Two cases of congenital rectus diastasis with Kagami-Ogata syndrome.

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**Introduction**

Congenital rectus diastasis is well misidentified as a large omphalocele due to the similar image of extraction of liver and intestine with attached cord vessels. Kagami-Ogata syndrome (KOS) is a rare disease with a paternal uniparental disomy 14. It has been known as a small bell-shaped thorax with coat-hanger appearance of the ribs on X-ray after birth, however polyhydramnios and abdominal wall defects are clinical features prenatally. We present two cases of KOS which were prenatally diagnosed polyhydramnios and atypical omphalocele, eventually diagnosed rectus diastasis.

**Cases**

Case 1: 37 y.o., gravida 2 para 2, woman showed polyhydramnios and an excessive growth (+2.3SD) around 20 weeks. Fetal ultrasound revealed large omphalocele with liver protrusion and vascular ring (Fig. 1). Chromosomal analysis revealed 46, XY. Due to progressive threatened preterm labor, cesarean section was performed at 27+6 weeks. With the features of face and chest Xp, KOS was highly suspected.

Case 2: 28 y.o. primi gravida was referred due to polyhydramnios and omphalocele (Fig. 1). Chromosomal analysis was 46, XX. Since the uterine contraction was hard to control, cesarean section was performed at 27+4 weeks. Other clinical features such as facial expression and gene analysis diagnosed as KOS.

**Table 1: Clinical outcomes of KOS in these cases**

<table>
<thead>
<tr>
<th>Case</th>
<th>AFI (cm)</th>
<th>Amniorr. reduction (times)</th>
<th>Mode of delivery</th>
<th>GA at birth (w)</th>
<th>Apgar score</th>
<th>BW (g, SD)</th>
<th>BH (cm)</th>
<th>Duration of intubation (days)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>40.8</td>
<td>3</td>
<td>eCS</td>
<td>27-6</td>
<td>1/5</td>
<td>1735 (+3.3)</td>
<td>38</td>
<td>104</td>
</tr>
<tr>
<td>Case 2</td>
<td>27.2</td>
<td>0</td>
<td>eCS</td>
<td>27.4</td>
<td>1/4</td>
<td>1594 (+3.7)</td>
<td>33</td>
<td>120</td>
</tr>
</tbody>
</table>

Table 1: Clinical outcomes of KOS in these cases

**Discussion**

① Prenatal clinical features of KOS patients have been shown as below.

- **Polyhydramnios**
  - Thoracic and abdominal abnormalities (Omphalocele was identified in about 1/3 of all patients, and rectus diastasis was found in the remainings.)
- Small bell-shaped small thorax with coat-hanger appearance of the ribs could be identified after birth

With these features, KOS should be differentiated.

② Differentiation of rectus diastasis and omphalocele
- Identify the cord insertion with color Doppler
- Angle of liver herniation or the presence of Wharton’s jelly

**Conclusions**

Although KOS is a rare disease with many clinical features, polyhydramnios and abdominal abnormalities would be a key sign for prenatal diagnosis. Prenatal evaluation of cord insertion with color Doppler might be helpful to distinguish between omphalocele and rectus diastasis.