Clinical Case Report

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A "blind" vascular ring in association with congenital cystic adenomatoid malformation

A case report

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Abstract

Rationale: The occurrence of congenital cystic adenomatoid malformation (CCAM) and vascular ring (VR) is extremely rare.

Patient concerns: We present a case of left CCAM with VR consisting of a left aortic arch and right descending aorta with left tracheal compression causing atelectasis.

Diagnoses: A high-risk male neonate with the diagnosis of left CCAM was diagnosed at 20 weeks gestational age by antenatal ultrasound. Chest CT revealed multiple cysts in the left inferior lung. Cardiac CT showed VR consisting of a left aortic arch and right descending aorta with left tracheal compression causing atelectasis.

Interventions: left inferior lobectomy was performed. Cardiac CT showed VR consisting of a left aortic arch and right descending aorta with left tracheal compression causing atelectasis. Descending aorta transposition was performed.

Outcomes: The patient recovered smoothly and remained asymptomatic during the 12-months of postoperative follow-up period.

Lessons: We report this rare case of CCAM with VR consisting of left aortic arch and right descending aorta with left tracheal compression causing atelectasis. From the findings of this report, early surgical treatment is recommended. Although the prognosis after surgery remained good, second surgery can be avoided if VR was detected early.

Abbreviations: CCAM = congenital cystic adenomatoid malformation, CT = computed tomography, CVR = CCAM volume ratio, MRI = magnetic resonance imaging, US = ultrasonography, VR = vascular ring.

Keywords: congenital cystic adenomatoid malformation, diagnosis, treatment, vascular ring

1. Introduction

Congenital cystic adenomatoid malformation (CCAM) is a developmental abnormality within the fetal lung, resulting from terminal bronchial proliferation and abnormal alveolar development. It is a relatively rare congenital malformation of the lungs, and occurs in approximately 1.04 per 10,000 births. CCAM is usually diagnosed prenatally by means of routine ultrasonographic analysis, and is typically unilateral, involving only a single lobe of the lung. Some studies have identified CCAM volume ratio (CVR) > 1.6, where polyhydramnios and hydrops are considered as significant predictors of neonatal respiratory distress.[1] Surgical removal of the lesion is considered the gold standard for all patients with CCAMs who became symptomatic.[2]

Vascular rings (VRs) refer to a variety of congenital abnormalities that are related to the position or branching of the aortic arch and this occurs in 1 to 2% of the general population.[3,4] Some anomalies of the aortic arch may form VRs, causing clinical symptoms due to tracheal or esophageal compression. Patients with clinical manifestations should be carefully examined and operated as soon as the diagnosis was made.

Cases of CCAM combined with aortic arch anomalies are less seen. To our knowledge, this is the first case report of CCAM with VR consisting of a left aortic arch and right descending aorta with atelectasis in English literature. Knowledge concerning the diagnosis and management of CCAM with VR is reviewed.

2. Case report

A 30-minute-old term infant initially presented to our hospital after detection of a left CCAM 5 months before delivery. The infant weighed 3110 g at 41 weeks estimated gestational age; cesarean section was performed for premature rupture of membranes. The Apgar scores were 9, 10, and 10 at 1, 5, and 10 minutes, respectively. He had normal vital signs, color, activity, and feeding, no respiratory difficulty, and normal stool and urine output.

The mother was 36 years old, and she had an uncomplicated pregnancy. Ultrasound (US) examination was performed at

