Limb body wall complex associated with placenta accreta: a mere coincidence or a sign of an etiopathogenic link?

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Background

- Limb Body Wall Complex (LBWC) is a rare and lethal congenital anomaly characterized by a spectrum of multiple deformities. The typical finding is the deformed closing of the thoracic-abdominal wall associated with limb deformities, kyphoscoliosis and short or absent umbilical cord. In addition, craniofacial deformities, single umbilical artery, intestinal atresia and spina bifida can be observed.
- The cause of the LBWC is unknown, but three theories are more accepted: intrinsic disorder of the ectodermal placodes, early amniotic rupture theory, and vascular disruption theory and tissue hypoxia.
- The pathogenesis of the placenta accreta is not fully understood either, although it is believed that a strong association exists between decidualization and vascularization damage in uterine scar areas.

Case Report

- Multigravida, 27 years old, history of four previous C-sections. The ultrasound scan assessment at 32 weeks showed a fetus with 1300 grams and an extensive open anterior abdominal wall defect, kyphoscoliosis and lower limbs displaced in relation to fetal trunk. Liver and small bowel were observed outside the abdominal cavity close to the placenta (figures 1A, 1B). The echocardiography showed double outlet right ventricle and ventricular septal defect.
- Complete placenta previa was observed filled with many vascular lacunae. The magnetic resonance imaging confirmed the ultrasound findings (figure 2). Elective C-section followed by total hysterectomy was performed at 34 weeks.
- The infant weighed 1900 grams with undefined sex. The death occurred within minutes after birth. Great thoracic-abdominal deformity was observed with amelia of the right upper limb (figure 3).

Discussion

- The closeness between the abdominal organs and placenta confirmed the non-obliteration of the extra-embryonic coelom. The umbilical cord was very short. No amniotic bands were observed. The placenta increta was confirmed by anatomopathological examination of the uterus.
- The association between LBWC with placenta accreta observed here shows that the vascular disruption theory can be considered one of the main causes.
- It is believed that the decidual hypoxia that provoke the anomalous invasion of the trophoblast also reach the embryo.
- Due to the descriptive nature of this study, it cannot be affirmed that a causal relation exists between poor blood perfusion of the decidua and LBWC.
- As placenta accrete is a complication strongly associated with uterine scar, LBWC also can be associated with this risk factor.