Case Report

iMedPub Journals
http://www.imedpub.com/

Critical Care Obstetrics and Gynecology
ISSN 2471-9803

2016
Vol.2 No.3:15

DOI: 10.4172/2471-9803.1000123

Post-partum Haemorrhage Following Elective Caesarean Section in a Patient with MELAS Syndrome: A Case Report

Natasha Prosser[1] and Christopher Griffin[1,2*]

[1]Department of Obstetrics and Gynaecology, King Edward Memorial Hospital, Subiaco, WA 6008, Australia

[2]Departments of Obstetrics and Gynaecology, St John of God Midland Public Hospital, Midland, WA 6056, Australia

[*]**Corresponding author:** Christopher Griffin, Department of Obstetrics and Gynaecology, St John of God Midland Public Hospital, Midland, WA 6056, Australia, E-mail: Christopher.Griffin@SJOG.org.au

Rec date: Apr 22, 2016; **Acc date:** May 17, 2016; **Pub date:** May 24, 2016

Citation: Prosser N, Griffin C. Post-partum haemorrhage following Elective Caesarian Section in a patient with MELAS syndrome: A case report. Crit Care Obst&Gyne. 2016, 2:15.

Abstract

Mitochondrial myopathies are a heterogenous group of diseases which may be caused by mutations of genes encoded by nuclear or mitochondrial DNA. Some of these disorders may affect single systems, or arise in a constellation of features representing distinct disorders. There is little information about MELAS in pregnancy. This is a case report upon a patient with mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes (MELAS) with a deterioriating picture of lactic acidosis precipitating disseminated intravascular coagulation (DIC) and a major post-partum haemorrhage (PPH) following an elective Caesarean section.

Keywords:

Caesarean section; Post-partum haemorrhage (PPH); MELAS syndrome

Introduction

Mitochondrial myopathies are a heterogeneously diverse group of disorders of the mitochondrial respiratory chain. MELAS A > G3256 mutation may occur as frequently as 236/100000 according to a recent Australian study [1]. Clinical presentation is protean in nature, involving muscle dysfunction, lethargy, vomiting, seizures and stroke-like episodes. There are few reported cases of MELAS and associated complications in pregnancy. We present a case of significant lactic acidosis following Elective Caesarian Section and associated DIC and PPH.

Case Report

A 28-year-old primigravid woman was diagnosed at 7 weeks gestation with MELAS syndrome. Due to earlier stroke-like episodes, the patient was started in the first trimester on aspirin 100 mg daily, which was ceased at 36 weeks.

The pregnancy was otherwise uneventful with a planned Caesarean section (CS) due to fetal malpresentation at 38 weeks gestation. The procedure was performed under a combined spinal-epidural anaesthetic. A healthy baby girl was delivered with no intraoperative complications and an estimated blood loss of 300 ml. The patient was transferred to the Adult Special Care Unit (ASCU) for post-operative monitoring and observation.

During post-partum observations, the patient was haemodynamically stable with a uterus that was firm, central and not enlarged. However, ongoing moderate per-vagina (PV) loss prompted a blood gas analysis and coagulation profile. The lactate had risen to 2.2 mmol/L from 1.8 mmol/L pre operatively. The fibrinogen was reduced at 1.5 g/L, APTT raised at 41 seconds and platelet count was normal at 187×10^9/L.

A Rotational thromboelastometry (ROTEM) (TEM International GmBH, Munchen, Germany) performed at this time showed fibrinogen deficiency, and 16 units of cryoprecipitate (CP) were administered. Despite this, bleeding continued and a repeat ROTEM indicated refractory low fibrinogen so a further 8 units of CP was administered. At this stage the uterus was noted to be enlarging and firm.

The patient was transferred to the operating theatre for examination under anaesthesia. An 800 ml intrauterine clot was evacuated and a Bakri balloon (Cook Medical Inc., Bloomington, IN, USA) was inserted. Intra-operative investigations showed the patient's haemoglobin had dropped to 69 g/L (preoperative 121 g/L), platelets now 67×10^9/L and lactate had risen to 3.6 mmol/L.

Further resuscitation with fresh frozen plasma, platelets, packed red blood cells (PRBC), tranexamic acid and calcium chloride was undertaken. An estimated total blood loss at this time was 1500 ml.

Due to the post-operative unstable metabolic and haematological picture, the patient remained ventilated and was admitted to the Intensive Care Unit for monitoring. Continuing blood loss occurred with a rise of the lactate to 4.6

mmol/L. Uterine artery embolization was undertaken with subsequent reduction in PV bleeding. Thereafter with supportive care the patient made an uncomplicated course of recovery occurred and the patient was discharged home.

