

Case Report

Volume 1 Issue 1 - January 2016
DOI: 10.19080/AJPN.2016.01.555553

Acad J Ped Neonatol

Copyright © All rights are reserved by Pace Carlotta

A Monoamniotic Pregnancy Discordant for Limb Body Wall Complex and Complicated by Acute TTTS

Pace Carlotta^{1*}, Masturzo Bianca¹, Gaglioti Pietro¹, Viora Elsa², Sciarrone Andrea², Botta Giovanni³, Todros Tullia¹

¹2U Gynecology and Obstetrics Unit, Sant'Anna Hospital, Italy

²Ultrasound and Prenatal Diagnosis Unit, Sant'Anna Hospital, Italy

³Gynecological and Obstetrical Pathology Unit, Sant'Anna Hospital, Italy

Submission: November 24, 2015; **Published:** January 05, 2016

***Corresponding author:** Pace Carlotta, Sant'Anna Hospital, Città della Salute e della Scienza, Via Ventimiglia 3, 10126, Turin, Italy, E-mail: pace.carlotta@gmail.com

Abstract

We are reporting a case of monoamniotic twin pregnancy discordant for Limb Body Wall Complex (LBWC), complicated by acute twin-to-twin transfusion syndrome. The twins were delivered by an emergency cesarean section at 29+6 weeks' gestation because of persistent fetal tachycardia of the unaffected twin. The phenotypically normal twin was anemic at birth (donor twin), while postmortem examination revealed a condition of overloaded fetal circulation of the LBWC twin (recipient twin). This is, to our knowledge, the first case of acute TTTS in a monoamniotic pregnancy reported in Literature. In monoamniotic pregnancies discordant for LBWC careful fetal monitoring should be granted, as the presence of the malformation masks the classic signs of twin-to-twin transfusion syndrome.

Keywords: Limb Body Wall Complex; Monoamniotic Pregnancy; Acute Twin to Twin Transfusion Syndrome

Abbreviations: LBWC: Limb Body Wall Complex; TTTS: Twin to Twin Transfusion Syndrome; GA: Gestational Age; AC: Abdominal Circumference; MCA: Middle Cerebral Artery; bpm: Beats per Minute

Introduction

Limb-body wall complex (LBWC) is a rare and lethal fetal malformation (1/7500 pregnancies, 1/14000 births [1]), characterized by a large abdominal wall defect with rudimental or absent umbilical cord and abdominal placental attachment. An amniotic sheet connects the skin margin of the ventral wall defect to the placental surface, forming a tube containing the eventrated organs. Severe kyphoscoliosis, limb deformities and several other structural anomalies are possibly associated. As limb-body wall complex is invariably fatal, it requires accurate diagnosis and appropriate counseling. In most cases termination of pregnancy is chosen. Of particular concern are those cases in which LBWC presents as a discordant malformation in monochorionic pregnancy. A review of the literature (1965-2013) reports 5 cases of monochorionic pregnancies discordant for LBWC, including 3 monoamniotic pregnancies [2,3]. We are reporting a case of monoamniotic twin pregnancy discordant for LBWC, complicated by acute twin-to-twin transfusion syndrome (TTTS).

Case Report

A 33 year-old woman, gravida 1, para 0, was referred to our center at 16 weeks' gestation because of a monochorionic monoamniotic twin pregnancy discordant for complex fetal anomaly. At the first ultrasound examination, one twin appeared to be growth-restricted but anatomically normal; the other twin was confirmed to have multiple malformations (severe scoliosis, small thorax, wide anterior wall defect with abdominal placental attachment, ventriculomegaly). The diagnosis of limb body wall complex was made and the patient was apprised of the negative outcome of the malformation and of the risks for the other twin. The hypothesis of selective foeticide by fetoscopic cord ligation or laser coagulation was excluded as technically impossible. Serial fetal ultrasound examinations confirmed growth restriction for the anatomically normal twin, with AC below 5th centile but normal umbilical artery, ductus venosus and MCA Doppler velocimetry. At 27 weeks' gestation the patient was admitted to hospital because of Clostridium difficile infection. She was administered antibiotic therapy and corticosteroid

prophylaxis for neonatal respiratory distress syndrome.

Fetal heart rate of the anatomically normal twin was monitored by cardiotocography three times a day, upon visualization of the other twin's heart beat by ultrasound. At 29+6 weeks' gestation fetal monitoring turned pathological for the anatomically normal twin, recording persistent fetal tachycardia at 190-200 bpm. An emergency cesarean-section was performed. The first twin was a phenotypically normal male, weighing 980 g, with Apgar scores of 6 at 1 minute and 9 at 5 minutes with ventilatory support. Neonatal haemoglobin at birth was 11 g/dl. He was admitted to neonatal intensive care unit and had a good outcome. The second twin was extracted together with the placenta, from which he could not be detached, and died in a few minutes. Post-mortem examination revealed marked vascular congestion of all the examined organs and abnormal dilation of hepatic centrilobular veins (Figure 1), stating a condition of overloaded fetal circulation. All of these features strengthened our hypothesis of this fetus being the recipient in an acute form of twin to twin transfusion syndrome. Histologic examination of the placenta showed typical features of TTTS as they are reported in Literature [4,5]: edematous villi with delayed maturation and small bloodless vessels (donor's villi, Figure 2) coexisted with mature villi with congested vessels (recipient's villi, Figure 3).

