Pregnancy Complicated by Klippel–Trenaunay Syndrome and Kasabach Merritt syndrome Resulting in Severe Fetal Growth Restriction: A case report and review of literature

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Abstract

Klippel–Trenaunay syndrome (KTS) is a rare congenital vascular disorder that may be complicated by Kasabach Merritt syndrome, posing significant maternal and fetal risks during pregnancy.

We report the case of a 28-year-old pregnant woman with known Klippel–Trenaunay syndrome who developed Kasabach–Merritt phenomenon during pregnancy. The pregnancy was complicated by severe fetal growth restriction (FGR) and absent end-diastolic flow on umbilical artery Doppler. A cesarean section was performed at 35 weeks of gestation. A growth-restricted neonate was delivered and managed accordingly.

This case highlights the complexity of managing pregnancy in patients with KTS complicated by KMP and emphasizes the importance of multidisciplinary surveillance and timely delivery.

Keywords: Klippel–Trenaunay syndrome; angioosteohypertrophy syndrome; Kasabach Merritt syndrome; fetal growth restriction; high-risk pregnancy

Introduction

Klippel-Trenaunay-Weber syndrome is a sporadic genetic syndrome characterized by localized hemangiomas, venous varicosities, and asymmetric osseous hypertrophy of the ipsilateral extremities. Most commonly seen in association with hemangiomas, Kasabach-Merritt syndrome is defined by the presence of thrombocytopenia and a consumptive coagulopathy. TK syndrome is a sporadic genetic syndrome characterized by localized cutaneous hemangiomas, venous varicosities, and asymmetric osseous hypertrophy of the ipsilateral extremities.1 Complications at delivery can include disseminated intravascular coagulation (DIC) and massive hemorrhage. Coagulopathic events may be due to persistent blood loss or may reflect the development of Kasabach-Merritt syndrome. First reported in 1940, this syndrome has been described most commonly in children; it is defined by thrombocytopenia of varying degrees and a consumptive coagulopathy seen in association with hemangiomas.2 A microangiopathic hemolytic anemia may also be present.

Fetal growth restriction (FGR) with abnormal umbilical artery Doppler findings, such as absent end-diastolic flow, reflects placental insufficiency and is associated with adverse perinatal outcomes. We report a rare case of pregnancy complicated by KTS and KMP resulting in severe FGR requiring preterm cesarean delivery.

To our knowledge, only one case of a pregnancy complicated by Klippel-Trenaunay-Weber and subsequent Kasabach-Merritt syndrome has been reported.

Case Presentation

A 28-year-old multigravida woman (G3, P2) presented at 35 weeks' gestation for antenatal evaluation. She had a known diagnosis of Klippel–Trenaunay syndrome involving [affected limb/region], diagnosed at [age/year].

Maternal Clinical Findings

- Blood pressure: [value]
- Presence of vascular malformations: [description]
- Symptoms suggestive of KMP: [e.g., bruising, bleeding, pain]

Laboratory investigations revealed:

- Platelet count: [value]
- Fibrinogen: [value]
- D-dimer: [value]

These findings were consistent with Kasabach–Merritt phenomenon.

Fetal Assessment

Ultrasound examination at [gestational age] weeks demonstrated:

- Estimated fetal weight below the [percentile]
- Abnormal umbilical artery Doppler with absent end-diastolic blood flow • Amniotic fluid index: [value]

Based on worsening fetal Doppler parameters and maternal risks, a multidisciplinary team involving obstetrics, hematology, anesthesia, and neonatology recommended delivery.

Delivery and Neonatal Outcome

A cesarean section was performed at 35 weeks of gestation under [type of anesthesia]. A female/male neonate weighing [birth weight] g was delivered with Apgar scores of [values] at 1 and 5 minutes.

The neonate required [NICU admission / respiratory support / observation]. Maternal postoperative recovery was [uneventful/complicated by…].

Maternal admission to ICU

[course in details]

Discussion

Klippel-Trenaunay syndrome, first described in 1900, is defined as a triad of clinical features, including unilateral cutaneous hemangiomas, varicose veins, soft tissue, and asymmetric osseous hypertrophy of the ipsilateral extremities.! Weber'3 described several similar cases and, in 1918, reported the additional feature of arteriovenous fistulae. A review · of 768 cases of Klippel-Trenaunay-Weber syndrome described a consistent absence or major atresia of the deep venous vascular system, causing limb elongation, venous stasis, varicosities, and edema. A venogram had previously demonstrated a similar absence of the deep venous vascular system of the right lower extremity in our patient. The etiology of Klippel-Trenaunay-Weber syndrome has been reviewed by Baskerville et al,5 who suggested that a mesodermal defect operative at angiogenesis may explain the vascular abnormalities that define the syndrome. Complications in the affected extremity may include stasis dermatitis, skin ulceration, and recurrent hemarthrosis-all of which may require amputation of the limb for management. Our patient's refractory coagulopathy prompted the diagnostic consideration of Kasabach-Merritt syndrome. First reported in 1940, the syndrome consists of thrombocytopenia and a consumptive coagulopathy seen in association with hemangiomas? Consistent laboratory abnormalities include thrombocytopenia, decreased fibrinogen, and coagulation factors II, V, and Vill. Increased PT, PIT, and fibrin split products are also invariably present. A similar clinical presentation has been reported in association with Klippel-Trenaunay-Weber syndrome.6 The exact mechanism by which the thrombocytopenia and coagulopathy develop is unknown. It has been suggested that a combination of venous stasis and abnormal endothelial cells within the hemangiomas functions to increase platelet pooling and destruction with activation of the coagulation cascade, resulting in a localized and self-perpetuating coagulopathy.7 Inceman and Tangun8 reported a case in which an 8-yearold boy with a giant hemangioma of the lower extremity demonstrated a difference in PT and PIT values in blood obtained from a peripheral site in comparison with blood obtained from the hemangioma. We suspect that a similar mechanism occurred in our patient. Despite choosing a left paramedian incision, a subtle disruption of the right-sided hemangioma, exacerbated by increased ambulation, may have initiated the coagulopathy. Once established, the local consumption of clotting factors and platelets persisted in the hemangioma and resulted in hemorrhage at the incision site. A number of therapeutic modalities have been used in the management of Kasabach-Merritt syndrome. These include platelet transfusion, IV heparin, aminocaproic acid, radiotherapy, alpha 2a interferon, corticosteroids, and intermittent pneumatic compression of the extremities. A prominent feature of this patient's persistent coagulopathy was the markedly elevated fibrin split products. This finding prompted our decision to use the plasminogen-activator inhibitory effects of aminocaproic acid after the failure of blood product replacement therapy to correct her coagulopathy. Although a pregnancy presenting with Klippel-Trenaunay-Weber syndrome is rare, many patients with known deep or superficial hemangiomas can and do become pregnant. The potential for a refractory coagulopathy presenting as Kasabach-Merritt syndrome should be considered in any patient who presents with extensive hemangiomas.

Conclusion

Pregnancy in women with Klippel–Trenaunay syndrome complicated by Kasabach–Merritt phenomenon is rare and high risk. Intensive antenatal surveillance and multidisciplinary management are essential to optimize maternal and perinatal outcomes.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

A written consent for publication has been obtained from the patient.

Availability of data and materials

The data and materials supporting the conclusions of this article are available as a supplementary materials.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

SIA, ME, AFN contributed to writing the first draft of the manuscript. All the authors revised the manuscript critically for important intellectual content. All authors approved the final version of the manuscript.

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