of a mucocele proximal to a centrally obstructed bronchus (right main; Fig. 1). The CCAM-volume ratio (CVR) was measured at 9. No normal right lung was visualized and the presumptive diagnosis was proximal right bronchial atresia. The mother received steroids in an attempt to limit growth of the fetal mass and possibly to enhance maturation of the normal, contralateral lung. Over the next several weeks the fetal hydrops progressed to maternal mirror syndrome, leading to an induced vaginal delivery of a hydropic, nonviable fetus at 29-6/7 weeks' gestation. The postmortem examination revealed atresia of the right mainstem bronchus with marked dilation of the distal, obstructed airways.

2.1.2. Patient 2

A 16 year-old primigravida was evaluated at 18 weeks gestation for sonographic evidence of an echogenic right chest mass. Fetal MRI at 21-2/7 weeks' gestation, revealed a large, right-sided, homogeneous mass occupying the entire right hemithorax and causing eversion of the hemidiaphragm, significant mediastinal shift, compression of the left lung, and ascites (Fig. 2A). The CVR was 2.6; there was no hydrops. The mother was given steroids, and the pregnancy continued without significant changes. Repeat fetal MRI at 32-1/7 weeks GA revealed a slight decrease in the size of the central cystic lesion (dilated bronchus), from $15 \times 12 \times 13$ mm to $14 \times 9 \times 12$ mm, and improvement in the diaphragmatic eversion. However, the leftward mediastinal shift and left lung hypoplasia remained significant (Fig. 2B–D).

The fetus was delivered at 34 weeks, intubated, and quickly required escalation of respiratory support to include high frequency oscillatory ventilation and nitric oxide therapy. Initial chest x-rays revealed a partially air-filled lung on the left side. The patient had progressive failure of ventilation despite maximum support, and was not considered a candidate for ECMO because of irreversible, severe pulmonary hypoplasia. An urgent right thoracotomy was done with the goal to decompress the mass-effect, and to consider right pneumonectomy. During operation, the neonate's respiratory status continued to decline. Given the poor prognosis, the operation was ended without resection. The infant died shortly after support was withdrawn. Postmortem examination confirmed the diagnosis of right mainstem bronchial atresia with a 2 mm atretic segment near the lung hilum. There was massive hyperplasia of the right lung (observed weight of right lung = 56.5 g; expected weight of both lungs combined = 33.5 g) and marked distention of the obstructed bronchi, mild bronchiectasis and bronchiolectasis, and patchy fluid-filled air spaces with double capillary alveolar walls. The right hemithorax was expanded with shift of the mediastinum into the left hemithorax causing posterior displacement of the left lung. The left lung was severely hypoplastic (observed weight = 8.7 g; expected weight = 33.5 g for combined right and left lungs).

2.1.3. Patient 3

A 35 year-old G6P4 woman with a history of 4 prior cesarean sections and gastric bypass was referred after fetal ultrasound at 19 5/7 weeks' gestation showed a right lung mass. Fetal center evaluation, including fetal MRI, at 20-6/7 weeks' gestation, showed evidence for right mainstem bronchial atresia. The entire right lung was massively over expanded (occupying 80% of the thorax) with compression of the right hemidiaphragm, no normal appearing right lung tissue, and a CVR of 5.7 (Fig. 3A and B). The fetus had findings of hydrops with abdominal ascites, scalp edema, and a small pericardial effusion. Echocardiogram revealed compression of the cardiac chambers and signs of early heart dysfunction, including atrioventricular valve regurgitation, flow reversal in the ductus venosus, and notching in the umbilical vein. After extensive multidisciplinary consultation, discussion of treatment options, and review of the case by the Fetal Therapy Board at Texas Children's Hospital and Baylor College of Medicine, the family asked to proceed with efforts at fetal surgical resection, understanding that the approach was reasonable but that the outcome of this intervention could not be predicted. Open fetal surgery was performed at 21 weeks. Following extensive lysis of adhesions, uterine exposure was achieved and a stapled hysterotomy performed. The right arm of the fetus was delivered and right chest exposed. Given the tamponade physiology, a 20 cc/kg bolus of 5% albumin was administered in the umbilical vein to preload the heart, and then a right thoracotomy was performed. Efforts were taken to exteriorize the right lung mass slowly to minimize abrupt changes in mediastinal pressure. Expected fetal bradycardia was treated with atropine and low dose epinephrine. Examination of the thoracic anatomy confirmed massive over inflation of all three lobes of the right

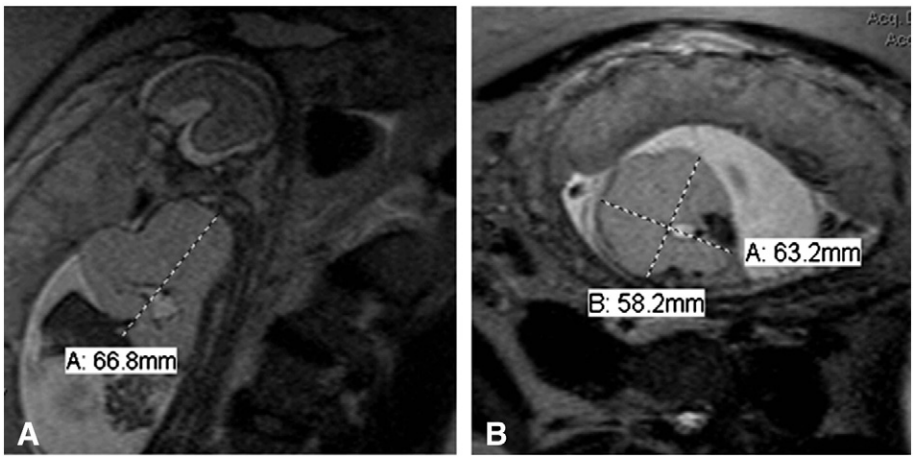

Fig. 1. T2-weighted fetal MR single shot imaging with additional bFFE images of a 22-3/7 weeks GA fetus. Sagittal (A) and axial (A) views demonstrate an extremely large, right side based chest mass with marked mass effect, including displacement of the heart and mediastinum, and inversion of the hemidiaphragms, and very little lung tissue visible on the left. There is a central high signal branching structure at the right hilum that is very suggestive of a mucocele and central bronchial atresia of the right lung. Measured CVR = 9.