A case of Postpartum Haemorrhage in a Patient with Classical Ehlers-Danlos Syndrome

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Background

- Ehlers-Danlos syndrome (EDS) is a group of inherited connective tissue disorders that are characterised by joint hypermobility, skin hyperextensibility and tissue fragility [1].
- A new international classification of EDS was established in 2017 that describes 13 subtypes. Hypermobile EDS is the most common followed by the classical and vascular subtypes [2].
- Obstetric complications in patients with EDS include, but are not limited to, placenta previa, preterm delivery secondary to preterm rupture of membranes, rapid labours, uterine and vascular ruptures, post partum haemorrhage (PPH) and significant perineal trauma [3]. These complications have been documented for individuals with hypermobile and vascular EDS however data is limited for the less common classical EDS [4].

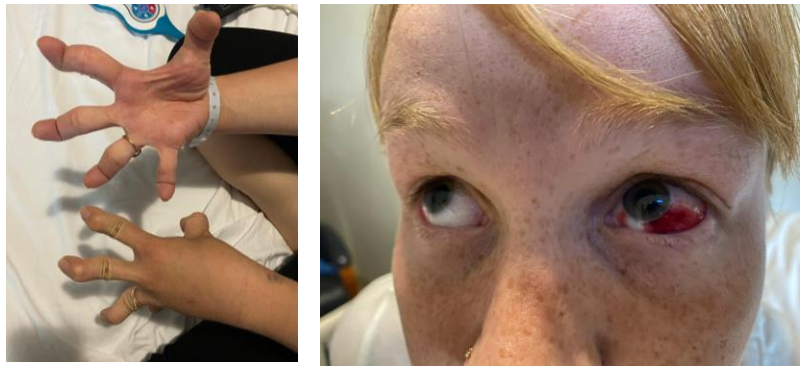

Figure 1: Clinical manifestations of EDS (written permission attained to publish images)

Case

A 31-year-old female with a background of classical EDS, mitral valve prolapse and varicose veins presented in active labour at 41+1 weeks gestation. She had a previous uncomplicated vaginal delivery and an elective caesarean section for placenta previa. She delivered a 4355g live infant by vaginal delivery with episiotomy. Active third stage management was undertaken with intramuscular ergometrine 500mcg-oxytocin 5IU and the placenta was delivered complete with controlled-cord-traction. The woman had a postpartum haemorrhage that was managed with fundal massage, oxytocin infusion, 1000mcg misoprostol per rectum and a catheter in birth suite before the prompt decision was made to transfer to theatre.

In theatre an examination under anaesthesia revealed the episiotomy site was not bleeding and no other perineal trauma could be visualised. A manual evacuation of the uterus demonstrated no retained placenta and the uterus was well contracted throughout however bleeding continued. A laparotomy was performed to rule out uterine rupture or previous caesarean scar dehiscence and no evidence of either was found. Intramuscular carboprost 0.25mg was given and 15 minutes later intramyometrial carboprost 0.5mg was given. The bleeding settled and the episiotomy site was sutured. Additional sutures were placed on both the laparotomy wound and episiotomy site given the increased skin laxity. The total blood loss was 2500mL.

Discussion

- The development of the diagnosis and management of EDS is still a relatively recent development in medicine and hence the obstetric manifestations and management options have limited literature.
- There is no agreed safer mode of delivery in patients with EDS and no literature on vaginal deliveries following caesarean section. This case is an example of a successful vaginal delivery following caesarean section in a patient with a background of classical EDS but highlights the risk of massive postpartum haemorrhage and potential concerns about uterine rupture.

References

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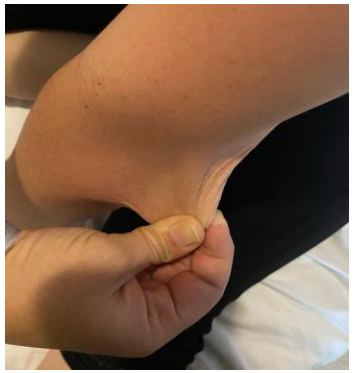

Figure 2: Clinical manifestations of EDS continued

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