

Anaesthetic Management of a Parturient with Klippel-Trenaunay Syndrome

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Introduction

Klippel-Trenaunay Syndrome is a rare congenital vascular disease which is characterised by capillary, venous and lymphatic malformations, leading to cutaneous haemangiomas, venous varicosities and limb hypertrophies, and is associated with both haemorrhagic and thrombotic complications. We describe the anaesthetic management of a 36 year old parturient with COVID and Klippel Trenaunay Syndrome, who underwent an elective caesarean section.

Very few such cases have been described in the past, and the multi-system nature of condition poses various challenges to both the obstetrician and the anaesthetist.





Discussion

KTS may be associated with underlying epidural and subdural vascular malformations, however, due to its rarity and inconsistent presentation, there may be some debate surrounding the defining characteristics of KTS. Gestation, and the physiological changes that come with it, may further complicate and exacerbate the manifestations of this syndrome, with increased obstetric risk, and should be avoided in these patients.
Counselling patients regarding obstetric risk is challenging and should be addressed on an individual basis. Whilst treatment of Klippel-Trenaunay syndrome is usually conservative unless specific complications occur, regular clinical and radiographic monitoring is crucial in order to assess the impact of pregnancy on disease progression.
The success in the management of patients with this syndrome requires the participation of a multidisciplinary team, consisting of obstetrician, anaesthetist, urologist, haematologist and vascular surgeon, with appropriate collaboration among the professionals involved.
Periodic imaging and coagulation tests are recommended to evaluate the evolution of vascular malformations in the pelvis, uterus, and vagina and to identify possible neuraxial changes, to guide the safest way of delivery and anaesthetic technique for these patients.

Case Report

We had a 36 year old,three previous normal vaginal deliveries,and an elective Caesarean section (CS) four years ago under general anaesthetic (GA) at 36 weeks gestation.She was told by a Vascular surgeon that she was not suitable for Regional Anaesthesia.There was no recent imaging of her back to rule out arteriovenous malformations.Her past history included Gastric Bypass surgery under GA two years ago.She also had depression,varicose veins and three previous deep venous thrombosis (DVTs) and was on prophylactic low molecular weight heparin (LMWH).She had tested positive for COVID 4 days ago,and had mild symptoms of cough and sore throat.After a multi-disciplinary discussion involving Obstetrician,Vascular Surgeon,Haematologist and Anaesthetist,a decision was made to proceed with GA for the elective CS in agreement with the patient.After securing two wide bore cannulae and adequate pre-oxygenation,a modified Rapid Sequence Induction was performed,and an oral endotracheal tube was secured.Anaesthesia was maintained with oxygen,nitrous oxide and sevoflurane.Post delivery of baby,Oxytocin five units,followed by an infusion,Midazolam,Morphine,Ondansetron and Dexamethasone were administered.Extubation was performed when the patient was fully awake.In recovery,further opioids were given for pain.The estimated blood loss was 200 ml.

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