Clinical Case Report

Pregnancy with Klippel–Trenaunay syndrome

MONICA GUPTA, GARIMA KACHHAWA, RAJESH KUMARI, ALKA KRIPLANI

ABSTRACT

Klippel–Trenaunay syndrome (KTS) is a rare congenital disorder characterized by the presence of vascular naevi, varicose veins and soft tissue or bone hypertrophy affecting one or more extremities. Due to the rarity of the syndrome, there is limited literature regarding the current practice in the management of pregnancy complicated with KTS. Successful management of pregnancy with KTS is a challenge for clinicians as pregnancy exacerbates the already increased risk of thrombosis and haemorrhage associated with this syndrome. We report a patient with KTS with previous poor obstetric history managed with favourable maternal and foetal outcomes.

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INTRODUCTION

Klippel–Trenaunay syndrome (KTS) is a congenital disorder characterized by cutaneous haemangioma, venous varicosities and bone or soft tissue hypertrophy affecting one or more extremities.[1] It is a rare birth defect of unknown origin and variable phenotypic expression, with a reported prevalence of 1 in 30 000–100 000 live births.[2] The vascular malformation in KTS is generally seen in extremities, but can cover large areas of the body and even extend up to the pelvis involving the internal and external genitalia. Pregnant patients with KTS can have aggravation of symptoms of KTS or present with life-threatening complications such as recurrent thromboembolism and severe postpartum haemorrhage.[3–5] There are a few case reports/case series in the literature describing severe complications in pregnancy such as uterine prolapse and paraplegia.[6,7] This may be due to rarity of the syndrome itself and physicians often advising patients with KTS against conception because of exacerbations of complications during pregnancy.[3,8] Information regarding obstetric care in these patients is hence scarce. We report a pregnant woman with KTS with previous poor obstetric outcome, who was managed successfully at a tertiary care hospital.

THE CASE

A 32-year-old female (gravida 2, parity 1, live 0) was referred to us at 13 weeks of gestation with previous poor obstetric history and known case of KTS. Her previous pregnancy, 2 years ago, was complicated with intrauterine growth restriction, severe oligohydramnios and absent diastolic flow on ultrasound for which she underwent an emergency caesarean section at 34 weeks' gestation. The baby died on day 11 of life due to sepsis. In the postpartum period, she was evaluated for leg swelling and varicosities. She had swelling of her right lower limb since childhood, but there was no history of thrombosis. Computed tomography peripheral angiography revealed generalized hypertrophy of the right lower limb, soft tissue and muscle; cortical thickening of the right lower limb long bones that was more obvious in the femur; extensive dilatation of the superficial veins of the right lower limb and a normal deep venous system. A diagnosis of KTS was made on the basis of these findings and the presence of venous varicosities in the right leg.

During the present pregnancy, her first trimester was uneventful with normal antenatal investigations, including haemoglobin of 12.5 g/dl and platelet count of 172×10^9/L. Her coagulation profile and thrombophilia screening tests were normal. She developed varicosities and increased swelling in her right leg since the fourth month of pregnancy (Fig. 1). The lower limb swelling increased progressively and became painful with advancing gestation. The swelling exacerbated after walking and reduced on lying, and the right leg measured 4 cm more in diameter than the left at the level of the mid-thigh. She was advised the use of elastic compression stockings and elevation of the foot end of the bed while sleeping to prevent thrombosis. After consultation with a vascular surgeon, she was given low molecular weight heparin (LMWH) 1 mg/kg subcutaneous twice daily and oral micronized purified flavonoid extract 500 mg twice daily from 28 weeks of gestation. An obstetric ultrasound at 28 weeks revealed a grade 2 placenta and prominent veins in the body of the uterine myometrium (Fig. 2). There was no limb hypertrophy, cystic lesions or growth restriction in the foetus. Foetal growth was monitored every fortnight thereafter. Magnetic resonance imaging (MRI) of the spine revealed no arteriovenous malformation. Doppler ultrasound of the lower limb showed no venous thrombosis.

At 37 weeks' gestation, pelvic examination revealed a favourable Bishop score and increased probability of success

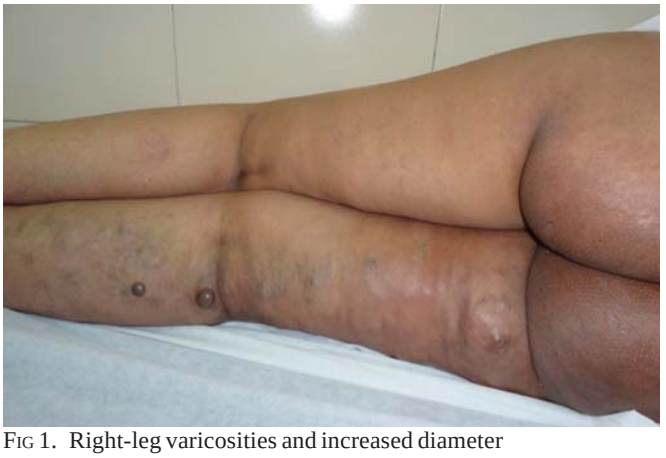

FIG 1. Right-leg varicosities and increased diameter

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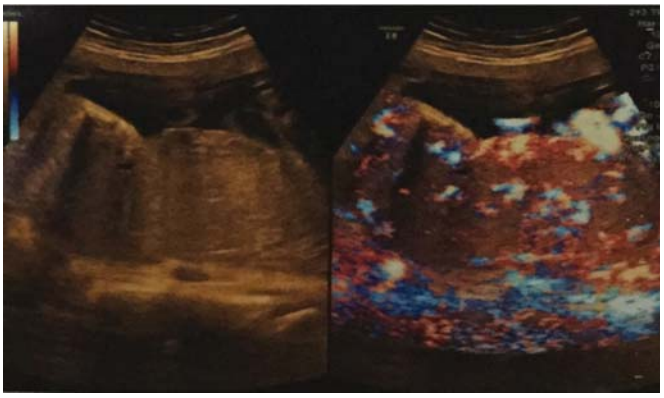

