J Cases Obstet Gynecol, 2015;2(4):75-77

Case Report

Case series of three patients of fetal limb body wall complex

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Abstract

Limb body wall complex (LBWC) is a rare fetal polymalformation of unknown etiology .The general incidence is less than 1 in 14,000- 22,000 pregnancies. We report 3 cases of LBWC identified at our institute within a span of 2 years. Total pregnancies seen were 2989, all of which underwent a detailed prenatal ultrasound imaging. Of these, 23 were anomalous fetuses/babies. All three cases were diagnosed prenatally on ultrasound .All were placenta- abdominal type of LBWC. Two of them had large abdominoschisis with single lower limb and scoliosis. The third fetus had a large omphalocele with right congenital talipes equinus varus and an amniotic band. All three pregnancies were terminated.

Key words:

Limb body wall complex, congenital anomaly, fetal polymalformation, rare fetal anomalies

Introduction

Limb body wall defect is an extremely rare fetal congenital abnormality. It is lethal and complex. It is very difficult to identify on prenatal ultrasound because several differential diagnosis exist including gastroschisis and omphalocele [1]. Until today its etiology remains unclear [2]. Knowledge about this condition among common clinicians remains scanty, making it a less identified and less reported congenital abnormality. We encountered three cases of fetal LBWC at our institute over a short span of two years.

Case presentation

Patient 1: A 24-year-old unbooked patient came to our hospital for her first antenatal visit at 5 months gestation. She suffered a spontaneous abortion in the past in a remote rural place in India for which she had no medical consultation. Hence, no records of that pregnancy are

Article history:
Received 30 01 2015
Accepted 21 04 2015

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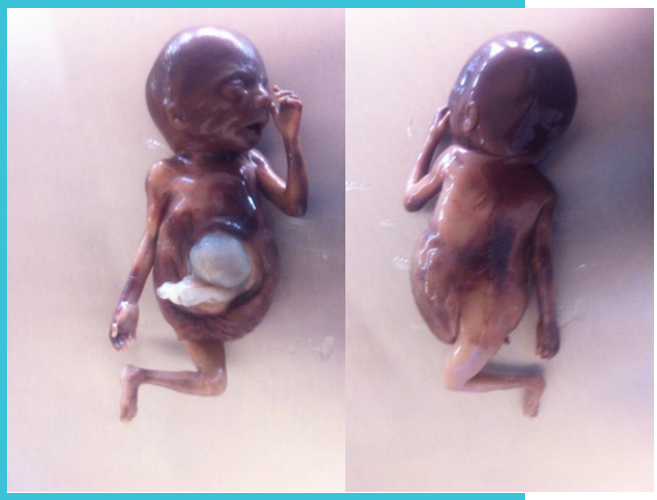
Figure 1.

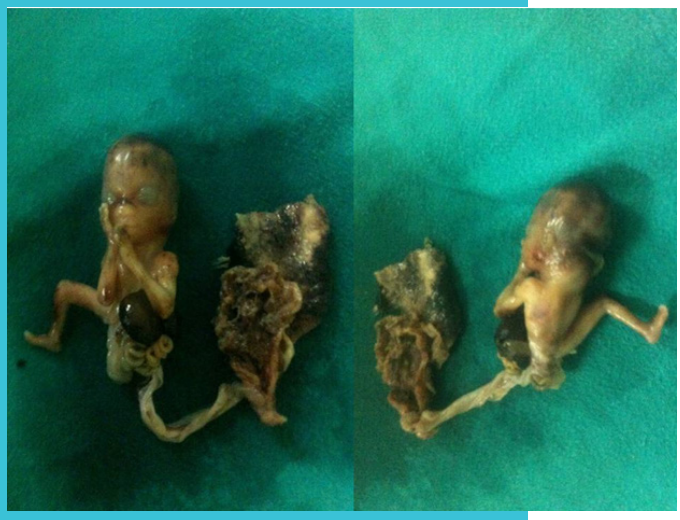


Photograph of anterior view of aborted fetus of patient 1 showing abdominoschisis with absent right lower limb and marked scoliosis (A) and photograph of posterior view of aborted fetus (B).

available for discussion. She did not suffer from any disease or medical problems in the past,had no previous visits at any hospital for any reason. She had no family history of any medical disorder or congenital anomalies. She also had no history of any kind of substance abuse. When a Targeted Imaging for Fetal Anomalies (TIFFA) was done, we received a report which showed multiple fetal malformations including gastroschisis, absent right lower limb. We terminated the pregnancy according to 'eugenic indication' of Medical Termination of Pregnancy (MTP) Act of India, 1971 rule. Labor was induced with Tablet Misoprostol 50 µg vaginally and complete expulsion of fetus and placenta took place. She aborted a male fetus weighing 150 grams with similar features as seen on ultrasound with an additional finding of scoliosis (Figure 1). Postmortem of the fetus confirmed these findings.

