Congenital diaphragmatic hernia and fetal lung lesions: a preferential association and a prenatal diagnosis challenge

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BACKGROUND

Congenital diaphragmatic hernia (CDH) is a serious condition that results in lung hypoplasia. There exists a preferential association between CDH and congenital bronchopulmonary defects.

OBJECTIVES

To describe a series of 4 pregnancies with congenital diaphragmatic hernia (CDH) associated to lung lesions: Congenital Cystic Adenomatoid Malformation (CCAM) and Broncopulmonary Sequestration (BPS).

MATERIALS & METHODS

Retrospective review of 4 pregnancies referred to our Fetal Medicine Unit with a final diagnosis of CDH and lung lesions. Initial diagnosis, GA at referral, imaging findings, prenatal diagnosis, perinatal outcomes and postnatal diagnosis are described. The evaluation at our Unit included detailed anatomical ultrasound, fetal ecocardiogram and MRI.

RESULTS

- Between 2002 and 2013, 94 cases of prenatal diagnosis of CDH, and 46 of lung lesions (CCAM and/or BPS) were evaluated at our Unit.
- Four cases of association between CDH and bronchopulmonary defects were observed (2 cases with CCAM, 1 case with BPS and one case with CCAM and BPS).
- Gestational age at diagnosis was between 24 and 31 wks.
- All were left sided CDH and 3/4 had liver herniation.
- O/E LHR were between 28 and 38% in all cases.
- No fetuses developed hydrops or required in utero intervention.
- Gestational age at delivery was 39 weeks in all cases.
- ECMO was utilized in 2/4 neonates.
- Three fetuses died in the first month of life and one was alive at two years.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>GA at dx (weeks)</th>
<th>Liver position</th>
<th>O/E at dx</th>
<th>GA at delivery (weeks)</th>
<th>ECMO</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>L CDH - CCAM</td>
<td>26</td>
<td>Down</td>
<td>37%</td>
<td>39</td>
<td>No</td>
<td>Alive at two years</td>
</tr>
<tr>
<td>L CDH - BPS</td>
<td>31</td>
<td>Up</td>
<td>28%</td>
<td>39</td>
<td>No</td>
<td>Deceased</td>
</tr>
<tr>
<td>L CDH - BPS</td>
<td>24</td>
<td>Up</td>
<td>30%</td>
<td>39</td>
<td>Yes</td>
<td>Deceased</td>
</tr>
<tr>
<td>L CDH – CCAM - BPS</td>
<td>25</td>
<td>Up</td>
<td>38%</td>
<td>39</td>
<td>Yes</td>
<td>Deceased</td>
</tr>
</tbody>
</table>

CONCLUSIONS

In our series, 4 % of prenatally diagnosed CDH were associated to ipsilateral bronchopulmonary lesions. In none of them the association was initially detected. CDH and CCAM/BPS are mutual differential diagnosis in the prenatal assessment of thoracic lesions. Thus, the coexistence of both defects makes a correct prenatal diagnosis a real challenge.

REFERENCES:


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