

CASE REPORT ON MANAGEMENT OF ANTENATAL PATIENT WITH KLIPPEL TRENAUNARY SYNDROME

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Objectives: To discuss the management of antenatal patient with vascular malformation of KTS and MDT input.

Design:
Case based Literature review

Case report:
G4 P3 who is a known case of KTS with a triad of Vascular malformation as Port wines, long left leg and Varicose veins was referred to high risk obstetrics clinic. Her pregnancy was managed with MDT input with a haematologist, anaesthetist, surgical team, Obstetrician and a paediatrician. Patient has a fetal growth surveillance at 28, 32 and 36 weeks and was on antenatal clexane due to her increased risk of thrombosis. She had IOL at 38+ weeks and she developed APH during labour with a blood loss of 250 mls with normal observations and soft abdomen. She had initial resuscitation with IV fluids, baseline bloods, ROTEM and 4 units of RBC cross matched. She progressed to normal delivery and needed syntocinon infusion for PPH and transexemic acid .



Discussion:

Klippel Trenaunay syndrome is a rare Cutaneous vascular malformation syndrome involving a triad of capillary & venous malformation and limb hypertrophy.

Most cases of KTS are caused by somatic mutations in the PIK3CA gene. A multi-disciplinary team approach with optimal care coordination and mainstay is symptomatic care with medical and occasional surgical intervention. During childhood, close monitoring for limb length discrepancy is needed. Patients may need Compression stockings, limb elevation, and intermittent pneumatic compression devices are used to minimize lymphoedema and venous insufficiency.

Sclerotherapy for vascular malformations and Laser treatment can be offered for port-wine stains.

Pregnant patients need anti-thrombotic therapy as prophylaxis due to a high risk of deep venous thrombosis and pulmonary embolism. Bleeding due to coagulopathy should be managed promptly with supportive care, fresh frozen plasma, and low molecular weight heparin.

Conclusion:

KTS has a wide spectrum of clinical findings. Its prognosis is related to the severity of vascular malformations which progress over time with ongoing overgrowth and worsening venous insufficiency. Acute bleeding from GI malformation carries the increased mortality risk. Recurrent deep venous thrombosis leads to the risk of PE which can be massive and fatal. Pregnancy can be associated with increased risk of thrombotic events as well as bleeding in the form of Antepartum and postpartum bleeding in these group of patients and outcomes can be good with multidisciplinary input.