Case Report

Bronchial atresia in a neonate with congenital cytomegalovirus infection

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Abstract:

Bronchial atresia (BA) is characterized by a mucus-filled bronchocele in a blind-ending segmental or lobar bronchus with hyperinflation of the obstructed segment of the lung. We describe a neonate who presented on his 9th day of life with respiratory distress. Chest computed tomography showed a soft tissue density involving the right middle lobe (RML). RML lobectomy confirmed the diagnosis of BA. Cytomegalovirus was detected by polymerase chain reaction in blood, urine, and tracheal aspirates which may provide further insight into the pathogenesis of BA.

Key words:

Bronchial atresia, congenital, cytomegalovirus, infection, neonate

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Bronchial atresia (BA) is an anomaly characterized by a mucus-filled bronchocele in a blind-ending segmental or lobar bronchus, with hyperinflation of the obstructed segment of lung.[1] It was first described in the literature by Ramsay in 1953.[2] Subsequently, in 1963, Simon and Reid[3] described it in detail in a series of three patients who had an atretic bronchus in the antero-apical region of the left upper lobe. We report a case of BA in a neonate with congenital cytomegalovirus (cCMV) infection.

Case Report

A 9-day-old boy presented with a history of increased work of breathing and cyanosis. He was born at 40 weeks gestation via vacuum extraction for fetal distress. His birth weight was 3.0 kg with normal Apgars and did not require any resuscitation. Meconium-stained liquor was noted at delivery. He was discharged home at 48 h of age. Antenatal ultrasound at 22 weeks of gestation demonstrated hyper-echoic changes in the thorax and abdomen which did not progress throughout pregnancy. At presentation to the hospital, he was in moderate respiratory distress and was commenced initially on continuous positive airway pressure, but he required mechanical ventilation for respiratory deterioration in the 2nd week of life. He had a normal white cell count and C-reactive protein. Blood cultures were negative. CMV was detected by polymerase chain reaction (PCR) in urine and blood, and in endotracheal aspirate samples. In addition, stored blood samples (from newborn screen) taken on day 2 of life were also positive for CMV by PCR. CMV immunoglobulin M antibody was also positive. A head ultrasound demonstrated diffuse peri-ventricular calcification. A chest computed tomography (CT) scan was performed [Figure 1a-c]. Flexible bronchoscopy and echocardiography were normal.

Due to an ongoing requirement for mechanical ventilation associated with left mediastinal shift and lung compression from an overinflated right middle lobe (RML), the child had a RML lobectomy [Figure 2].

Pathology revealed a bronchocele with an 11-mm mucus plug [Figure 2] in a sub-segmental bronchus of the RML. Although bronchi were seen to arise from the cyst and communicate with the distal lung causing marked over-inflation, no direct continuity was identified between the bronchocele and proximal bronchi. Cytomegalovirus inclusions with minimal surrounding inflammation were noted on microscopy of the peripheral lung.

After excision of the RML, the patient was extubated and gradually weaned off oxygen. However, following anesthesia for a central line placement at 1 month of age for a 6-week course of ganciclovir for cCMV infection, he developed further respiratory distress. A repeat CT scan demonstrated persistent hyperinflation of the residual right lung, especially the right lower lobe. Instead of further lobectomy with permanent loss of lung mass, right lung volume reduction surgery was performed. There was subsequent marked clinical improvement and he was discharged home aged 3½ months and continues to thrive with no respiratory distress.

Discussion

The etiology of BA remains unknown. It was thought to be caused by an antenatal vascular insult around the 16th week of gestation during

