Case Report

Congenital Cystic Adenomatoid Malformation, Type II: A Rare Cause of Haemoptysis

Pradipprava Paria¹, Saptarshi Das¹, Sibnath Gayen¹, Sibarjun Ghosh¹, Rajarshi Basu² and Goutam Bandyopadhyay³

Departments of Paediatrics¹, Cardiothoracic Surgery² and Pathology³, R.G. Kar Medical College, Kolkata (West Bengal), India

Abstract

Congenital cystic adenomatoid malformation (CCAM) occurs secondary to the cystic adenomatous over-growth of terminal bronchioles, which results in the secondary inhibition of alveolar growth. In most of the cases, respiratory distress is the presenting feature during the neonatal period. In about 90% of patients, recurrent respiratory infections necessitating chest imaging reveal CCAM before the age of two years. We describe here the occurrence of congenital cystic adenomatoid malformation of right lung in a 12-year-old girl presenting with haemoptysis and hypovolaemic shock. She underwent right middle lobectomy; and histopathological examination confirmed the diagnosis. She has been doing well on follow-up. [Indian J Chest Dis Allied Sci 2016;58:53-57]

Key words: Congenital cystic adenomatoid malformation, Haemoptysis, Lobectomy.

Introduction

Congenital cystic adenomatoid malformation (CCAM) of lung is a rare anomaly with incidence around 1-4/100,000 births.¹ It results from aberration in the developmental process of terminal airways leading to hamartomatous dysplastic changes, usually confined to one lobe.² Congenital cystic adenomatoid malformation are classified into three different types based on their gross appearance (Stoker’s Type I, II and III).³ Majority of cases (50%) are Stoker’s Type I. Type II (40%) has bad prognosis due to its severity in clinical course and propensity to have sarcomatous or carcinomatous transformation later.⁴ Type III is extremely rare (<10%). With the advancements in prenatal diagnosis, 60% of the cases of CCAMs can be detected in utero.⁵ Rest of the cases are detected mostly within two years of age. Congenital cystic adenomatoid malformation can rarely present in an adult with recurrent respiratory tract infections, pneumatocele or pneumothorax.⁶ We describe a case of Stoker’s Type II CCAM presenting for the first time in a 12-year-old girl with massive haemoptysis and hypovolaemic shock. The case is being reported because of its extreme rarity in incidence as well as presentation.

Case Report

A 12-year-old-girl presented in a state of hypovolaemic shock following a sudden onset, large bout of haemoptysis. She did not complain of fever. There was no history of recurrent respiratory tract infection, foul smelling sputum, breathlessness, cyanosis. There was no history of prolonged bleeding after trauma, history of contact with tuberculosis. Perinatal history was also uneventful. She was resuscitated with intravenous fluids and three units of blood transfusion. Antibiotics were added. General physical examination revealed severe pallor with hypotension. There was no cyanosis or clubbing. Neck veins were not engorged. No cervical lymph nodes were palpable. On systemic examination, upper airway was normal. Trachea was in midline position. No visible localised or generalised bulge or depression or restriction of movement of chest wall was present. There was dullness on percussion over infrascapular and interscapular region on the right side. Over the same regions, auscultatory findings revealed diminished vesicular breath sound, no adventitious sounds and diminished vocal resonance. No cardiac murmur was heard. Liver and spleen were not palpable.

Chest radiograph revealed right-sided lower and middle zone opacity (Figure 1). Laboratory testing revealed haemoglobin 10gm/dL (following transfusion of 3 units of whole blood); total leucocyte count was 6100/mm³ with a differential count of polymorphs 63%, lymphocytes 35%, eosinophils and monocytes 1% each; platelet count was 2.1 lac/cc. Erythrocyte sedimentation rate (ESR) was 35mm at the end of the first hour. Blood coagulation profile including prothrombin time, international normalisation ratio, augmented partial thromboplastin time were normal.

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Correspondence and reprint requests: Dr Pradipprava Paria, Department of Pediatrics, R.G. Kar Medical College, 1, Khudiram Bose Sarani, Kolkata-700 004 (West Bengal), India; E-mail: drpradip83@gmail.com
Serum biochemistry revealed serum aspartate aminotransferase (SGOT) 30IU/L, serum alanine aminotransferase (SGPT) 16IU/L, serum alkaline phosphatase (ALP) 110IU/L, serum total bilirubin 1.3mg/dL (direct bilirubin 0.5mg/dL). Mantoux test (5 Tuberculin Units) was non-reactive. Sputum smear examination for acid-fast bacilli was negative. Computed tomography (CT) of the thorax showed a mass in the middle zone of the right lung with minimal collapse and areas of ground-glass opacification (Figure 2). Considering the possibility of arteriovenous (AV) malformation, bronchoscopy was planned but could not be performed, as she again had another episode of massive haemoptysis during preparation for the procedure and required another 3 units of whole blood transfusion for resuscitation. A CT-guided bronchoscopy was done, which revealed obstruction at the right middle lobe bronchus with gradual luminal compromise (Figure 3).

