

CASE REPORT

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Antenatal management of twin anemia-polycythemia sequence

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Abstract

Twin-twin transfusion syndrome (TTTS) and recently discovered twin anemia-polycythemia sequence (TAPS) are well-known severe complications of monochorionic pregnancies which are associated with placental anastomoses. TAPS may occur spontaneously with a prevalence of 3% to 5% in monochorionic pregnancies. It also can occur iatrogenically after laser treatment of TTTS in 2% to 13% of monochorionic pregnancies. Optimal perinatal management for TAPS has not been established yet. It has been reported that early well-timed diagnosis of the disease is very important for reducing fetal adverse events. Because of the rarity, the literature is limited to the description of small series or case reports of twins affected by TAPS. Here, we describe diagnosis and management of TAPS case that was diagnosed after laser treatment of TTTS and review the literature about clinical characteristics including clinical features, prenatal characteristics, diagnosis, treatments and short-term outcomes of the TAPS.

Keywords: Twin anemia-polycythemia sequence, twin to twin transfusion, polyhydramnios

Introduction

The widespread use of assisted reproductive technologies along with advancing maternal age has resulted in a prominent rise in the frequency of multiple pregnancies which of 98% are twins [1]. They are associated with a higher risk of morbidity and mortality than singleton pregnancies include gestational diabetes, miscarriage, hypertensive disorders, preterm delivery, fetal genetic and congenital abnormalities, stillbirth and cerebral palsy [1,2].

Twin-twin transfusion syndrome (TTTS) and recently discovered twin anemia-polycythemia sequence (TAPS) are well-known severe complications of monochorionic pregnancies which are associated with placental anastomoses [3].

TAPS may occur spontaneously with a prevalence of 3% to 5% in monochorionic pregnancies. It also can occur iatrogenically after laser treatment of TTTS in 2% to 13% of monochorionic pregnancies. Fetoscopic laser coagulation of vascular anastomoses is currently the best available therapeutic option for TTTS. It usually presents within 1–5 weeks after the initial laser surgery [4]. Perinatal outcome in TAPS vary and appears to change according to the severity of the disease. Optimal perinatal management for

TAPS has not been established yet. It has been reported that early well-timed diagnosis of the disease is very important for reducing fetal adverse events [4,5].

Because of the rarity, the literature is limited to the description of small series or case reports of twins affected by TAPS. Here, we describe diagnosis and management of TAPS case that was diagnosed after laser treatment of TTTS and review the literature about clinical characteristics including clinical features, prenatal characteristics, diagnosis, treatments and short-term outcomes of the TAPS.

Case Report

A 33-year-old gravida 1 para 0 pregnant was referred to our clinic at 24 weeks' gestation with signs of TTTS (Donor fetus had reversed flow in the ductus venous Doppler- Quintero stage III) that confirmed using Voluson 730 Expert, GE Medical Systems, Milwaukee, Wisconsin, U.S.A. Patient was treated with fetoscopic laser coagulation of vascular anastomoses under general anesthesia (Figure 1). A 3.0-mm Karl Storz 26161 U trocar (Karl Storz GmbH & Co. KG, Tuttlingen, Germany), a 2.0-mm Karl Storz 26008AA Hopkins II Telescope with 0 degree and a 600-µm diameter diode laser fiber, with a maximum power of 40 W (Diode laser, Dornier MedTech) was used during the procedure. Trocar was inserted percutaneously into the sac of the recipient twin and the telescope with 0 degree was passed through the trocar. Five AV anastomoses from donor to recipient were first identified and coagulated,

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followed by coagulation of 3 AV from recipient to donor with a maximum power of 50 W. The time lapse between coagulation of the first and the last anastomosis was 5 minutes.

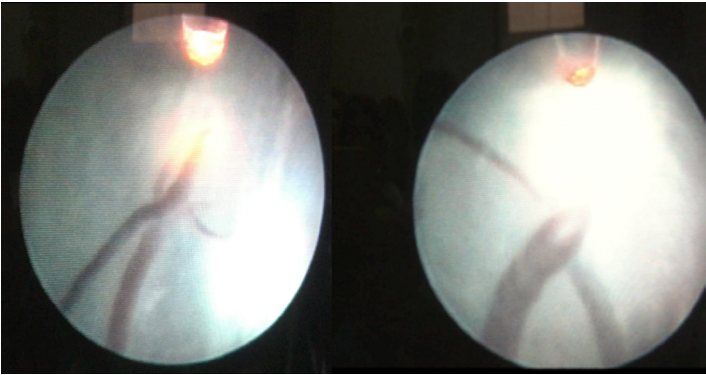


Figure 1. Fetoscopic laser coagulation of the anastomotic vessels

Four days later normalization of the amount of amniotic fluid in both sacs and bladder filling in both fetuses were confirmed. The Doppler findings were normal.

