CASE REPORT

Klippel–Trénaunay–Weber syndrome associated with fetal growth restriction


Department of Obstetrics and Gynecology 'A', Sertin Maternity Hospital, Tel Aviv Sourasky Medical Center and Sackler Faculty of Medicine, Tel Aviv University, Israel

1To whom correspondence should be addressed at: Department of Obstetrics and Gynecology 'A', Sertin Maternity Hospital, Tel Aviv Sourasky Medical Center, PO Box 7079, Tel Aviv 61070, Israel

Klippel–Trénaunay–Weber syndrome is a rare congenital deep-vein malformation. Pregnancy in patients with this syndrome is rare and only a few cases have been reported. Known obstetrical risks in pregnant patients with this syndrome include bleeding from angiomata in the genitalia, and coagulation disturbances. We present a 31 year old woman with this syndrome who, on two occasions, delivered small-for-gestational-age neonates. This may have been due to placental insufficiency caused by angiomatosis related to the syndrome.

Key words: fetal growth restriction/Klippel–Trénaunay–Weber syndrome

Introduction

Klippel–Trénaunay–Weber syndrome (KTWS) is a rare, congenital malformation, characterized by varicosity, hypertrophy of soft tissues and bones, haemangioma (Klippel and Trénaunay, 1900; Baskervill et al., 1985a), deep-vein anomaly (Lindenauer, 1965), and arteriovenous fistulae (Parks-Weber, 1918), in various organs including the pelvic organs and vagina (Powolony, 1986). The cause of the syndrome is unknown, but most likely is the result of a diffuse mesodermal abnormality (Baskervill et al., 1985b). Recently, the syndrome was associated with a 5:11 balanced translocation (Whelan et al., 1995).

Prenatal diagnosis by ultrasound was first reported by Hatjis et al. (1981) and since then additional cases have been reported. Pregnancy in patients with KTWS is rare and only a few pregnancies have been reported (Schmutzler, 1968; Kopp and Rielbrock, 1969; Powolony, 1986; Fishman and Paldi, 1989; Verheijen et al., 1989; Neubert et al., 1995; Pollack et al., 1995).

We present a patient with KTWS who, on two occasions, delivered small-for-gestational-age neonates, which may be causally related to the pathological manifestations of the syndrome in the placenta.

Case report

The patient was a 31 year old white woman with KTWS. Since childhood she had suffered from haemangioma, varicosity and soft-tissue hypertrophy of the left lower extremity for which she had been treated conservatively. At the age of 25 years she conceived for the first time. Pregnancy was uneventful until 35 weeks gestation when a significant enlargement of the affected limb was noted. Although deep-vein thrombosis was not demonstrated on Doppler ultrasound, prophylactic subcutaneous heparin was initiated. Despite this treatment, the patient's clinical condition did not improve.

During admission, ultrasonographic examination revealed (for the first time) fetal growth restriction, and a work-up for growth restriction that included screening for chronic maternal disease, serology for infectious and autoimmune diseases, and targeted ultrasound for fetal malformations, was normal. Induction of labour was attempted at 39 weeks gestation but due to fetal distress (detected on fetal heart monitoring), Caesarean delivery was performed. A healthy male neonate weighing 2380 g was delivered, with an Apgar score of 10 at 1 and 5 min.

Postpartum, the patient was evaluated by a vascular surgeon and, based on the clinical findings and absence of deep vein thrombosis, KTWS was diagnosed. The patient was operated on and the superficial femoral vein, as well as some of the varicosities were removed, resulting in slight improvement of the left limb hypertrophy.

Six years later the patient conceived again. Aspirin, 100 mg/day, was started from 12 weeks gestation because of the previous delivery of a small-for-gestational-age newborn. The pregnancy course, including a monthly ultrasonographic examination, was uneventful until 35 weeks gestation, when fetal growth arrest was detected. Again work-up for fetal growth restriction was negative.

A non-stress test and a biophysical profile were performed on a weekly basis until the 38th gestational week when, due to fetal growth restriction, uterine scar and unripened cervix, the delivery was accomplished by elective Caesarean section. A healthy male neonate weighing 2100 g was delivered, with an Apgar score of 10 at 1 and 5 min. Preoperative, low-molecular-weight heparin (20 mg/day s.c.) was administered prophylactically, and continued for the first postpartum week.

The operation and postoperative course were uneventful. Pathological examination revealed a 540 g placenta with a fresh placental infarct, surrounded by an excess of syncytial knots and areas of chorangiosis (Figure 1), with villous vascular congestion.

© European Society for Human Reproduction and Embryology
Pregnancy in patients with KTWS is rare. Maternal risks during both vaginal and Caesarean delivery are excessive bleeding and coagulation disturbances due to stasis of blood in the angiomas (Baskervill et al., 1985a).

Indeed, in three of the reported cases (Schmutzler, 1968; Powolony, 1986; Neubert et al., 1995), the patients underwent elective Caesarean section because of angiomas at the external genitalia. Two of these patients had coagulation disturbances after surgery (Schmutzler, 1968; Neubert et al., 1995), resulting in the death of one patient due to disseminated intravascular coagulation and acute renal failure, probably because of stasis and bleeding from the angiomatosis areas in the pelvis. A fourth patient underwent Caesarean delivery but no indication was reported (Verheijen et al., 1989). Three patients were reported to have delivered vaginally without complications (Kopp and Rietbrock, 1969; Fishman and Paldi, 1989; Pollack et al., 1995), one of whom delivered twins (Kopp and Rietbrock, 1969).

In our patient an initial attempt was made at vaginal delivery. However, due to obstetrical indications, uncomplicated Caesarean sections at both deliveries were performed.

Fetal risks in patients with this syndrome are unknown. Five of the seven fetuses reported in the literature, including the twin neonates, were appropriate for gestational age (Kopp and Rietbrock, 1969; Powolony, 1986; Fishman and Paldi, 1989; Neubert et al., 1995; Pollack et al., 1995). In one report the fetus was defined as slightly growth restricted (Verheijen et al., 1989). The birth weight of one fetus was not reported (Schmutzler, 1968). Our patient delivered small-for-gestational-age neonates twice. As extensive work-up performed for fetal growth restriction was negative, it may be speculated that angiomatosis of the placenta, detected in the pathological examination, may result in shunting of blood from the placenta and possibly an infarct due to stasis, which may therefore contribute to placental insufficiency and fetal growth restriction.

Mode of delivery should be individualized in patients with this syndrome. When arteriovenous anomalies and angiomas of the external genitalia are noted, Caesarean delivery is probably safer due to possible uncontrollable bleeding. However, during Caesarean delivery caution should be exercised to avoid possible bleeding from angiomas in the uterine region. Patients without external vascular abnormalities may be candidates for vaginal delivery. Regardless of the mode of delivery, large amounts of blood should be available and intensive haemodynamic monitoring is indicated. Peripartum prophylactic anticoagulation with low-molecular-weight heparin should be considered.

To the best of our knowledge, the aforementioned case presents, for the first time, the possible association between KTWS and fetal growth restriction. Therefore, the fetus should be closely monitored for growth restriction by frequent ultrasound, and possibly Doppler flow, examinations.

References

Received on May 20, 1996, accepted on September 17, 1996.