Case Report

Microcystic congenital pulmonary airway malformation of the lung causing non immune hydrops fetalis and Mirror syndrome: a case report

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ABSTRACT

Congenital Pulmonary Airway Malformation (CPAM) is a rare lung anomaly with a highly variable prognosis depending on the presence of fetal hydrops and the size of the cyst. CPAM associated with fetal hydrops carries a grave fetal prognosis and development of “mirror syndrome” calls for immediate delivery of fetus as this condition is potentially life threatening for the mother, if no intervention is done. We report on the pathological findings of a non-immune hydropic fetus with cardiac shift to right side, moderate hydromnios and a rare maternal condition which “mirrors” the sick fetus known as “mirror syndrome” by developing edema and high blood pressure due to microcystic CPAM at 26 weeks of gestation.

Keywords: Microcystic congenital pulmonary airway malformation (CPAM), Non-immune hydrops fetalis, Mirror syndrome

INTRODUCTION

Congenital Pulmonary Airway Malformation (CPAM) is a rare hamartomatous lung lesion and is also known as congenital cystic adenomatoid malformation (CCAM). It was first described by Ch’in and Tang in 1949. It is characterized by abnormal embryogenesis in the form of overgrowth of terminal bronchioles (adenomatoid) at the expense of alveolar spaces. CPAM comprises of heterogeneous group of cystic and solid/microcystic lung lesions. The microcystic lesions are invariably fatal as they cause non-immune hydrops fetalis. The hydrops fetus will place the mother at a slightly increased risk for “mirror syndrome”. Mirror syndrome (Ballantyne’s syndrome) is defined as the development of maternal edema in association with fetal hydrops. John W. Ballantyne in 1892 was the first to describe the association of maternal edema in pregnancy with fetal hydrops due to rhesus isoimmunization. We report here such a rare case of microcystic CPAM causing non immune hydrops fetalis and mirror syndrome.

CASE REPORT

A 20-year-old primigravida was referred to the department of obstetrics at 26 weeks of gestation with gestational hypertension, pedal edema, moderate anemia, mild proteinuria and with suspicion of a non-immune fetal hydrops.

Ultrasonography revealed fetal ascites, cardiac shift to right side, large hyper-echogenic edematous left lung and moderate hydromnios (Figure 1a, 1b and 1c).
The bad fetal prognosis and maternal morbidity were explained to the parents and they opted for termination of pregnancy. The 26-week male fetus was delivered by using inducing agents and sent for fetal autopsy.

Gross examination: revealed distention of abdomen and generalized edema. No other fetal defects were found, nor any signs of fetal anemia. On gross examination the thoracic viscera showed enlarged left lung with mediastinal shift to the right thoracic cage (Figure 2a & 2b). The right lung appeared hypoplastic, about a third of the size of the left lung (Figure 2c). Cut sections of enlarged left lung appeared solid without any obvious cyst formation (Figure 2d). All other thoracic (trachea, esophagus, right lung, heart and major vessels) and abdominal visceral organs appeared to be normal in their anatomy.

**Figure 1a:** USG showing fluid in the peritoneal cavity.

**Figure 1b:** USG showing edematous hyper-echogenic left lung.

**Figure 1c:** USG showing cardiac shift to right side.

**Figure 2a:** Gross of fetus: Thoracic viscera shows enlarged left lung.

**Figure 2b:** Gross of fetus: mediastinal shift to the right thoracic cage.
Figure 2c: Gross photo of viscera showing hypoplastic right lung as compared to left.

Figure 2d: Cut sections of left lung: solid without obvious cyst formation.

Microscopic examination: Sections studied show bronchiole-like structures, lined by tall columnar mucinous ciliated epithelium (Figure 3a & 3b). Alveoli are lined by non-ciliated cuboidal epithelium. There was absence of bronchial cartilage and bronchial tubular glands.

Figure 3a: H&E stain, 5x: showing bronchiole-like structures.

Figure 3b: H&E stain, 10xs: ciliated, columnar mucinous epithelial lining.

Based on histopathology, a final diagnosis of Congenital Pulmonary Airway Malformation (CPAM) type III was considered.