She was then transferred to Cardio-thoracic Surgery Department and underwent right middle lobectomy under one lung ventilation. On gross pathological examination, the excised specimen revealed multiple, evenly spaced cysts attached to the bronchial tree. Histopathology of the segment showed foetal lung pattern with honey-combing and multiple tiny cysts measuring 0.5-1.0 cm in diameter. Examination of multiple sections revealed collapsed cystic spaces lined by bronchiolar type pseudo-stratified columnar epithelium. The walls of the cysts were thin and deficient of muscle coat. Cysts contained mucinous material and were surrounded by collapsed alveoli with haemosiderin laden macrophages. All the above features suggest congenital cystic adenomatoid malformation of lung, type II (Figure 4). The patient recovered successfully after the operation. Ultrasonography of the abdomen did not reveal any abnormality. Chest radiograph showed expansion of the lung. One year post-operative follow-up was uneventful. Published reports of CCAM occurring in adults are summarised in the table.6-25
Table. Published cases of adult congenital cystic adenomatoid malformation.

<table>
<thead>
<tr>
<th>Study, Year of Publication</th>
<th>Age (Years), Gender</th>
<th>No. of Cases</th>
<th>Clinical Features</th>
<th>Site</th>
<th>Stoker’s Classificationa</th>
<th>Treatment Done</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vicidomini et al, 1997</td>
<td>62, M</td>
<td>1</td>
<td>Recurrent pulmonary infection</td>
<td>Right middle and lower lobe</td>
<td>II</td>
<td>Lobectomy</td>
</tr>
<tr>
<td>Hellmuth et al, 1998</td>
<td>18-65, M 11 F 12</td>
<td>23</td>
<td>Recurrent pulmonary infection</td>
<td>Left lower lobe 11 Right lower lobe 7 Right upper lobe 1 Right middle lobe 2</td>
<td>I 19 II 3 Not specified 1</td>
<td>Lobectomy 16 Cyst resection 2 Pneumonectomy 1 Not specified 4</td>
</tr>
<tr>
<td>Vicidomini et al, 2000</td>
<td>21, M</td>
<td>1</td>
<td>Recurrent pulmonary infection</td>
<td>Right lower lobe</td>
<td>I</td>
<td>Lobectomy</td>
</tr>
<tr>
<td>Hellmuth et al, 2002</td>
<td>0.5-23, M 8 F 4</td>
<td>12</td>
<td>Recurrent pneumonia 9 Pneumothorax 1 Chance finding 2</td>
<td>Left lower lobe 3 Right lower lobe 6 Left upper lobe 2 Right upper lobe 1</td>
<td>I 7 II 4 Not specified 1</td>
<td>Lobectomy 8 Segmentectomy 2 Local resection 2</td>
</tr>
<tr>
<td>Hellmuth et al, 2004</td>
<td>51, M</td>
<td>1</td>
<td>Recurrent pulmonary infection</td>
<td>Right lower lobe</td>
<td>I</td>
<td>Wedge resection</td>
</tr>
<tr>
<td>Hellmuth et al, 2005</td>
<td>46-47, M 1 F 1</td>
<td>2</td>
<td>Recurrent pulmonary infection</td>
<td>Right lower lobe 1 Left lower lobe 1</td>
<td>I</td>
<td>Lobectomy</td>
</tr>
<tr>
<td>Hellmuth et al, 2006</td>
<td>24, F</td>
<td>1</td>
<td>Recurrent pneumothorax</td>
<td>Left lower lobe</td>
<td>II</td>
<td>Wedge resection</td>
</tr>
<tr>
<td>Hellmuth et al, 2007</td>
<td>17-64, M 2 F 5</td>
<td>7</td>
<td>Recurrent pulmonary infection</td>
<td>Left lower lobe 2 Left upper lobe 1 Right upper lobe 2 Right middle lobe 1 Right lower lobe 1</td>
<td>I-3 II-4</td>
<td>Lobectomy 6 Wedge resection 1</td>
</tr>
<tr>
<td>Hellmuth et al, 2007</td>
<td>18, M</td>
<td>1</td>
<td>Recurrent pulmonary infection</td>
<td>Right lower lobe</td>
<td>I</td>
<td>Lobectomy</td>
</tr>
<tr>
<td>Morelli et al, 2007</td>
<td>38, M</td>
<td>1</td>
<td>Asymptomatic</td>
<td>Right middle lobe</td>
<td>I</td>
<td>Lobectomy</td>
</tr>
<tr>
<td>West et al, 2007</td>
<td>18, M</td>
<td>1</td>
<td>Recurrent pulmonary infection</td>
<td>Left lower lobe</td>
<td>I</td>
<td>Lobectomy</td>
</tr>
<tr>
<td>Khan et al, 2008</td>
<td>33, M</td>
<td>1</td>
<td>Asymptomatic</td>
<td>Left upper and lower lobe</td>
<td>I</td>
<td>Pneumonectomy</td>
</tr>
<tr>
<td>Lai et al, 2009</td>
<td>40, F</td>
<td>1</td>
<td>Asymptomatic</td>
<td>Right upper lobe</td>
<td>I</td>
<td>Surgical resection</td>
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<tr>
<td>DiScioscio, 2010</td>
<td>34, F</td>
<td>1</td>
<td>Recurrent pneumonia, Haemoptysis</td>
<td>Right upper lobe</td>
<td>I</td>
<td>Conservative</td>
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<tr>
<td>Kwak et al, 2011</td>
<td>58, F</td>
<td>1</td>
<td>Cough &amp; blood tinged sputum</td>
<td>Right middle lobe</td>
<td>I</td>
<td>Lobectomy</td>
</tr>
<tr>
<td>Feng et al, 2012</td>
<td>26, 28, F 2</td>
<td>2</td>
<td>Persistent cough, fever</td>
<td>Left lower lobe</td>
<td>II</td>
<td>Lobectomy</td>
</tr>
<tr>
<td>Poflee et al, 2014</td>
<td>19, M</td>
<td>1</td>
<td>Persistent cough, expectoration</td>
<td>Right lower lobe</td>
<td>I</td>
<td>Lobectomy</td>
</tr>
<tr>
<td>Rao et al, 2014</td>
<td>37, M</td>
<td>1</td>
<td>Chronic cough and dyspnoea on exertion</td>
<td>Left upper and lower lobe</td>
<td>I</td>
<td>Left pneumonectomy</td>
</tr>
<tr>
<td>Sergiacomi et al, 2014</td>
<td>44, M</td>
<td>1</td>
<td>Progressive dyspnoea</td>
<td>Lingual and both lower lobe</td>
<td>II</td>
<td>Conservative management</td>
</tr>
<tr>
<td>Malinova et al, 2015</td>
<td>21, M</td>
<td>1</td>
<td>Recurrent productive cough</td>
<td>Right middle and lower lobe</td>
<td>II</td>
<td>Resection</td>
</tr>
</tbody>
</table>