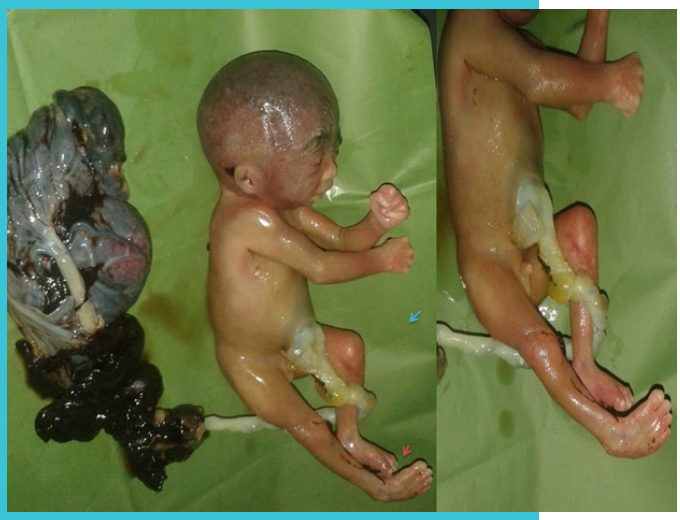
Patient 2: A 21-year-old primigravida who was a regular booked case at our hospital was advised a nuchal translucency scan. Her ultrasound also revealed similar defects in the fetus including absent left lower limb and a large abdominoschisis with bowel and liver extrusion (Figure 3). We terminated this pregnancy also and she again delivered a fetus with absent left lower limb and left gastroschisis. The umbilical cord was not involved. There were no discernible external genitalia. There was anal atresia (Figure 2). Autopsy was not done as the patient's relatives did not give the consent to do so, although karyotyping revealed a 46XY pattern.
Patient 3: A 25- year-old-second gravida came to us during her first visit for an anomaly scan at 20 weeks gestation which revealed right congenital talipes equinus varus, a large omphalocele, choroid plexus cyst and an amniotic band. The pregnancy was terminated. She delivered a male fetus of weight 600g and all the findings completely correlated with the ultrasound findings (Figure 4). Autopsy was not done as the patients relatives did not give consent to do so.

Figure 2.



Photograph of anterior view of aborted fetus of patient 2 showing large abdominoschisis with evisceration of liver and bowel, anal atresia and absent external genitalia and absent left lower limb (A) and photograph of posterior view of fetus showing short umbilical cord partially covered by amniotic membrane attached to the eviscerated internal organs directly (B).

Figure 3.



Photograph of fetus of patient 3 showing omphalocele with right congenital talipes equinus varus with placenta and amniotic band.

Discussion

Limb body wall defect is a complex and lethal fetal malformation. It is diagnosed when the fetus has any 2 of the 3 features regarding (i) encephalocele/exencephaly with facial cleft, (ii) thoraco/abdominoschisis, (iii) limb defect [3]. There are two types of LBWC: Placento-cranial type which involves upper limb defects with thoracoschisis or exencephaly/encephalocele and placento-abdominal type which involves lower limb defects with abdominoschisis. They can be associated with scoliosis, short umbilical cord, neural tube defects, internal organ defects like absent diaphragm, bowel atresia and renal dysplasias [4]. LBWC can be diagnosed prenatally by ultrasound and elevated alpha-fetoprotein [5].

In our institute, 2989 pregnancies were seen over a two year span from December 2012 till December 2014. All patients underwent a detailed prenatal ultrasound at our institute. A total of 23 anomalous fetuses/ babies were identified and delivered of which 3 fetuses were diagnosed with limb body wall complex. A doubt arises if LBWC is commoner than we think. Its incidence has been reported traditionally in literature as 14,000 – 22,000 pregnancies [5,6]. Others reported higher incidence as 1 in 4000 pregnancies [3] and 1 in 7500 pregnancies [7].

Most of them get diagnosed in the prenatal period on ultrasonography and are not carried on till birth. The incidence at birth is much lesser is therefore about 11/428,599 births [8]. Probable underreporting of LBWC is occurring because its diagnosis is not being established easily. This could be due to limited awareness regarding the condition and limited availability of research material to refer to. Standard obstetrics textbooks do not include much information on LBWC. It is important for clinicians to expand their knowledge on LBWC because it seems now that LBWC is more common than we believe. The other possibility is a rise in the incidence of LBWC. If this is true it is difficult for us to evaluate the cause because little is known about its etiology.

No clear etiology could be designated to this condition although several theories have been postulated like disruption of embryonic vessels, early amnion rupture amniotic bands that amputate limbs and malfunction of body wall ectodermal placode that can cause defective embryonal folding leading to body cavity defects. Some researchers have also spoken of vasoconstrictive agents like cocaine, nicotine to cause LBWC. No risk factors or teratogenic factors could be identified in all these patients. All of them were sporadic in origin.

In conclusion, LBWC is a rare fetal malformation that is incompatible with life. Little is known about its true incidence, etiology or pathogenesis. Clinicians should make an effort to report every case of LBWC they encounter in order to establish its real incidence. Widespread clinical material and inclusion about this condition in standard obstetric textbooks are required to increase its awareness among clinicians to avoid misses in the diagnosis.

Conflict of Interest
None

References

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