The anaesthetic management of a parturient with vascular Ehlers-Danlos syndrome

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Introduction

Vascular Ehlers Danlos Syndrome (EDS type IV) is a rare autosomal dominant disorder of type III procollagen which predisposes to severe vascular complications including arterial dissection and rupture.[1] We describe the delivery of a woman successfully managed by caesarean section (CS) under neuraxial anaesthesia with careful titration of vasoactive drugs.

Case Report

A 34-year-old woman presented in the second trimester of her fourth pregnancy after investigation for a family history of sudden cardiac death due to aortic rupture. She had clinical features suspicious of EDS including joint hypermobility and skeletal disproportion. Imaging for vascular abnormalities was unremarkable but is a poor predictor of bleeding. The patient's mother died of an aortic dissection with near-normal aortic root dimensions on previous imaging.

To minimise the vascular stress of advancing pregnancy, she underwent an elective caesarean section at 34+6 weeks. A general anaesthetic was considered relatively contraindicated due to the hypertensive effect of laryngoscopy and the need to monitor for symptomatic dissection. A combined spinal-epidural was selected to avoid these issues, allow flexibility for top-up to maintain anaesthesia in the event of procedural complications and to allow the mother to be awake for the birth. We used a spinal dose of 2.4mls 0.5% heavy bupivacaine and 300mcg diamorphine. In addition to standard monitoring, we used invasive arterial blood pressure monitoring.

A healthy infant was delivered with an estimated blood loss of 550 mL in the mother. A slow oxytocin bolus was sufficient to manage uterine tone. There were no postoperative complications with mother and baby discharged on the fifth postoperative day.

The diagnosis was confirmed postnatally by genetic studies and a heterozygous COL3A1 mutation (c.4294C>T, p.Arg1432Ter) which has been widely reported in Vascular EDS.[2]

Discussion

- Vascular EDS has a substantial impact on long-term patient outcomes with a median survival of 54 years for women. 80% of patients experience a medical or surgical complication by 40 years of age.
- High rates of arterial rupture present substantial peripartum and anaesthetic challenges. Case series have given conflicting estimates of pregnancy-related mortality, with one noting an 11.5% risk of death, whilst a subsequent analysis found that women below 30 years demonstrated higher rates of death but not conclusively explained by pregnancy-related complications.
- There is limited evidence to guide obstetric and anaesthetic decision making. Delivery via CS can be successfully achieved by proactive planning and avoidance of haemodynamic fluctuations driven by bolus administration of vasoactive medications commonly used in pregnancy.

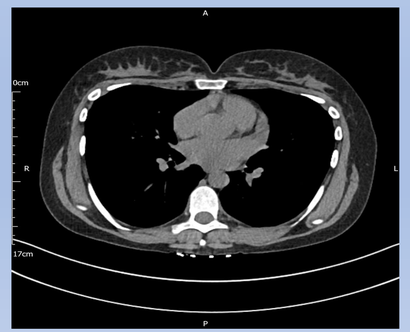


Fig.1 Patient's CT Angiogram Aorta

References

1.Pepin M, Schwarze U, Superti-Furga A, et al. Clinical and genetic features of Ehlers-Danlos syndrome type IV, the vascular type. N Engl J Med 2000; 342: 673-80

2.Pepin M, Schwarze U, Rice K et al Survival is affected by mutation type and molecular mechanism in vascular Ehlers–Danlos syndrome (EDS type IV). Genetics in Medicine. 2014 Dec;16(12):881-8

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