Limb Constriction Secondary to Pseudoamniotic Band Syndrome after Selective Fetoscopic Laser Surgery: Report of a Case with a Favorable Outcome

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Established Facts

- Twin-to-twin transfusion syndrome is a serious complication of monozygotic twins with monochorionic placentation.
- Few studies have addressed the issue of complications related to selective fetoscopic laser photocoagulation in twin-to-twin transfusion syndrome pregnancies.
- There are little or no data on the incidence of pseudoamniotic band syndrome.

Novel Insights

- In our report the surviving twin had surgical correction with excellent cosmetic and functional results presenting no major postnatal sequelae of PABS.

Key Words

Limb constriction • Pseudoamniotic band syndrome • Fetoscopic laser surgery • Twin-to-twin transfusion syndrome

Abstract

Pseudoamniotic band syndrome (PABS) is an iatrogenic complication that causes entanglement of fetal parts in a constrictive sheet of detached or ruptured amniotic membrane after an invasive procedure, namely amniocentesis, amnioreduction or septostomy in twins. The incidence and

risk factors for PABS after fetoscopy-guided laser have not been documented [Winer et al.: Am J Obstet Gynecol 2008;198:393.e1–393.e5]. We report a case of monochorionic diamniotic twin pregnancy submitted to selective fetoscopic laser photocoagulation for twin-to-twin transfusion syndrome at 16 weeks of gestation. The procedure was complicated by the death of one of the fetuses at 24 weeks of gestation. Moreover, the surviving twin was diagnosed postnatally with pseudoamniotic band syndrome, presenting with affected limbs. The newborn was submitted to surgical correction of these lesions with a successful outcome and was discharged on day 15.

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Fig. 1. Constriction ring on the upper left arm.

Case Report

A 32-year-old Caucasian woman, gravida 1, was referred to our Maternal-Fetal Unit at 9 weeks of gestation for spontaneous monochorionic diamniotic twin pregnancy.

At 12 weeks of gestation, the twins presented concordant crown-rump lengths and amniotic fluid indexes, with nuchal translucencies of 2.3 and 0.8 mm.

An ultrasound examination performed at 16 weeks of gestation revealed Quintero stage II twin-to-twin transfusion syndrome (TTTS). The parents were informed of the risks of TTTS and decided to submit to selective fetoscopic laser photocoagulation (SFLP) of vascular anastomosis. The procedure took place at 16 weeks and 3 days of gestation, at Hospital Clinic in Barcelona, and no immediate complications were noticed. At 17 weeks and 6 days, both twins were alive with concordant amniotic fluid indexes, visible bladders and umbilical artery pulsatility index on the recipient of 2.49 with absent diastolic waves, and on the donor of 1.68. However, at 24 weeks of gestation, demise of the recipient twin was diagnosed. Subsequent scans showed normal growth and activity of the surviving fetus, and pregnancy was otherwise uneventful.

At 39 weeks and 5 days of gestation, a cesarean section for cephalopelvic disproportion was performed, and a 3,910-gram boy was delivered with Apgar scores at the 1st and 5th minutes of 9 and 10, respectively. Physical examination showed a constriction ring on the upper left arm with edema of the distal limb, which was not necrotic (fig. 1), and multiple constriction rings involving the digits of both feet, with amputation of the distal phalange of the second digit of the right foot (fig. 2). Pathological examination of the placenta and amnion confirmed the diagno-

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Fig. 2. Digits of both feet with multiple constriction rings, with secondary amputation of the distal phalange of the second digit of the right foot.

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Fig. 3. Operated upper left arm on the 8th day of life.

sis of pseudoamniotic band syndrome (PABS). The placenta was morphologically compatible with gestational age. It presented significant villous sclerosis, perivillous fibrin deposition and microcalcifications, with maceration and mummification of the umbilical cord and amniotic membrane of the demised fetus, secondary to his death. The remaining placenta had no anomalies, except for chorioamnionitis and vasculitis of umbilical cord of the surviving twin. The pathology of the demised fetus, although limited by advanced state of mummification, showed no malformations and biometry was compatible with 18 weeks of gestation.