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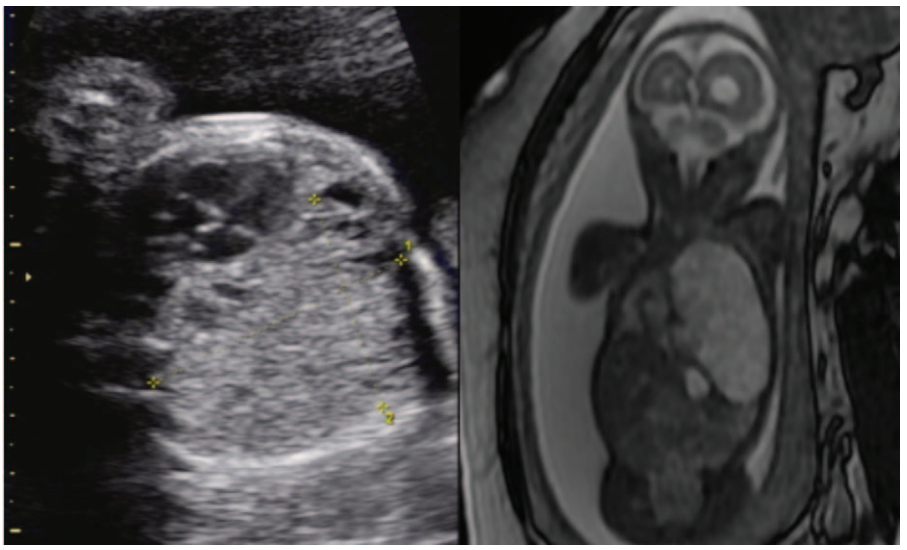

Figure 1. Ultrasound and MRI showed relatively well-defined solid and mixed cystic lesions in the left hemithorax, pleural effusion, and polyhydramnios. MRI = magnetic resonance imaging.

20 weeks gestational age, which showed relatively well-defined solid and mixed cystic lesions in the left hemithorax. When rechecked by using US and fetal magnetic resonance imaging (MRI), the lesions were increased in size to 7.0 cm × 5.4 cm × 4.5 cm with mild polyhydramnios and mediastinal shift (Fig. 1). After prenatal evaluation, he was considered to be a high-risk fetus, due to CVR value of 2.31, pleural effusion, and polyhydramnios. Maternal blood test results were all negative for active infection by adenovirus, cytomegalovirus, or toxoplasma.

On admission, the neonate showed respiratory distress and required urgent oxygen supplementation. Chest CT revealed a 4.6 cm × 5.6 cm × 6.2 cm multiple cystic lesions in the left inferior lung (Fig. 2A and B). The left inferior lobectomy was performed, and x-ray showed left atelectasis after surgery. Bronchofiberscopic

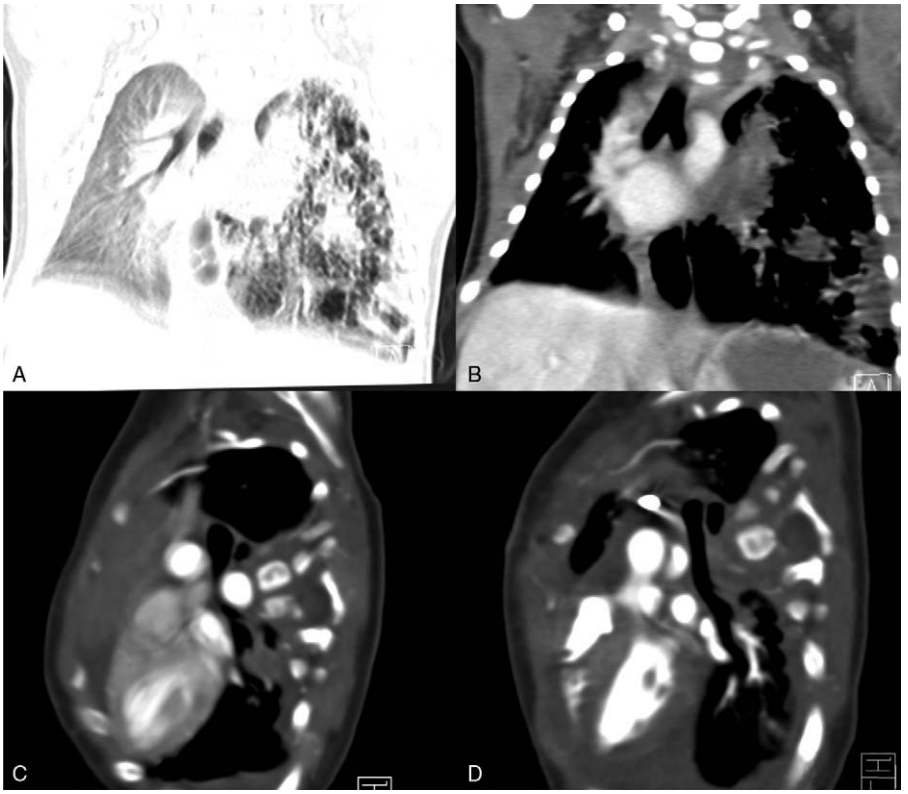

Figure 2. (A, B) Chest CT demonstrated a multiple cysts lesions (4.6 × 5.6 × 6.2 cm) in the left inferior lung. (C) Cardiac CT demonstrated a left aortic arch and right descending aorta with left tracheal compression causing atelectasis. (D) Postoperative cardiac CT demonstrated restoration of lung compression after relief of bronchogenic compression. CT = computed tomography.