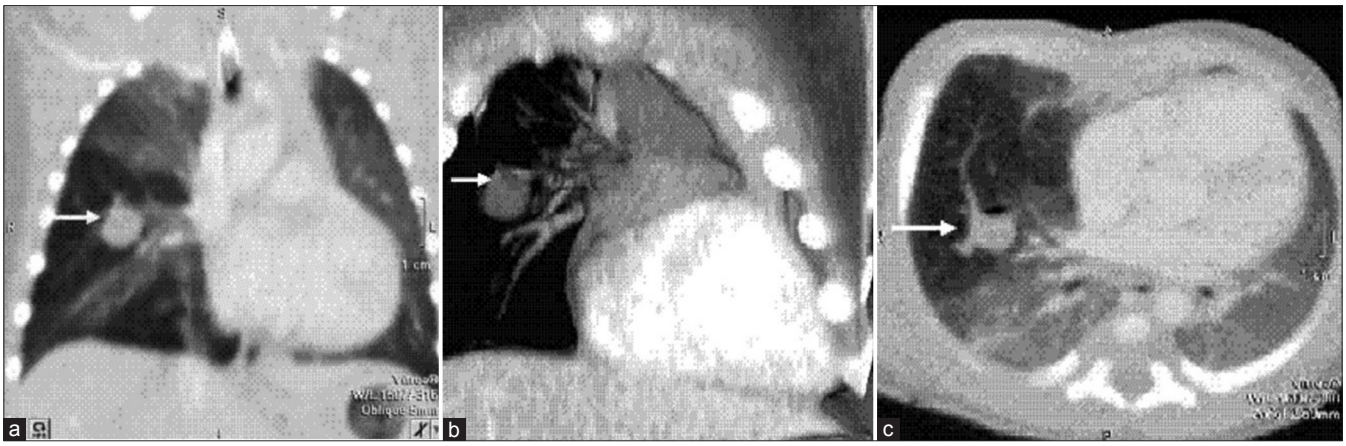

Figure 1: (a) The coronal view of the computed tomography shows a soft tissue density involving the right middle lobe with hyper expanded segments just distal to it (b) A 3D saggital view shows that the 1 cm lesion is not communicating with any adjacent vessel or bronchus (c) The lesion seems to be fluid filled with parts of the expanded lungs herniating anteriorly to the other side as seen in this axial view

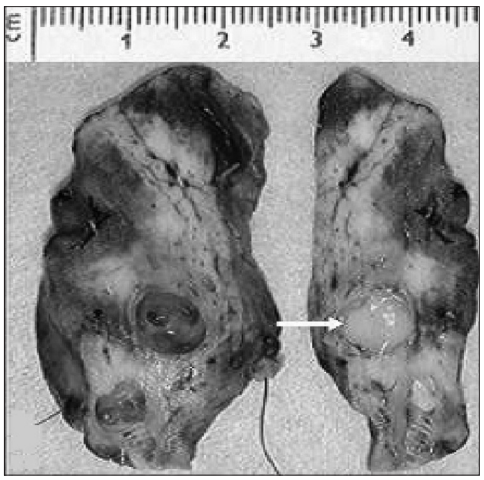

Figure 2: The bronchocele with an 11 mm mucous plug

the late stages of lung development.[3-5] However, other theories suggest that a nest of proliferating cells loses connection with the distal tip of the developing bronchial bud and continues to branch independently. As a result, normal branching distal to the atresia is maintained without actual connection to the central airway. It is hypothesized that this would occur around the 5th-6th week of gestation which is the time the proximal airways develop. This is the time when bronchogenic cysts are thought to develop.[6]

As there is no direct communication with the central airways, the hyperinflation distal to the atretic segment is thought to be due to aeration by collateral air drift through the intraalveolar pores of Kohn, the bronchoalveolar channels of Lambert, and the interbronchiolar pores of Martin. This theory is supported by newer imaging techniques using Xenon ventilation CT.[7]

In children, BA usually has a symptomatic presentation with cough, respiratory distress, or recurrent infections and has a female predominance (59%). It occurs most commonly in the right lower lobe (39%) followed by left or right upper lobes (23%).[7]

A prenatal diagnosis of BA using ultrasound and fetal magnatic resonance imaging MRI has seldom been made.[5,8-10] Postnatally, chest radiographs and CT are the main tools in diagnosis and may show segmental hyperinflation and mucus impaction. Surgical resection of the affected segment should be considered in symptomatic patients.

Our patient is particularly interesting because of the congenitally acquired CMV.

CMV was identified by PCR in urine, blood, and respiratory secretions in the 2nd week of life and from stored blood on newborn screen on day 2 of life. These, together with the clinical presentation confirm symptomatic congenital CMV infection. One case previously reported congenital lobar emphysema in a patient with cCMV infection.[11] but BA, to the best of our knowledge, has never been reported in cCMV-infected patients. It is possible that the CMV infection caused the BA, either due to a vascular insult or secondary inflammation at a crucial time of bronchogenesis causing atresia of the affected bronchus, as was previously hypothesized.[11] This finding is supported by the presence of CMV inclusion bodies in the resected lobe.

In summary, this is the first reported case of BA occurring with cCMV infection which may give further insight into the pathogenesis of this rare condition.

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