However, ten days later, Doppler measurement revealed a gradual increase of the MCA-PSV in the ex-recipient, with a maximum MCA-PSV value was 62 cm/sec (1.93 multiples of the median (Mom)), suggesting fetal anemia and the other baby’s maximum MCA-PSV value was 27 cm/sec (0.8 MoM), suggesting fetal

polycythemia (Stage 2 TAPS). The combination of both findings led to the clinical suspicion of TAPS, most probably because of the presence of residual anastomoses. The findings of the fetuses had progressed in the following days. Two weeks later, TAPS re-staged as stage 3 based on TAPS diagnostic criteria at 26th weeks of the gestation (Table 1). The various therapeutic options were discussed with the parents, including re-intervention with fetoscopic laser, intrauterine transfusion, selective feticide of the hydropic twin by radiofrequency laser ablation, and expectant management. The parents chose the expectant management. Scalp and skin edema, ascites, and pericardial effusion that suggest fetal hydrops gradually developed in the ex-recipient at 27th weeks of gestation eventually preterm premature rupture of membranes (PPROM). Later a Caesarean section was performed because of fetal heart rate anomalies associated with reduced fetal movement. There was no evidence of placental abruption at delivery. One of the twin was a plethoric with symptoms of respiratory distress requiring intubation for surfactant therapy. The echocardiography showed moderate right ventricular hypertrophy, right atrial dilatation, and a moderate pericardial effusion. Initial hemoglobin concentration was 20 mg/dl and the baby died after 2 hours delivery. Second twin was a pale with 7mg /dl hemoglobin level and died after 1 day.

Placental examination was difficult because the chorionic plate was torn extensively along the area of the prior laser procedure. The placenta macroscopic findings showed discordant placental proportions (Figure 2).

Table 1. Staging of TTTS and TAPS

Stage	TTTS (Quintero et al.) [10]		Prenatal TAPS (Slaghekke et al.) [13]		Postnatal TAPS (Slaghekke et al.) [13]
		Comment [14]		Comment [14]	Inter-twin Hb Discordance, g/dl
1	Polyhydramnios* and oligohydramnios** sequence. Bladder of donor visible.	Mild volume imbalance	Recipient MCA PSV < 1.0 MOM and donor MCA PSV > 1.5 MOM	Mild Hb discordance	>8
2	Polyhydramnios* and oligohydramnios** sequence. Bladder of donor not visible.	Progressive volume imbalance	Recipient MCA PSV < 0.8 MOM and donor MCA PSV > 1.7 MOM	Increasing Hb discordance	>11
3	Polyhydramnios and oligohydramnios sequence. Bladder of donor not visible.	Progress to cardiovascular dysfunction	Abnormal Doppler scan***	Progress to cardiovascular dysfunction	>14
4	Abnormal Doppler scan***	Overt heart failure	One or both fetuses have hydrops	Overt heart failure	>17
5	One or both fetuses have hydrops	High risk for secondary organ damage if one co*twin survives	One or both fetuses have died	High risk for secondary organ damage if one co-twin survives	>20

Hb: hemoglobin, *> 8 cm maximal pocket, **<2 cm maximal pocket, ***Absent or reversed end-diastolic velocity in the umbilical artery, pulsatile flow in the umbilical vein or reversed flow in the ductus venosus.

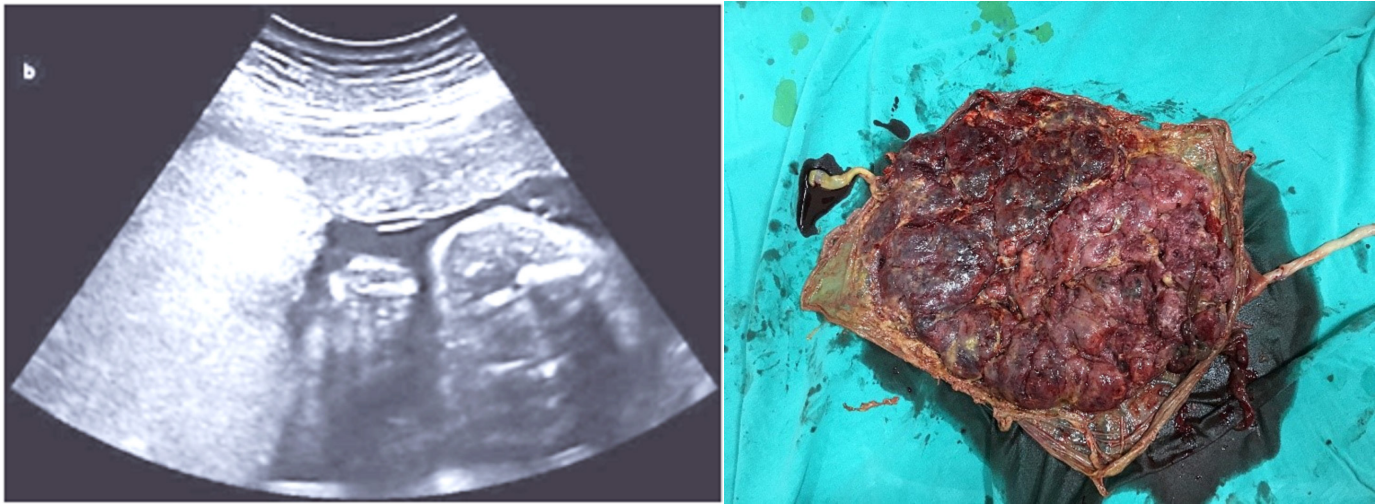


Figure 2. The placental ultrasonography and macroscopic findings showed discordant placental proportions

Discussion

Multiple pregnancies are classified as mono- or dizygotic based on number of eggs fertilized at conception. Monozygotic twins occur when one egg is fertilized by one sperm followed by division of the embryo in to two from 2 to 14 days after fertilization [1,2,7].