Definition of abbreviations: M=Male; F=Female.
Congenital cystic adenomatoid malformation usually results from aberration of terminal airway development, usually within 5-6 weeks of gestational age, before perichondrial tissue is formed. Males are affected as frequently as females. The left lung is involved as often as the right one; and single lobe disease observed four times more often than multi-lobe disease. It has been described that it can cause still-born and premature birth. Immediately after birth, respiratory distress is frequent in most cases. As the child grows, repeated respiratory infections and pneumothorax become frequent. The CCAM is most commonly detected in the neonatal period and up to 90% of diagnoses are made within the first two years of life. It is rare to get first time presentation of CCAM in the adolescents and adults; and after extensive literature search, we found about 60 such reported cases (Table). Our case is an unusual one due to the adolescent age of onset. Another rarity of this case is its initial presentation as hypovolemic shock due to massive haemoptysis.

Congenital cystic adenomatoid malformations are classified into three different types based largely on their gross appearance. Type I has large (>2cm) multiloculated cysts, lined with ciliated pseudo-stratified-columnar epithelium. The walls of the cysts contain smooth muscle cells and elastic tissue. One-third of the cases have mucus secreting cells. Cartilage is rarely seen in the cyst wall. Most of the adult cases diagnosed are of Type I (60%-70%). Type II has smaller uniform cysts with similar histopathologic pattern as Type I. The presence of microcystic lesions in Type II, identified on prenatal ultrasonography, has been reported to be associated with hydrops foetalis and still-births, thus, carrying a poor prognosis. Type II is associated with other congenital anomalies like kidney growth failure, diaphragmatic hernia, jejunal atresia, and colon growth failure. Our case, though of Type II, presented at a later age with uneventful perinatal history and no associated congenital anomaly. Type III is not grossly cystic, referred to as the “adenomatoid” type. Microscopically, the lesions are not true cysts, but communicate with the surrounding parenchyma. Differential diagnosis of CCAM includes other cystic lesions, like pulmonary sequestration, bronchogenic cyst, congenital lobar emphysema, diaphragmatic hernia and cystic bronchiectasis. To differentiate, radiographic findings are very dependable. Typically the chest radiograph in CCAM shows multi-cystic lesions. The most frequent findings on computed tomography of thorax are nodules with a well-defined boundary accompanied by a ground-glass appearance. Though imaging can give clue regarding the diagnosis, definitive diagnosis is made only by histopathological examination. Surgical resection is the treatment of choice in most symptomatic cases. Elective excision is recommended even in asymptomatic cases because of its potential for malignant transformation.

In conclusion, in adolescents and adults, CCAM may be a rare cause of haemoptysis. Early recognition and timely surgical intervention is essential to prevent the consequences and potential risk of malignant transformation.

References