FIG 2. Prominent veins in uterine myometrium

of trial of labour after caesarean (TOLAC). Although there were no vulval varicosities, vaginal mucosal vascularity was increased. The patient was counselled regarding the risks and benefits of caesarean versus vaginal delivery, and she consented for TOLAC. The patient went into spontaneous labour at 38 weeks and delivered vaginally a male child weighing 3.09 kg. There was excessive bleeding from the episiotomy site from dilated and engorged vessels, which were ligated, and the episiotomy wound was stitched in layers. Vaginal packing was done with antiseptic-soaked roller gauze for 24 hours to control diffuse oozing from the vaginal mucosa. The blood loss was approximately 800 ml. The patient received two units of packed red blood cells and injectable antibiotics for 48 hours. She was advised elastic stockings and thrombo-prophylaxis with LMWH after delivery until 6 weeks postpartum. Her postpartum period was uneventful.

DISCUSSION

KTS also known as 'Naevus vasculosus osteohypertrophicus', was first described by two French physicians, Maurice Klippel and Paul Trenaunay, in 1900.[2] The disease is characterized by the triad of vascular malformation of capillaries, veins or lymphatic (port-wine stains) and/or venous varicosities along with limb hypertrophy due to overgrowth of bone or soft tissue.[2] At least two of the three features should be present for diagnosis. The predominant sites of the characteristic port wine stains are the legs followed by arms, trunk and rarely on head or neck. Our patient had swelling in the leg since childhood while varicosities developed in pregnancy, but port wine stains were absent, and hence diagnosis was delayed. There was increase in varicosities and hypertrophy of soft tissue and bone during pregnancy, leading to differences in leg dimension.[1]

KTS is mostly sporadic, and rarely an autosomal dominant pattern of inheritance has been reported in some cases.[9] Since the disease can be hereditary in rare cases, it is important to exclude limb hypertrophy or multiloculated cystic lesion in thorax, abdominal wall or the limbs of foetus by colour Doppler ultrasound.[10] There is a risk of intrauterine growth restriction and hence growth of the foetus should be monitored during pregnancy.[11] Our patient had growth-restricted foetus in her first pregnancy, but the present pregnancy was uneventful and congenital anomaly scan was also normal.

Coagulopathy is the most commonly reported serious complication of this syndrome. Postoperative venous thrombosis is ten times more common in patients with KTS, and it tends to be recurrent.[5] Both antiplatelets (aspirin) and anticoagulants (LMWH) can be considered in these patients. In patients with a history of thrombosis, therapeutic anticoagulation is recommended.[3] Prophylactic anticoagulation is generally recommended during the postpartum period. In one of the reported cases, only postpartum anticoagulation was given, and the patient developed superficial venous thrombosis of the leg.[12] In another report, the patient with KTS developed postpartum pulmonary thromboembolism despite both antepartum and postpartum thrombo-prophylaxis.[13] In our patient, LMWH was started at 28 weeks due to progressive increase in swelling and varicosities of the right leg, which were also painful. Other measures for the prevention of thrombosis are micronized purified flavonoid extract and elastic compression stockings. Micronized purified flavonoid extract along with LMWH combination has been found to be superior to LMWH alone in the prevention of thromboembolism.[14] Our patient did not have thrombosis despite the grossly increased swelling and varicosities. Horbach *et al*.[4] have estimated a risk of 83 venous thromboembolic events per 1000 KTS pregnancies from their nationwide cross-sectional data. This estimate is more than the cut-off for prophylactic anticoagulation (70/1000 pregnancies)[15] as suggested by the recent guidelines and hence, they recommend prophylactic anticoagulation during pregnancy and 6 weeks postpartum.

Normal physiological changes of pregnancy such as increased cardiac output, venous pressure, limb oedema and venous stasis can exacerbate the already increased risk of thromboembolism and haemorrhage in KTS.[3] The morbidity in KTS is related to complications due to venous malformations resulting in cellulitis, venous ulcers, thrombophlebitis and thromboembolism. There may be extension of these malformations to intra-abdominal and pelvic organs and to the skin of the lower abdomen, pelvis and genitalia, leading to haemorrhage at these sites, hence complicating the mode of delivery.[5] The mode of delivery in patients with KTS should be individualized. Both modes have their own risk of complications. Due to vulvovaginal varicosities, there is an increased risk of bleeding into the rectum and haemorrhage during delivery. MRI spine should be done in these patients to rule out neuraxial vascular anomalies, which could complicate locoregional anaesthesia. Moreover, caesarean section can be complicated by abdominal wall or uterine varices and haemangiomata as in our patient and hence should be avoided except for obstetric indications.[16,17]

Conclusion

KTS in pregnancy can be associated with life-threatening complications. Obstetricians should counsel patients with KTS regarding these risks in the pre-conception period. Thrombo-prophylaxis should be considered for patients during pregnancy and in the postpartum period. A successful outcome of pregnancy complicated with KTS is possible with a multidisciplinary approach involving the gynaecologist, anaesthesiologist and haematologist or vascular surgeon.

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Conflicts of interest. None declared

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