Monozygotic twins are associated with higher perinatal morbidity and mortality than dizygotic twins. Chorionicity is determined by the timing of embryo division in monozygotic twins [7].

Previous studies showed that while dichorionic twins almost never have placental vascular anastomoses virtually all monochorionic twins have vascular anastomoses connecting the two fetal circulations. Artery to artery (AA), vein to vein (VV), and artery to vein (AV) vascular anastomoses can be seen in monochorionic placentas [8]. AV anastomoses that are known as ‘deep anastomoses’ are unidirectional anastomoses in which an artery of one twin is connected with a vein of the co-twin via a shared cotyledon and blood flow one direction from artery to vein. AA and VV anastomoses are ‘superficial’ anastomoses since they lie on the placental surface and they are bi-directional. Almost all monochorionic twins share a single placenta with inter-twin vascular anastomoses that allows blood to transfer from one fetus to the other and vice versa [8,9]. Both TTTS and TAPS are the chronic form of fetus to fetus transfusion. The main difference between TAPS and TTTS is absence of oligohydramnios and polyhydramnios in TAPS. It is not known exactly why the recipient twin does not develop polyhydramnios and the donor twin in TAPS does not develop oligohydramnios, like TTTS.

TTTS that complicates 10-15 % of monochorionic twins, is seen usually at second and third trimester. Although almost all monochorionic twins share vascular anastomoses and thus exist in a state of constant inter-twin transfusion only a minority develop clinical TTTS. Quintero staging is widely used for staging TTTS (Table 1) [10-14].

TAPS whose placentas typically have few small and mostly unidirectional AV anastomoses was described as a new form of chronic feto-fetal transfusion first time in 2007 by Lopriore et al [11]. TAPS develop when a chronic net transfusion of red

blood cells through very small diameter (< 1 mm) unidirectional anastomoses takes place [11,15]. TAPS can occur sporadically, or iatrogenically following fetoscopic laser ablation for TTTS [8,13-15]. It has been reported that spontaneous and post laser TAPS have different placental angioarchitecture. As compared with twins with post-laser TAPS, spontaneous TAPS had a higher number of AA anastomoses [8,15].

The middle cerebral artery peak systolic velocity (MCA-PSV) is used for prenatally diagnosis of TAPS. Increased value of MCA-PSV (greater than 1.5 MoM) of the donor twin suggestive of fetal anemia and decreased MCA-PSV (less than 1.0 MoM) in the recipient twin, suggestive for fetal polycythemia (Table 1) [10-14]. In TAPS, the fetus who present with anemia is usually former recipient [8].

Diagnosis for postnatal TAPS requires that the inter-twin hemoglobin difference must be more >8 g/dl and also one of the two following criteria: either a reticulocyte count ratio of >1.7 or the presence of only small vascular anastomoses (diameter 1 mm) on placental inspection [13] (Table 1). TAPS must be differentiated from uteroplacental insufficiency, discordant manifestation of intrauterine infection, and any cause of fetal anemia [13].

The perinatal mortality and morbidity frequency in TAPS is not known yet, probably due to the heterogeneity of TAPS outcome is vary. Spontaneous resolution of antenatal TAPS has also been described [14]. Once the TAPS is diagnosed, management depends on gestational age and accessibility of the vascular equator [14]. Management options includes blood transfusion of the anemic twin (to prolong gestation) with or without exchange transfusion of the twin with polycythemia; selective feticide, re-laser treatment for residual anastomoses or conservative management [14]. It has been reported a case of post-laser TAPS which resolved spontaneously without any intervention [16].

We report a case of TAPS after fetoscopic laser surgery leading to signs of severe anemia and fetal hydrops in the ex-recipient and polycythemia in the ex-donor. These symptoms persisted for several weeks and eventually caused PPRM and a preterm delivery.

Conclusion

Although the only causal treatment for TAPS is fetoscopic laser coagulation of the residue vascular anastomoses, it can be technically more challenging than in TTTS. It is mainly due to absence of polyhydramnios and a stuck twin, which makes the visualization of the vascular equator more difficult. The other reason is placental anastomoses in TAPS are known to be only few and may be missed during fetoscopy because of their smallness. Prevention of post-laser TAPS can be achieved by reducing the rate of residual anastomoses by using 'Solomon' technique that coagulate entire vascular equator on the chorionic surface instead of selectively coagulating each visible anastomosis [13,15]. Ideally randomized trials are needed to determine the optimal management option for TAPS.

Competing interests

The authors declare that they have no competing interest

Financial Disclosure

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