Placento-cranial adhesion: A report of two cases

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CASE REPORTS

Case 1
A preterm, eight hour-old male with cranio-facial deformities was born normally at a hospital to a G, P, 22 year-old mother on June 24, 2006. The mother had no significant antenatal history. The child weighed 1.9 kg including the placenta, which was adherent to the head. Also evident were associated exencephaly, cleft of face, lip and palate and bilateral anophthalmos.

Constriction marks over the right arm, hypoplasia of the right radius and ulna and oligodactyly in both hands were also present [Figure 1]. There were no thoracic or abdominoschisis and both lower limbs as well as other systemic examinations were normal. He survived for only 48 h after birth.

Case 2
A full-term male was born to a G, P, 25 year-old mother by lower segment Caesarean section (LSCS) performed due to obstructed labour on August 27, 2006. Antenatal history was nothing significant. He weighed 2.5 kg including placenta, which was adherent to his head. Also evident were associated exencephaly and cleft of lip and palate on the right side [Figure 2]. He also had multiple constriction marks over both lower limbs and associated bilateral congenital talipes equino varus (CTEV). Other systemic examinations were normal. He survived for only 12 h after birth.

DISCUSSION

Limb body wall complex (LBWC) is a rare, complicated, polynamalformative fetal malformation syndrome with the essential features of: (a) exencephaly / encephalocele with facial clefts, (b) thoraco- and / or abdominoschisis and (c) limb defects.[1] There is no sex or familial predilection and two phenotypes have been described as placento-cranial and placento-abdominal adhesions.[1] The LBWC is also described as body stalk anomaly,
Limb body wall complex (LBWC) is a rare congenital anomaly characterized by a disruption of the cranio-facial structures during early gestation. It is often associated with other severe structural defects, and the prognosis for affected fetuses is poor. Early antenatal diagnosis and medical termination of the pregnancy are essential. The risk of recurrence being negligible, the prognosis for a later pregnancy remains positive.

In a fetopathologic evaluation with amniotic bands, Moerman et al. described three types of lesions: (1) constructive tissue bands, (2) amniotic adhesions and (3) more complex lesions such as LBWC. Most of the cranio-facial defects (encephalocele and/or facial clefts) occurring in these fetuses are the result of a vascular disruption sequence with or without cephalo-amniotic adhesion.

The diagnosis of this entity is based on the presence of two of the three features described above. Ultrasonographic detection of abdominoschisis, scoliosis, a single umbilical artery, a short umbilical cord, an extremely elevated level of maternal serum alpha-fetoprotein and abnormalities of the lower extremities are the keys to early diagnosis.

Major structural defects associated with LBWC include limb defects, scoliosis, craniofacial defects, cardiac defects, bowel atresia, renal agenesis/dysplasia, urogenital anomalies, absent or short umbilical cord etc. LBWC should be differentiated from more common anterior abdominal wall defects such as gastroschisis, omphalocele, etc.

As the existence of a fetus with LBWC is always endangered and universally fatal; it necessitates an early antenatal diagnosis and medical termination of the pregnancy. The risk of recurrence being negligible, the prognosis for a later pregnancy remains positive. Both of our patients showed the placental-cranial phenotype. Neither any further investigations nor postmortem examinations have been done in these two cases because of the fatal nature of the disease.

**REFERENCES**


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