

Klippel Trenaunay syndrome with pregnancy – A rare presentation

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Abstract

Introduction: Klippel Trenaunay syndrome (KTS) is a rare congenital disorder, the etiology of which is unknown and it is characterised by vascular malformations. The incidence of pregnancy with Klippel Trenaunay syndrome is extremely rare with very few of them being reported and documented. To achieve successful maternal and fetal outcome a multidisciplinary team approach is very important.⁽¹⁾

Keywords: Klippel Trenaunay Syndrome, KTS, Congenital disorder, Vascular malformations, Pregnancy.

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Clinical Case

A 26 years old diagnosed case of KTS, referred to our hospital as a case of G₂A₁ with 40⁺² weeks of gestation in early labour with failed induction. She was a booked case at a private nursing home where her antenatal period was uneventful. She had no past history of thrombosis but did undergo venous malformation excision surgery about 15 years back.

On general physical examination, vitals were stable, she had gross swelling of the left upper limb extending to the left chest and left scapular region with venous varices, multiple nodules and papules, some discrete and some grouped, bluish black to deep red over port wine stain. Per abdomen examination revealed uterus full term size, relaxed, cephalic presentation with fetal heart sounds well heard. Internal examination revealed an unfavourable Bishop score with average pelvic adequacy. Relevant investigations were within normal limits, coagulation profile was also normal. Uterine Doppler study did not reveal the presence of uterine venous malformation. A multidisciplinary team involving obstetricians, neonatologist, vascular surgeon, sonologists and anesthetists were involved during the management of the patient. She underwent an elective LSCS on the following day of admission in view of past dates and failed induction, a healthy live male baby of 3.28 kgs was delivered with no external congenital anomalies. Intraoperatively there were no uterine vascular malformations. Post-op period was uneventful, she was managed with limb elevation, compression stockings and heparin prophylaxis. Currently mother and child are doing well.



Discussion

Klippel Trenaunay syndrome was first described in 1900 by French physicians Maurice Klippel and Paul Trenaunay who recognised the vascular anomalies which were causing defects in the skin, limbs and other organs and referred it as 'naevus vasculoosus osteodystrophicus'.⁽²⁾ Frederick Parkes Weber, a German-British physician described arteriovenous fistula formation in the hypertrophic limbs in cases which were similar but were not identical to those described by Klippel and Trenaunay and those cases were referred as Klippel-Trenaunay-Weber syndrome,⁽³⁾ a rare congenital vascular disorder.

Klippel Trenaunay Syndrome (KTS) is a rare congenital disorder presenting with a clinical triad consisting of vascular malformations or deformities with extensive port wine stains, hypertrophy of soft tissues or bones involving limbs sometimes affecting pelvic and abdominal organs.

KTS in pregnancy is an extremely rare presentation and till date there are reports of only about twenty cases. The etiology of KTS is very much unknown and there are many theories associated as causing factor. Probability of mesodermal abnormality during the

intrauterine fetal development or abnormalities of the sympathetic system causing arteriovenous anastomotic dilatations,⁽⁴⁾ involvement of angiogenic factor AGGF1^(5,6,7) has also been associated. In 1% cases of KTS a single gene defect associated with translocation at(8;14)(q22.3;q13) has been proposed. Rarely it is inherited as autosomal dominant trait.^(8,9)

In the peri-conceptual period, imaging studies like ultrasonography, Doppler studies and contrast enhanced magnetic resonance imaging will help confirm the diagnosis of KTS and the extent of the lesion which will help in planning for management and intervention if required. Treatment for KTS is not definitive and mainly involves symptomatic management like excision surgery for venous varicosities to reduce risks and complications. Vascular anomalies in KTS causes morbidities like venous insufficiency, cellulitis, thrombophlebitis, thromboembolic disease and limb disparity.⁽³⁾ Active intervention is indicated in cases of localized lesions and in the presence of serious complications like bleeding or cardiac failure. Treatment options available for treating KTS are surgery, sclerotherapy, radiotherapy and compression therapy.⁽¹⁰⁾ Newer treatment methods are endovenous thermal ablation and laser treatment. Cellulitis and thrombophlebitis can be managed with analgesics, antibiotics, corticosteroids and limb elevation. Vertebral scoliosis and gait abnormalities are caused due to hypertrophy of a limb. KTS patients with stable disease need monitoring atleast annually or more often if clinically indicated. In case of progression of disease imaging studies should be performed and appropriate medical or surgical intervention is required.

KTS with pregnancy exacerbates the normal physiological changes occurring during pregnancy like venous stasis, increased venous pressure, leg edema and cardiac output and increases the risk of adverse events during pregnancy.⁽¹¹⁾ Pregnancy exacerbates KTS complications and obstetric risks are increased by ten fold in these women compared to normal pregnant females like deep venous thrombosis and other thromboembolic events.

Uterine venous malformations complicating caesarean section can be detected by Colour Doppler flow studies. Presence of limb hypertrophy and multiloculated cystic lesions in the fetus can be diagnosed by prenatal colour Doppler flow.⁽¹²⁾ Intrauterine growth restriction has been related with this syndrome sometimes.⁽¹³⁾

Vaginal mode of delivery is contraindicated in the presence of venous malformations in the uterine cervix or vaginal wall. Though epidural anesthesia is considered as best option during labour, studies of hemostasis and magnetic resonance imaging prior to delivery to detect vascular malformations next to the spinal cord will prevent increased risk of epidural haematoma.^(14,15) Uterine venous malformations is associated with increased risk of torrential intra and

postpartum haemorrhage.⁽¹⁶⁾ Earlier hysterectomy was considered as the definitive treatment for uncontrollable bleeding in the presence of uterine venous malformations, presently alternate methods like uterine artery embolization or internal artery ligation can be employed. Earlier pregnancy was contraindicated in KTS^(17,18,19) but successful pregnancies without resorting to hysterectomy can be achieved by managing these high risk cases with multidisciplinary team management at tertiary care centres.^(20,21)

Pregnant women with this syndrome need appropriate counselling and continued support regarding the course and management in pregnancy, labour and puerperium to achieve successful maternal and fetal outcome.

It was challenging for us to handle such a rare condition with pregnancy, fortunately it was uneventful and safe for the patient and the treating team.

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