A PREGNANCY AFFECTED BY Klippel-Trénaunay Syndrome

Dr Zahraa Ahadzada (B MedSci (Adv), MBBS), **Dr Sumeera Samarakkody** (MBBS, FRANZCOG)
Obstetrics and Gynaecology Department, Hervey Bay Hospital

Introduction

Klippel-Trénaunay syndrome (KTS) is a rare congenital vascular malformation syndrome involving capillary, venous and lymphatic systems (1). Morbidity from the syndrome results secondary to vascular abnormalities, namely venous insufficiency, thromboembolism or haemorrhage (1,2). Pregnancies complicated by KTS are rare. The physiological changes of pregnancy, such as increased venous return, venous stasis and increased cardiac output, can further exacerbate the physiology of these women (1). Studies have shown that pregnancies complicated by KTS have a significant risk of venous thromboembolic events (VTE) as well as severe postpartum haemorrhage (1-3).

Objective

To present a case of pregnancy with KTS and the management strategies undertaken.

Case

A 20-year-old G1P0 presented for a late booking visit at 38+3 weeks and was found to have KTS. Varicose veins were her mainstay symptom of KTS, managed with graduated compression stockings during the rest of her pregnancy. She opted for a caesarean section as her preferred mode of birth. The caesarean section was carried out in an elective setting at 39+6 weeks with spinal anaesthesia and an estimated blood loss of 600ml. During the procedure, she received carbetocin and tranexamic acid with no complications. The baby weighed 3580g. Given the increased risk of VTE, she was discharged on a prophylactic enoxaparin regimen for 6 weeks.

Discussion

This case illustrates minimal antenatal care in a pregnancy with KTS and known venous varicosities secondary to the condition. A review of the literature revealed that pregnancies with KTS have a higher risk for thrombotic events over haemorrhagic complications (1,2). Thromboprophylaxis should be considered in pregnancies with KTS, both antenatally and postnatally (1). In the small cohort of cases reported, mode of delivery has been guided by patient choice with consideration of factors secondary to reported conditions in the literature i.e. AV malformations over lower segment (2,3). Favourable obstetric outcomes can be achieved in KTS with multidisciplinary team input. However, being a rare disease, there is a paucity of evidence to guide management which can pose challenges for pregnancy-related care.

References

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