Case Report: Congenital Cystic Adenomatoid Malformation of an Entire Lung in a 17-Year-Old Girl Coexisting With VSD.

Dr. Bssem Sahsah Mohammed, Dr. Byan Ahmed Altorbak, Dr. Ghadeer Saleh Alenzi & Ziad alhomidan

Abstract: Congenital cystic adenomatoid malformations (CCAM), also known as congenital pulmonary airway malformation, is a rare, non-inherited developmental abnormality of lung with incidence of about 1 in 30,000. It arises from excessive unsystematic proliferation of tubular bronchial structures that forms non-working cystic lesion of abnormal lung tissue.

We present an unusual case of a 17-year-old girl whom presented with congenital cystic adenomatoid malformation of an entire lung and VSD.

Keywords: Congenital cystic adenomatoid malformation, CCAM.

Introduction

Congenital cystic adenomatoid malformations is rare with incidence of about 1 in 30,000, it is a non-hereditary anomaly of lung structural and functional development so that there is adenomatoid proliferation of cysts, rather than normal tissues, it was classified into five types by Stocker in 2002. CCAM is usually manifests in the first 2 years of life in respiratory distress or by repeated respiratory infection later on.

We present a rare case of a 17-year-old girl with CHD whom presented with shortness of breath and right side chest pain, later on to be diagnosed with congenital cystic adenomatoid malformation of an entire lung.

Case Report

A 17-year-old girl presented to ER with a history of shortness of breath and right side intermittent chest pain that increase with respiration and movement since 6 days ago with intermittent cough, no hemoptysis. She came from a low-income family and has VSD, had past history of repeated similar conditions in the past.

Abdominal examination:
Soft, lax no organomegaly.
Vital signs:
Temperature: 36.4 °C
Pulse=103 /minute
Respiratory Rate=22/minute.
BP=134/60 mm hg

On X ray
Hyper inflated right lung
White left lung
left mediational shift

On CT
Hypo plastic left lung with multiple macro cysts
And will indentified mass in the upper right lung

Course of treatment:

No other medical conditions or diseases.
No family history of diseases or congenital anomalies.

On Examination:
Patient looked ill, was conscious and oriented
Chest examination:
Decreased air entry on the left side
Rt lung Crepitation
S1+S2 with pan systolic murmur.
O2 supplementary 4 L/ min Mask  
Perfalgan 1G IV  
Tab Lasix 20 mg OD  
Amoxicillin tab every 8 hours

Injection Zinforo dose 760 mg  
Every 8 hours  
Steam inhalation

She was admitted for 7 days into medical ward. After improvement discharged.

However, she deteriorated 2 days later, developed acute respiratory failure, and had to be intubated then shifted to ICU.

<table>
<thead>
<tr>
<th>First day:</th>
<th>Second day:</th>
<th>Third day:</th>
</tr>
</thead>
<tbody>
<tr>
<td>WBC=10.19</td>
<td>WBC=11.9</td>
<td>WBC=12.42</td>
</tr>
<tr>
<td>RBCs=9.18</td>
<td>RBCs=7.91</td>
<td>RBCs=8.61</td>
</tr>
<tr>
<td>HGB=16.3</td>
<td>HGB=14.5</td>
<td>HGB=15.3</td>
</tr>
<tr>
<td>PLT=294</td>
<td>PLT=236</td>
<td>PLT=283</td>
</tr>
</tbody>
</table>

Other tests:

- Blood urea nitrogen=1.3 (low)
- Creatinine=38.18 (low)
- Total bilirubin=32.5 (high)
- Creatine kinase MB=27 (high)
- Phosphorous=2.276 (high)
- Potassium=3.3
- Glucose =6.11

The patient developed septic shock and type 1 respiratory failure and arrested she did not respond to resuscitation according to ACLS protocol and was announced died after 3 days since admission to ICU. Autopsy could not be perform.

**Discussion**

Congenital cystic adenomatoid malformation is a rare hematoma of the lung due to faulty embryonic development and proliferation of Tubular bronchial structures, often affecting one lung lobe and both lungs equally. CCAM is usually discovered prenatally or in neonatal period presenting with respiratory distress due to surrounding structures compression. Rarely is it discovered in adulthood with repeated respiratory infections. CCAM can predisposes to the development of neoplasm, such as rhabdomyosarcoma and pneumothorax.

Congenital cystic adenomatoid malformation was first described by Ch’in and Tang in 1949; although it is named congenital cystic adenomatoid malformations only 3 out of the 5 types have cysts.

In 2002 Stocker classified it into 5 types:

<table>
<thead>
<tr>
<th>Types</th>
<th>Frequency</th>
<th>Prognosis</th>
</tr>
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<tbody>
<tr>
<td>Type 0</td>
<td>Rare</td>
<td>Usually Fatal</td>
</tr>
<tr>
<td>Type 1</td>
<td>≈ 65%</td>
<td>good prognosis</td>
</tr>
<tr>
<td>Type 2</td>
<td>≈ 15%</td>
<td>poor prognosis</td>
</tr>
<tr>
<td>Type 3</td>
<td>≈ 5%</td>
<td>poor prognosis can be fatal</td>
</tr>
<tr>
<td>Type 4</td>
<td>≈ 10%</td>
<td>good prognosis</td>
</tr>
</tbody>
</table>
Early diagnosis and management is crucial for prevention of deterioration of patient condition and prevention of complications. Surgical intervention is the definitive management in symptomatic patient to allow normal tissue expansion and prevention of deterioration of preexisting problems.

Conclusion

Congenital cystic adenomatoid malformation is a hematoma of the lung without an identifiable cause that could cause problems in fetal period (hydrops and stillbirth), neonatal period (respiratory distress) or rarely later on manifesting in respiratory distress, neoplasm or repeated respiratory infection among others or could remain symptomless.

Early identification is important to allow for surgery along other modalities for optimal results.

Footnotes:

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References:


