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A Case Report of Anaesthesia of a Parturient with Moyamoya Disease

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Case Report

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ABSTRACT

The Moyamoya (MMD) Syndrome is a progressive intracranial occlusive arteriopathy. It is mainly prevalent in east Asia and in Japan population. Anaesthesia for Cesarean section could be challenging due to the normal physiological changes during pregnancy, and it can be more challenging to keep normal haemodynamic goals if pregnancy is associated with moyamoya disease. The available evidence suggests that the mode of anaesthesia and maintenance of haemodynamic status is priority over the exact method of delivery in Moyamoya patients a special anaesthesia technique was provided to our patient to control the fluctuations in Blood pressure and thus maintaining brain perfusion in our patient. The patient had a successful anaesthesia modality without any perioperative complications and the outcome was a healthy 2.9 kgs baby.

INTRODUCTION

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The Moyamoya Syndrome is a progressive intracranial occlusive arteriopathy. It is a rare disease that is mainly prevalent in East Asia and in Japan population. The incidence of this condition is highest in Japan with an incidence rate of 0.54 per 100,000 person-years[1]. It is almost twice as frequent in women as in men [2]. Anaesthesia for Caesarean section can be challenging due to the normal physiological changes during pregnancy, and it can be more challenging if pregnancy associated with MMD disease. We are reporting the first case of a pregnant multigravida MMD disease parturient from the Coombe Women and Infant Hospital, who had planned Elective Caesarean Section under combined spinal epidural anaesthesia.

CASE REPORT

32 years-old multigravida, weighing 55 kilograms, native Irish women with known diagnosis moyamoya, presented at 37 weeks of gestation for a planned caesarean section. She had history of neurological sensory disturbance of the left body side and headaches. Her symptoms started 7 months before the beginning of pregnancy she had previously episodes of severe headache and dizziness. Past Ct angiography showed Rt middle cerebral artery occlusion with hypertrophy at the origin of rt Anterior cerebral artery consistent with a small 2mm aneurysmal dilation. Abdominal and pelvic Ultrasound revealed a singleton live foetus with normal foetal heart, only noted to have Foetal Malposition with an oblique lie. Placental was Upper anterior. On physical examination, the patient was alert and oriented female with insignificant systemic exam. The patient was admitted in hospital for close monitoring as she was found to have anaemia (Hb 8.8g/dl). She received Intravenous Iron treatment two weeks before her planned section [3].

The patient was attending clinics of a combined multidisciplinary care team including Obstetrician, internal medicine, neurology, and anaesthesiology). A combined Spinal epidural anaesthesia was the chosen mode of anaesthesia for the caesarean section. Two large bore IV cannulas were inserted. A left radial Arterial Line was also applied. Standard monitoring as per ASA guidelines was all applied and the patient was prepared for the Anaesthesia in sitting position. Fluid preloading of 500 mls of warm Hartmann's solution was administered. Phenylephrine infusion was also attached primed and ready to administer [4] (Table 1).

Table 1. Symptoms experienced by the patient during the Case study.

Disease Symptoms	Experienced symptoms
Occasional Dizziness	Yes
Photophobia	Yes
limb Weakness	No
Visual Symptoms	No
Heating Symptoms	No
Vomiting	Yes
Headache	Yes

A spinal block was performed with 1.5 mls of 0.5% heavy bupivacaine mixed with 25 mcg fentanyl administered Intrathecally. Then an epidural catheter was placed and tested with 5mls of 0.25% bupivacaine. The patient was then kept supine with left lateral uterine displacement in place. Phenylephrine infusion was started at 3ml/hr and titrated to always maintain the systolic blood pressure between 110-130 mmgh. A second 5 mls incremental bolus of 0.5% bupivacaine were given at a 5 min interval (total of 15 mls) till a sensory block at T5 was achieved. Total

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volume of Phenylephrine infused was 500 mcg during surgery, and it was off 20 min after baby delivery. Ultrasonography revealed a single live fetus with a fetal heart rate of 130 bpm, in cephalic presentation, with fundal placenta.

Procedure was uneventful throughout; Total blood loss was estimated to be 450 mls. The patients received total IV fluids of 1500 mls. The patient was kept normothermic, normocarbia, and normotensive. Bp always kept within target systolic blood pressure range. A healthy baby with a normal APGAR score was delivered. Patient received Paracetamol and Diclofenac IV for analgesia. Prospective planned close monitoring of vitals in the HDU, hydration status and pain controls were maintained while women were admitted in HDU. The women were provided with Low Molecular weight heparin according to hospital protocols for deep vein thrombosis prophylaxis.

DISCUSSION

MMD is rare condition in non-Japanese population. This report is presenting a case of moyamoya disease in an Irish Parturient. The case was diagnosed after delivery of the first baby as she presented with stroke like symptoms without any residual long term neurological deficit.

Given the complexity of this case, the patient was cared for by a multidisciplinary team. The team worked to produce the safest delivery plan and to optimize her condition to the best outcomes [5]. Key components to keep hemodynamic stability intrapartum included avoiding blood pressure fluctuations, minimizing pain and give adequate intravascular fluid filling. The prevalence and incidence is approximately 3.16 cases and 0.35 cases per 100,000 people, respectively with a female-to-male ratio of 1.8:1. Mortality rates are approximately 10% in adults and 4.3% in children. MMD is associated with other diseases, including thyrotoxicosis, sickle cell anaemia, Down's syndrome, coarctation of the aorta and hypertension, which warrant consideration due to their implications during anaesthesia and surgery [6]. Pharmacotherapy in MMD patients includes the use of antihypertensives, anticoagulants or antiplatelet agents (e.g. Aspirin).

CONCLUSION

Anesthetic management needs both careful evaluation and management of both sickle cell disease and moyamoya disease. Mismatch of cerebral blood flow, Cerebral Metabolic Rate of Oxygen ($CMRO_2$), due to hypotension can aggravate ischemia, for example, dehydration secondary to vomiting in the last trimester of pregnancy and blood loss during delivery. In our local institution there are no consistent guidelines for managing pregnancy and delivery in women with MMD. Pregnancy-induced increase in estrogen and progesterone can accentuate vasodilation, and physiological increase in blood flow can lead to rupture of fragile blood vessels resulting in intracranial bleed, and this is more common in the antepartum period especially after 24 weeks of gestation. Neuraxial anaesthesia was recommended in the literature in almost all cases of MMD. Some studies had reported use of epidural anaesthesia a safe option; however this report is reporting a case of MMD under Combined Spinal Epidural anaesthesia.

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