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Limb Body Wall Complex: A Case Report

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Abstract
Limb body wall complex (LBWC) is a rare clinicopathological entity in which a abdominal wall and limb defect are major fetal anomaly. Very few cases have been reported in literature. Early and appropriate pre natal diagnosis is necessary for timely management. We report a case of LBWC in antenatal scan with congenital absence of lower limb.
Keywords: *Limb body wall complex, serinomelia.*

Introduction

LBWC is a rare polymalformative fetal syndrome which has been characterized by a wide spectrum of anomaly. Van allen et al.[1] criteria which has been traditionally used for the diagnosis has three major criterias out of which atleast two should be present.

1. Exencephaly/ encephalocele and facial cleft
2. Thoraco- and/or abdominoschisis; and
3. Limb defects

The incidence at birth is about 0.21-0.31 per 10,000 deliveries since majority of fetus affected with LBWC undergo intrauterine death[2,3]. Early antenatal ultrasonography helps in diagnosing the anomaly. Since there is no cure for LBWC the termination of the pregnancy is only option till date.

Case Report

A 26 year old primigravida female with 5 months amenorrhea who was referred to our institute for a second opinion after antenatal scan which was done 3 days prior to date of presentation in outside hospital and mentioned gastroschisis. She had normal blood profile and O positive blood group, Hb 10.6 gm/dl. Patient was referred to department of radiodiagnosis for reassessment of antenatal ultrasonography. The scan showed normal head with normal biparietaldiameter (BPD) and head circumference (HC) which corresponded to 19 week 1 day gestation period. However, a large abdominal wall defect with herniation of liver, stomach, bowel loops through it in the amniotic cavity was seen (fig 1,2). Spine was deformed with kyphoscoliotic changes (fig 1). Further study revealed serinomelia /unilateral absence of lower limb with polydactyly involving both hand and

foot (fig 4). Thorax was found to be small in size compressing the heart. Few parts of fetus appear to be intracoelomicon USG scan. Color Doppler showed short and two vessel umbilical cord with single umbilical artery (fig 3). No anomaly was detected in face, eye, orbits, noseand lips. Anterior thoracic wall was found to be normal. No chromosomal anomaly was seen in the karyotyping examination. There was no previous history of any drug intake expect for Iron and folic acid supplementation. There was no medical history of hypertension or diabetes. The patient was counseled regarding the fatal outcome of the pregnancy and was referred back to the department of Obstetrics and gynecology for termination of pregnancy. Finding of the ultrasonography was confirmed after the delivery by the obstetrician.

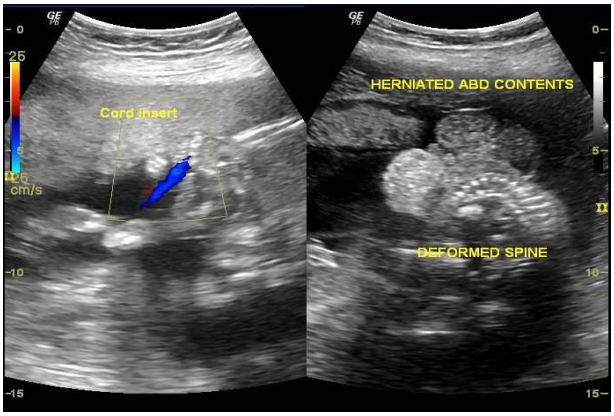

Fig 1: Ultrasound image showing deformed spine and large abdominal wall defect with herniated abdominal content

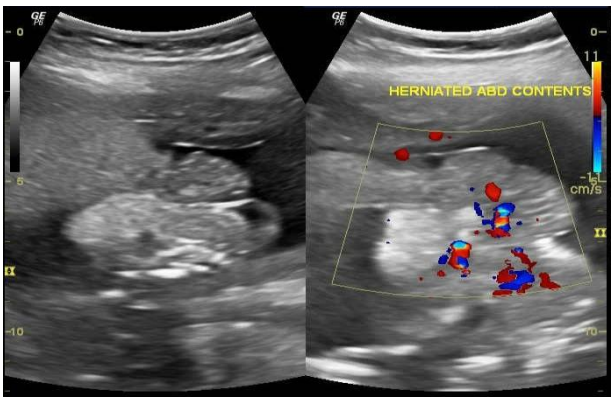

Fig 2: Ultrasound image showing large abdominal wall defect with herniated abdominal content