findings of bronchogenic stenosis showed external left main bronchial stenosis. Cardiac CT showed left aortic arch and right descending aorta with left tracheal compression causing atelectasis (Fig. 2C). Descending aorta revascularization was performed. The patient recovered smoothly and remained asymptomatic during the 12-months of postoperative follow-up period (Fig. 2D).

3. Discussion

CCAM combined with VR is an extremely rare condition. To our knowledge, this is the first case report of CCAM with VR consisting of left aortic arch and right descending aorta with left tracheal compression causing atelectasis in English literature. In the present case, VR was misdiagnosed as pulmonary lesion causing mediastinal shift.

CCAM is a rare pulmonary developmental disorder with replacement of normal pulmonary tissues by dilated bronchiolar-like airspaces having variable size and distribution. It occurs sporadically, and no genetic predisposition or gender predilection was observed. Recently, increased incidence of CCAM has been reported with improved prenatal ultrasound techniques. The natural history of prenatally diagnosed CCAM ranges from respiratory distress at birth with neonatal death to entirely asymptomatic lesions discovered when incidentally performing a chest radiograph for recurrent pneumonias or spontaneous pneumothorax during adulthood.[5] Majority of the pregnancies with an affected fetus demonstrated an excellent outcome. When infants with CVR>1.6, polyhydramnios, and hydrops are at high risk.[1] After prenatal evaluation, the present case was considered to be a high-risk fetus, due to the CVR value of 2.31, pleural effusion and polyhydramnios as detected by US. This case required delivery in a hospital that has pediatric thoracic surgery department. The patient had shortness of breath soon after birth, and this finding was identical with those reported in the literature.

VRs refer to a variety of congenital abnormalities that are related to the position or branching of the aortic arch, occurring in 1 to 2% of the live births. Some anomalies of the aortic arch, particularly those with aberrant branching, may form VRs, causing clinical symptoms from tracheal or esophageal compression.[4] The definition of left or right aortic arch refers to the bronchus that is crossed by the arch, not to the side of the midline where the aortic root ascends.[4] Fiber bronchoscope and cardiac spiral CT were considered as reliable diagnostic modalities for direct diagnosis of VR. The case described here had incomplete VR consisting of left aortic arch and right descending aorta. This subsequently led to left tracheal compression causing atelectasis, and was diagnosed by fiber bronchoscope and cardiac spiral CT.

The relationship between CCAM and VR is still unclear, and we make some speculations below. Firstly, previous studies have demonstrated that CCAM is the "attack" of vessels, such as in transient ischemic attack, during the critical bronchopulmonary development stage of normal lung,[6] and VR is a vascular abnormality, that results from CCAM. Secondly, this case had large lung lesion, which may lead to vascular displacement, resulting in VR. In addition, no relations between CCAM and VR were observed. Despite this report, the specific relationship between CCAM and VR has remained largely unexplored. Hence, further research is needed to clearly understand the relationship between CCAM and VR.

Surgery of symptomatic lesions remains relatively straightforward for both CCAM and VR, and hence, early surgery is recommended. The prognosis after surgery remained good and there is no evidence of recurrence or metastasis after surgery. Lobectomy or pneumonectomy is often recommended for CCAM. For VR, descending aorta revascularization was performed. The patient recovered smoothly and remained asymptomatic during the 12-months of postoperative follow-up period.

4. Conclusion

In summary, we report this rare case of CCAM with VR consisting of left aortic arch and right descending aorta with left tracheal compression causing atelectasis. From the findings of this report, early surgical treatment is recommended. Although the prognosis after surgery remained good, second surgery can be avoided if VR was detected early.

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