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Case Report

Prenatal sonographic diagnosis of limb-body wall complex: case series of a rare congenital anomaly

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ABSTRACT

Three case reports of a rare congenital anomaly "limb-body wall complex" also known as "body stalk syndrome" are presented with prenatal ultrasonographic diagnostic features, immediate after delivery evaluation, and histopathologic analysis.

Introduction

Limb-body wall complex (LBWC) is a rare, complicated, polymalformative fetal syndrome with essential features of:

(1) Exencephaly and/or encephalocele with facial clefts,
(2) Thoraco and/or abdominoschisis, and
(3) Limb defects [1].

The sonographic hallmarks of LBWC are neural-tube abnormalities, severe scoliosis, positional deformities, and abnormalities of fetal membranes [2]. Generally, the diagnosis is based on any 2 of the 3 previously mentioned features. Two adhesion phenotypes have been described, the "placentocranial" and "placentoabdominal." LBWC is also known as "body stalk syndrome." Unfortunately, there is no cure for LBWC, and it is generally considered to be incompatible with life (fatal) [3]. The poor prognosis of LBWC necessitates an early antenatal diagnosis and termination of pregnancy.

Case reports

Case report 1

A 26-year-old primigravida female with a 7-month amenorrhea was referred to our department for routine ultrasonography and fetal well-being study. She had normal blood

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profile and O +ve blood group. Hemoglobin was 10.5gm/dL. The ultrasonography revealed normal head with biparietal diameter (BPD) corresponding to 26 weeks 3 days. However, a large abdominal wall defect, with liver and gut coils herniating through it, into the liquor amnii was seen. The herniated organs formed an entangled complex covered by fetal membranes. Spinal dysraphism along with a large meningocele was also visualized in the lumbosacral region (Fig. 1A). Further study revealed bilateral clubfoot deformity in the fetus (Fig. 1B). Color Doppler study showed single umbilical artery supplying the liver (Fig. 1C). No anomaly was detected in the eyes, palate, lips, face, and thoracic region. The female was referred back to department of Gynecology and Obstetrics for further management with diagnosis of LBWC. On counseling, the patient about the fatal outcome of the anomaly, the pregnancy was terminated and ultrasonography findings confirmed thereof (Fig. 1D and E). Histopathologic analysis of the umbilical cord revealed single umbilical artery (Fig. 1F) and hypoplastic right internal iliac artery.

Case report 2

A 22-year-old G_2P_1 female with a 6-month amenorrhea was referred for fetal ultrasonography to the Radiology department. The blood profile revealed decreased Hemoglobin (8.5%), A +ve blood group and normal coagulation profile. The ultrasonography revealed gestational age of 22 weeks using BPD. Furthermore, it revealed a large abdominal wall defect

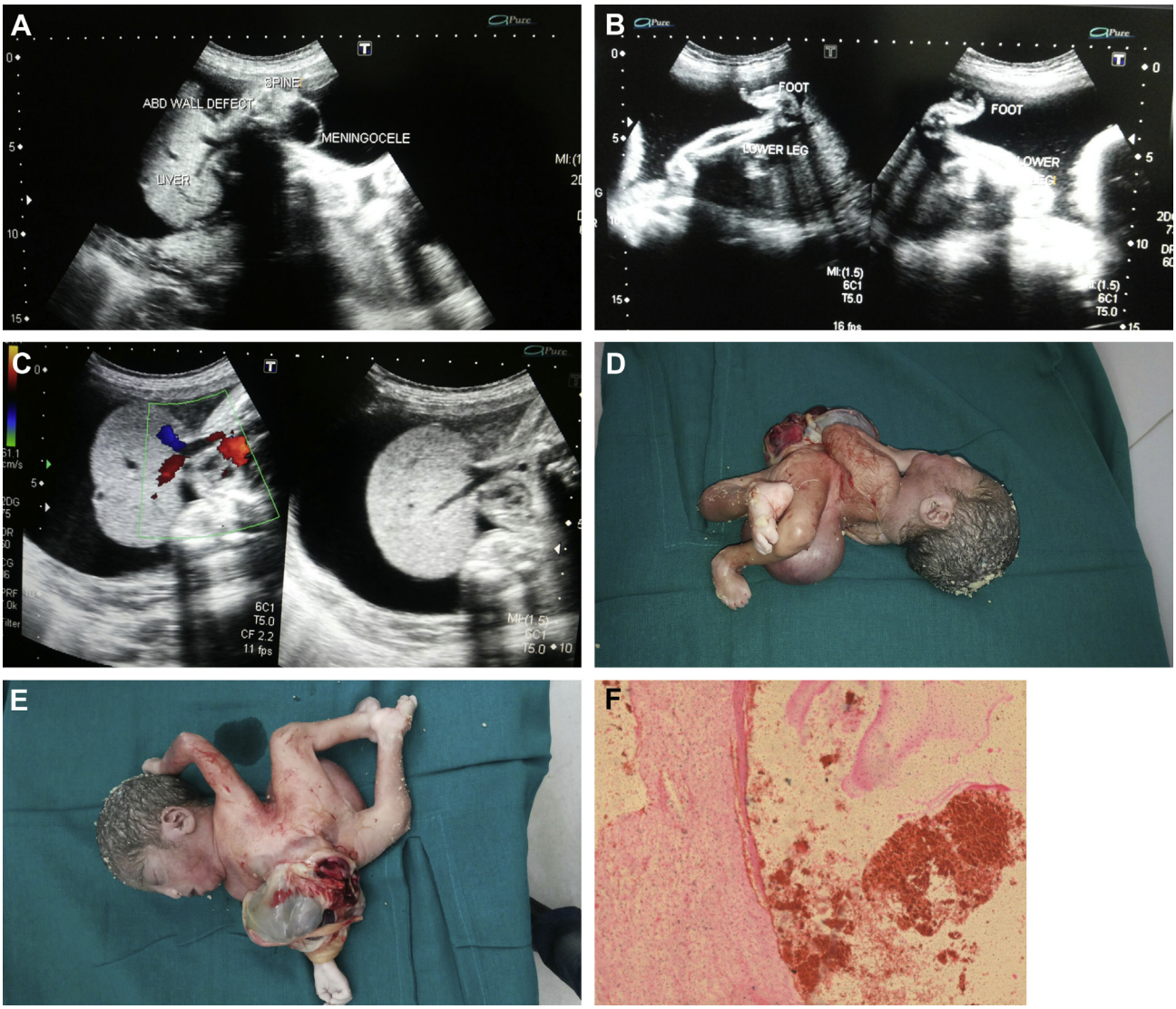

Fig. 1 – (A) Longitudinal view of ultrasound image showing the large abdominal wall defect with liver and gut-coils herniating through it into the liquor amnii. Also depicted is the meningocele in lumbosacral region and spinal dysraphism. (B) USG images demonstrating bilateral clubfeet deformity. (C) Color Doppler image showing the blood flow into liver of the fetus via the single umbilical artery. (D) Immediate post delivery image showing the large meningocele, herniated abdominal contents through abdominal wall defect, spinal dysraphism and club foot deformity is also seen confirming USG findings. (E) Post delivery image showing the large abdominal defect through which abdominal organs are seen herniating. Sex of the fetus is not distinguished. (F) Histopathology slide showing single umbilical artery with RBC's in the lumen.