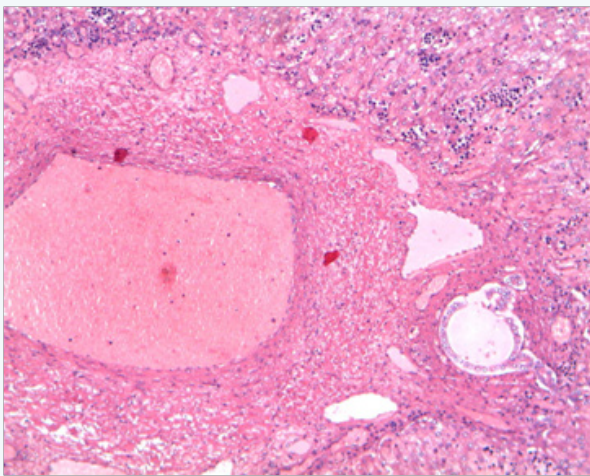


Figure 1: Hepatic tissue section of the LBWC twin, showing vascular congestion and abnormal dilation of hepatic centrilobular veins, stating a condition of overloaded fetal circulation (recipient twin).

Discussion

Monochorionic twin pregnancies represent 20% of multiple pregnancies and about 1.5% is monoamniotic [6]. Monoamniotic pregnancies are at the greatest risk of perinatal mortality and morbidity (death rate around 15% [7,8]), as they share all risks of monochorionic pregnancies (prematurity, structural anomalies, TTTS), but are also at risk for cord entanglement [9-11]. TTTS

has a reported incidence of 10% in monochorionic pregnancies and a lower incidence in monoamniotic pregnancies (3-9%) [12]. The diagnosis of TTTS, according to Quintero's staging system [13], is based on the presence of an amniotic membrane separating the twins. In monoamniotic pregnancies this staging system cannot be applied and the diagnosis is established on the observation of polyhydramnios in the receiver, non-visualization of the bladder of the donor fetus, discordant biometry and abnormal Doppler velocimetry (as a sign of anemia). In this specific case the presence of LBWC prevented a correct evaluation of fetal biometry and the visualization of the bladder in the affected twin (a cloacal exstrophy was signaled at the autoscopic examination). Umbilical artery Doppler-velocimetry was not assessable because of abdominal-placental attachment in the affected twin, and it was normal in the co-twin.

We could only monitor MCA Doppler-velocimetry in both fetuses and we had no signs of chronic fetal anemia, neither polyhydramnios. Despite the absence of premonitory signs an acute fetal impairment developed, which we have been able to identify thanks to the intensive fetal monitoring we set up since 28 weeks' gestation. The diagnosis of acute TTTS was made post-partum on the basis of the observation of anemia in the surviving fetus (neonatal haemoglobin: 11 g/dl) and it was confirmed by post-mortem examination of the recipient fetus and placental histology (Figure 1-3). As far as we know, this is the first case of acute TTTS in a monoamniotic pregnancy reported in Literature. Most reported cases of TTTS are chronic forms. In a series of 241 monochorionic pregnancies, the reported incidence of acute TTTS was 2.5%: all cases occurred in diamniotic pregnancies which had undergone vaginal delivery [14]. It has been demonstrated that intensive fetal monitoring in monoamniotic pregnancies reduces perinatal morbidity, mostly due to early recognition of fetal distress.

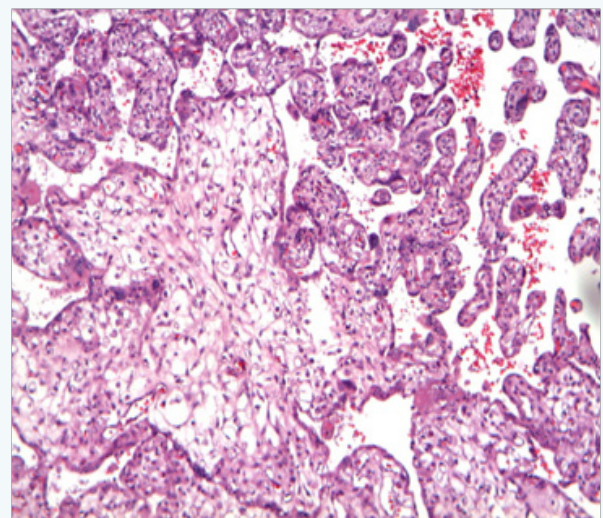


Figure 2: Section of the placenta, showing edematous villi with delayed maturation and small bloodless vessels (donor's villi).