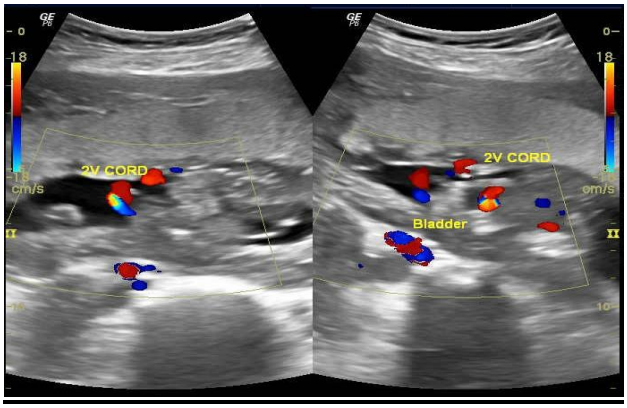

Fig 3: Ultrasound image showing two vessel umbilical cord (with absence of one umbilical artery)

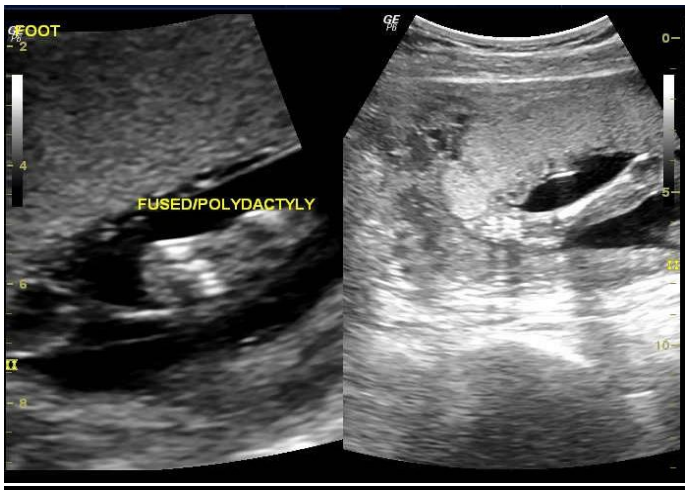

Fig 4: Ultrasound image showing right lower limb with polydactyl

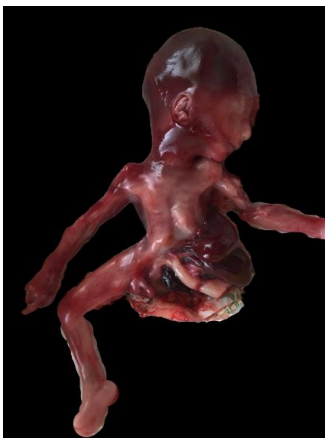

Fig 5: Gross image of abortus showing congenital absence of left lower extremities

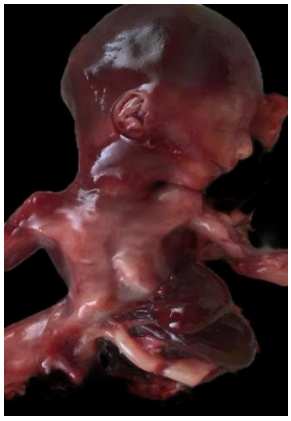

Fig 6: Gross image of abortus showing large abdominal wall defect with herniation of abdominal content and normal thoracic wall

Discussion

LBWC was described for the first time by Van Allen et al.[1] in 1987. It is also known by the other names like "Body stalk anomaly" "Congenital absence of umbilical cord" and "cyllosomus and Pleurosomus"[4,5] The diagnostic criteria for LBWC is still debatable, the commonly used one was put forth by Van Allen et al. in1987 as stated earlier.

Congenital malformations of the ventral abdominal wall occur in many forms, ranging from exomphalos to gastroschisis to more complex malformations, such as pentalogy of Cantrell and LBWC.

The most accepted theory is early embryonal dysplasia put forward byHartwig et al.[6] in 1989. According to this, there will be an abnormal embryonic folding related to malfunctioning of the body wall ectodermal placode. This leads to defective closure of embryonic abdominal wall, umbilical abnormality and persistence of extra embryonic coelom communicating with the abdominal cavity. These ectodermal placodes also add cells to mesoderm of the trilaminar embryonic disc, which is destined to form the genitourinary tract.

This anomaly does not have any sex predilection and recurrence of this condition is observed in two families suggesting a possible genetic etiology.[7]

There are two distinct phenotypes of LBWC described by Russo et al.[8] the placentocranial adhesion phenotype which may comprise craniofacial defect, facial cleft, amniotic adhesion and amniotic band sequence. The second is the placentoabdominal adhesion phenotype in which there is no craniofacial defect but may have imperforate anus, urogenital abnormalities, lumbosacral meningomyelocele and kyphoscoliosis. In our case report the presentation looks like the placentoabdominal phenotype.

Conclusion

LBWC is a lethal anomaly hence it has to be differentiated from treatable causes like omphalocele or gastroschisis, but sometimes oligohydramnios may mask the underlying anomalies and therefore, challenge the radiologist.

Conflict of Interest: None

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