A web reconstruction of the upper left arm was performed via Z-plasty on the 8th day of life. The newborn was discharged on day 15. Functional recovery of the left arm and hand was confirmed after 1 month of follow-up (fig. 3).

Discussion

TTTS is a serious complication of monozygotic twin gestations, with an incidence that ranges from 10 to 15% of the monochorionic diamniotic gestations [2]. It is defined sonographically by the combined presence of polyhydramnios in one sac and oligohydramnios in the other [3]. Intertwin placental anastomoses are the basis for the onset of TTTS. Although intertwin transfusion is a constant and usually balanced phenomenon in monochorionic twins, a chronic imbalance in net flow can trigger TTTS [4].

TTTS is one of the most lethal perinatal complications, with an 80–100% mortality rate and a 15–50% risk of disability in survivors without treatment [5].

Therapeutic interventions in this disorder are all invasive, with success rates related to gestational age at diagnosis and treatment, severity of the disease at the time of treatment and type of intervention. These procedures include amnioreduction, septostomy, laser photocoagulation of the placental anastomoses, and umbilical cord occlusion by bipolar diathermy [6].

In the Eurofoetus randomized controlled trial, SFLP of the chorionic vessels was considered a more effective first-line treatment than serial amniodrainage in severe TTTS diagnosed before 26 weeks of gestation. SFLP was shown to result in at least one survivor at birth and intact survival at 6 months of age in 76 and 56% of the cases, respectively, as compared with 56 and 51%, respectively, in cases treated by amniodrainage [7]. Salomon et al. [8] prospectively studied the neurologic and neurodevelopmental outcomes up to the age of 6 years in infants included in the Eurofoetus trial, and concluded that SFLP therapy is associated with an almost 40% reduction in the risk of fetal death or long-term major neurological impairment and that infants treated in the SFLP group had higher ASQ scores at the age of 5 years (60 months).

Few studies have addressed the issue of complications related to SFLP in TTTS pregnancies. According to a recent review by Beck et al. [9], iatrogenic preterm prelabor rupture of membranes remains the most important complication of minimally invasive fetal procedures. Some techniques like amniopatch [10] have been attempted to reduce the rate of complications, with variable results. Little or no data are presented on the incidence of PABS, where there is entanglement of fetal parts in a constrictive sheet of detached or ruptured amniotic membrane after invasive procedures such as amniocentesis [11–13], amnioreduction [14], or septostomy

in twins [15]. In a smaller retrospective study by Habli et al. [16] involving 152 patients with TTTS who were treated with SFLP at 16–26 weeks of gestation, 5 cases (3.3%) had PABS. Winer et al. [1] in a prospective study of 438 consecutive TTTS cases treated at 15–26 weeks of gestation reported PABS in 8 cases (1.8%). All the cases described by Winer survived the perinatal period and the affected twin was always the former recipient. In our case the recipient was demised at 24 weeks of gestation and the donor presented sequels of PABS. As the procedure was reported without immediate complications, we can only speculate on a nondiagnosed intertwin membrane rupture or detachment, since during the delivery we did not identify any amniotic membrane between both fetuses. Inadvertent septostomy has been reported to occur in 7% of cases after fetoscopic laser therapy, and is associated with an increased risk of adverse perinatal outcome and PABS [17]. The diagnosis of PABS was made prenatally in only 2 of 8 cases (25%) [1], highlighting the poor sensitivity of prenatal diagnosis for this condition. In our case, ultrasonographic diagnosis of the situation could not be made prenatally as the abnormalities were subtle.

Though a rare condition, we should be aware of the possibility of PABS in pregnancies that are treated by SFLP for TTTS, especially when fetal death of one twin occurs. In our report, the surviving twin had surgical correction with excellent cosmetic and functional results presenting no major postnatal sequelae of PABS. Nonetheless, some authors suggest the use of targeted ultrasound follow-up examination with Doppler studies and three-dimensional sonography [18, 19] because this may contribute to better diagnosis, counseling, treatment and long-term outcome of cases that are complicated with PABS.

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