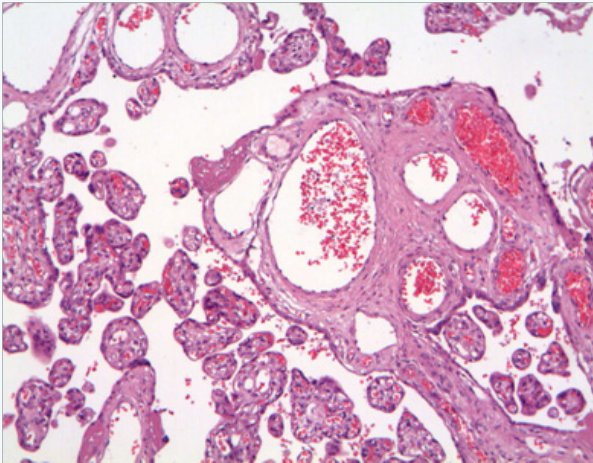


Figure 3: Section of the placenta showing mature villi with congested vessels (recipient's villi).

Heyborne et al. [15] suggest that even cord accidents are subacute events that can be prevented with strict monitoring. Similar speculation can be made regarding TTTS: the acute form of the syndrome might be subacute instead. Placental histology supports this theory, as villar modifications are unlikely to be acute. Intensive fetal monitoring in this case has highlighted the first signs of fetal distress and allowed prompt management. In conclusion, this case draws attention to specific issues related to the presence of LBWC in a monoamniotic pregnancy: on one side, the presence of this complex malformation nullifies the risk of cord entanglement; on the other side, the malformation itself masks the classic signs of twin-to-twin transfusion syndrome, thwarting the diagnosis of this rare but possible complication.

References

1. Spiller E, Salvador L, Bogana G, Neri F, Ambrosini G, et al. (2008) Body stalk anomaly: Management of two dichorionic-diamniotic pregnancies. *J Matern Fetal Neonatal Med* 21(10): 758-759.
2. Vidaeff AC, Delu AN, Silva JB, Yeomans ER (2005) Monoamniotic twin pregnancy discordant for body stalk anomaly: case report with nosologic implications. *J Ultrasound Med* 24(12): 1739-1744.
3. Daskalakis GJ, Nicolaides KH (2002) Monozygotic twins discordant for body stalk anomaly. *Ultrasound Obstet Gynecol* 20(1): 79-81.
4. de Laat MW, Manten GT, Nikkels PG, Stoutenbeek (2009) Hydropic placenta as a first manifestation of twin-twin transfusion in a mono chorionic diamniotic twin pregnancy. *J Ultrasound Med* 28(3): 375-378.
5. Sunagawa S, Kikuchi A, Kurihara N, Hiroma T, Ono K, et al. (2008) Mono chorionic twin fetuses showing a reversal of donor-recipient phenotypes in severe twin-twin transfusion syndrome without oligo-polyhydramnios sequence. *Congenit Anom (Kyoto)* 48(2): 92-96.
6. Dickinson JE (2005) Monoamniotic twin pregnancy: a review of contemporary practice. *Aust N Z J Obstet Gynaecol* 45(6): 474-478.
7. Hall JG (2003) Twinning. *Lancet* 362(9385): 735-743.
8. Hack KE, Derks JB, Schaap AH, Lopriore E, Elias SG, et al. (2009) Perinatal outcome of monoamniotic twin pregnancies. *Obstet Gynecol* 113(2 Pt 1): 353-360.
9. Lim KI, Dy C, Pugash D, Williams KP (2005) Monoamniotic twins discordant for anencephaly managed conservatively with good outcomes: two case reports and a review of the literature. *Ultrasound Obstet Gynecol* 26(2): 188-193.
10. Rossi AC, Prefumo F (2013) Impact of cord entanglement on perinatal outcome of monoamniotic twins: a systematic review of the literature. *Ultrasound Obstet Gynecol* 41(2): 131-135.
11. Murata M, Ishii K, Kamitomo M, Murakoshi T, Takahashi Y, et al. (2013) Perinatal outcome and clinical features of mono chorionic monoamniotic twin gestation. *J Obstet Gynaecol Res* 39(5): 922-925.
12. Morikawa M, Yamada T, Yamada T, Sato S, Minakami H (2012) Prospective risk of intrauterine fetal death in monoamniotic twin pregnancies. *Twin Res Hum Genet* 15(4): 522-526.
13. Quintero RA, Morales WJ, Allen MH, Bornick PW, Johnson PK, et al. (1999) Staging of twin-twin transfusion syndrome. *J Perinatol* 19(8 Pt 1): 550-555.
14. Lopriore E, Holtkamp N, Sueters M, Middeldorp JM, Walther FJ, et al. (2014) Acute peripartum twin-twin transfusion syndrome: incidence, risk factors, placental characteristics and neonatal outcome. *J Obstet Gynaecol Res* 40(1): 18-24.
15. Heyborne KD, Porreco RP, Garite TJ, Phair K, Abril D, et al. (2005) Improved perinatal survival of monoamniotic twins with intensive inpatient monitoring. *Am J Obstet Gynecol* 192(1): 96-101.