A CASE OF MYOTONIC DYSTROPHY LIKELY CONTRIBUTING TO A MASSIVE POSTPARTUM HAEMORRHAGE

BINURI ATTANAYAKE[1,2]
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BACKGROUND

Myotonic dystrophy is a progressive multi-system, autosomal dominant genetic disorder which is characterised by impaired muscle relaxation after voluntary contraction due to repetitive depolarisation of the muscle membrane. It can be subdivided into two: DM1 (Steinert disease) and DM2 (proximal myotonic myopathy). Steinert disease is the more common and severe subclassification. While it's not well researched in pregnancy, the main risks include miscarriage, preterm labour, preeclampsia and postpartum haemorrhage (PPH).

CASE REPORT

A 19 year old nulliparous patient with a background of Steinert disease and limited antenatal care, had a vaginal birth complicated by a 3.5L PPH causing haemodynamic instability. PPH management involved bimanual compression, urgent transfer to theatre, uterotonics, a Bakri balloon and 5 units packed red blood cells while the anaesthetics and ICU teams worked to stabilise her. The PPH was thought to be due to atony and retained membranes. She recovered well postpartum and was given an iron infusion prior to discharge. She was also extensively counselled postpartum regarding future pregnancies and encouraged to engage in antenatal care in the future to help optimise her haemoglobin antenatally and plan for potential complications arising from her myotonic dystrophy.

DISCUSSION

The mechanism by which myotonic dystrophy increases the risk of pregnancy-related complications is not well researched. One theory is that weakening of the uterine muscles can lead to uterine atony, therefore increasing the risk of PPH.
Studies have shown an increase in other pregnancy-related complications including miscarriage, preterm labour, polyhydramnios and prolonged labour. Additionally, congenital onset of myotonic dystrophy can result in neonatal complications such as respiratory and swallowing abnormalities, poor muscle tone, clubfoot and ventriculomegaly. Therefore, pregnancy planning and a multi-disciplinary approach to managing a high risk pregnancy is imperative. This would include genetic counsellors pre-pregnancy and antenatal care by an obstetric team and midwifery team. Diagnostic genetic testing, ultrasounds and counselling by maternal fetal medicine and management of other medical complications of myotonic dystrophy by obstetric medicine. Peripartum counselling by neonatology and anaesthetics is also beneficial. This multi-disciplinary approach can help to optimise maternal and neonatal outcomes. Unfortunately, in this case the patient had limited engagement with antenatal care, therefore addressing barriers to engagement can also be beneficial for optimizing outcomes.

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