DISCUSSION

Congenital Pulmonary Airway Malformation (CPAM) is a rare developmental abnormality of lung. Incidence being 1 in 25000 to 35000 pregnancies. Males are affected slightly more common than females. CPAM usually involves an entire lung or a single lobe or rarely involve the both lungs and this abnormal tissue will never function as normal lung tissue. The natural history of CPAM is likely related to its growth pattern during embryologic development. Lesion growth may increase or plateau during gestation. The size of lesions and variable growth behavior results in either absence of symptoms/development of non-immune hydrops fetalis.

Causes The underlying cause of CPAM is unknown.

- Resected CCAMs show signs of increased cell proliferation and decreased apoptosis.
• Studies have investigated the role of HOXB5 gene and protein expression, as well as other growth factors such as mesenchymal platelet-derived growth factor-BB.

• As development of the respiratory system begins at 3 weeks of gestation, and as a consequence of abnormal embryogenesis during the first 6-7 weeks of pregnancy, involving maldevelopment of terminal bronchioles leads to CPAM.

Although the routine antenatal imaging studies may suggest the possibility of CPAM, histopathological examination remains the main stay of diagnosis. Diagnostic criteria include:

1. Absence of bronchial cartilage (unless it is trapped within the lesion);
2. Absence of bronchial tubular glands;
3. Presence of tall columnar mucinous epithelium;
4. Overproduction of terminal bronchiolar structures without alveolar differentiation, except in the subpleural areas and
5. Massive enlargement of the affected lobe that displaces other thoracic structures.

Stocker has divided CCAM into five different categories based on the proposed site of defect in the tracheobronchial tree.8

Table 1: CCAM in five different categories based on the proposed site of defect in the tracheobronchial tree divided by Stocker.

<table>
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<th>Stocker classification</th>
<th>Gross and histological Differentiation</th>
<th>Prevalence (%)</th>
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<tbody>
<tr>
<td>Type 0</td>
<td>Lungs are firm and small Involved all lung lobes. Acinar dysplasia;</td>
<td>&lt;2</td>
</tr>
<tr>
<td>Type I</td>
<td>Single or multiple cysts &gt;2 cm in size. Cyst are lined by ciliated columnar to pseudo-stratified tall column epithelium overlying a thin to moderately thick fibromuscular layer.</td>
<td>60-70</td>
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<tr>
<td>Type II</td>
<td>Single or multiple evenly distributed cysts &lt;2 cm in size. Small, uniform cysts, irregular proliferation of ectatic structures resembling bronchioles.</td>
<td>15-20</td>
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<tr>
<td>Type III (microcystic)</td>
<td>Solid lesions &lt;0.5 cm size. Irregular curving channels and small air spaces lined by plump cuboidal epithelium.</td>
<td>5-10</td>
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<tr>
<td>Type IV</td>
<td>Large air filled cysts. Multilocular, large cysts lined by flattened alveolar lining cells.</td>
<td>&lt;10</td>
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These lesions may be asymptomatic, or can lead to enlarged abnormal lung tissue, causing s hypoplasia of normal lung, mediastinal shift, cardiovascular compromise and fetal cardiac failure leading to non-immune fetal hydrops.

The hydrops fetus will place the mother at a slightly increased risk for “mirror syndrome.”

Treatment depends on the size of the lesion and complications.

Treatment of choice for CPAM is lobectomy, performed at approximately 2 to 6 months of age. Thoracocentesis allows drainage of a large cyst with immediate decompression of the CPAM. Fetal surgery (thoracoamniotic shunt) that continually drains fluid from the CPAM to the amniotic space is preferred in some cases.

Maternal termination of pregnancy is the treatment of choice if CPAM is associated with hydrops fetalis and “mirror syndrome.”

CONCLUSIONS

Congenital pulmonary airway malformation is a rare fetal lung anomaly with excellent prognosis in the absence of fetal hydrops. CPAM associated with fetal hydrops carries a grave fetal prognosis and development of “mirror syndrome” calls for immediate delivery of fetus as this condition is potentially life threatening for the mother.

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REFERENCES


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