Discussion

MELAS is a mitochondrial disease caused by mutations of mitochondrial DNA which may be as high as 236/100000 of the general population 1. MELAS can arise from mutations in one of several genes of mitochondrial DNA, however 80% of patients have A > G3243 mutation. These mutations impair mitochondrial function, specifically manufacturing of proteins essential for respiratory chain function, energy production and oxygen usage [2].

MELAS is inherited via the maternal line, as mitochondria are contributed to the zygote by the ovum; however both males and females can be affected [2].

Diagnosis is suspected based on clinical picture and confirmed by ragged-red fibers on histopathological examination of muscle tissue biopsy [3]. The pathogenesis of MELAS is poorly understood and it has a complex array of presenting symptoms. Typically, individuals have a period of normal development before the appearance of signs and symptoms in early childhood. Early symptoms may include recurrent headaches, muscle weakness, pain, and loss of appetite, vomiting and seizures. Stroke-like symptoms usually occur before the age of 40 and involve altered consciousness, hemiparesis, migraine type headaches and seizures. Other conditions are also associated with MELAS, namely diabetes and cardiovascular disease such as cardiomyopathy, Wolff-Parkinson-White syndrome and disturbances in myocardial conduction [2,3].

There are a small number of reported cases of women with MELAS syndrome experiencing coagulation and bleeding issues during labour. Aggarwal reported PPH in a woman with MELAS syndrome following spontaneous vaginal delivery secondary to placental accretes, requiring 4 units of PRBC [4]. Dessole described a non-elective caesarian section for labour dystocia complicated by PPH and subsequent DIC secondary to atonic uterus, resulting in a hysterectomy to control blood loss [5]. Of interest, in this case the DIC was characterized by an increase in cross-linked degredation products, initially normal platelets and low borderline fibrinogen, which the authors attribute to the MELAS syndrome. However, case reports of emergency caesarians in three women with MELAS syndrome reported no bleeding complications [6-8]. Rosaeg observes a single patient who delivered her first child via vaginal delivery, second by Non-elective CS and third by Elective CS with no bleeding issues noted [6,7].

DIC is a systemic process characterized by pathological activation of the coagulation cascade. Excessive fibrin deposition leads to clot formation causing micro vascular occlusion and organ dysfunction. Clotting proteins and platelets are excessively consumed from ongoing coagulation resulting in severe bleeding [9].

Acidosis is associated with clinical coagulopathy however the contributing mechanisms are poorly understood. Acidosis can develop as a result of trauma, hypoxia and blood loss, however it is also postulated that lactic acidosis can lead to DIC. To date, there are limited studies with inconsistent results examining such causality. These are largely in vitro and animal models; however an in vivo study demonstrates a reduction in clotting time with acidosis and an early animal study showed the development of DIC in four out of five dogs following infusion of lactic acid [10-13].

The principles of management of lactic acidosis are to identify and correct the underlying condition, and to restore adequate perfusion and oxygen delivery to tissue. Treatment with an alkalizing agent such as sodium bicarbonate is not supported in the evidence so far and may contribute to worsening intracellular acidosis [14].

This is the first case in the published literature to describe the evolution of a PPH in a MELAS affected patient with the salient features of a rising lactate and falling fibrinogen preceding the clinical signs and diagnosis of PPH.

Patients with MELAS syndrome are at high risk of PPH due to the high circulating level of lactate. Minimal deterioration of the metabolic status of the patient ie the rising lactate level following the initial surgery is thought to have contributed to the subsequent massive PPH. Recognizing this very high risk point for predisposing patients with MELAS syndrome is important in the post-delivery care and prevention of major PPH.

Consent

Written informed consent was obtained from the patient for publication of this case report.

Conflict of Interest

The authors do not have funding or conflicts of interest to declare.

References

1. Manwaring N, Jones MM, Wang JJ, Rochtchina E, Howard C, et al. (2007) Population prevalence of the MELAS A3243G mutation. Mitochondrion 7: 230-233.
2. Taylor RW, Turnbull DM (2005) Mitochondrial DNA mutations in human disease. Nat Rev Genet 6: 389-402.
3. Pavlakis SG, Phillips PC, DiMauro S (1984) Mitochondrial myopathy, encephalopathy, lactic acidosis, and strokelike episodes: a distinctive clinical syndrome. Ann Neurol 16: 481-488.
4. Aggarwal P, Gill-Randall R, Wheatley T (2001) Identification of mtDNA mutation in a pedigree with gestational diabetes, deafness, Wolff-Parkinson-White syndrome and placenta accreta. Hum Hered 51: 114-116.
5. Dessole S, Capobianco G, Ambrosini G (2003) Postpartum hemorrhage and emergency hysterectomy in a patient with

mitochondrial myopathy: a case report. Archives of Gynecology and Obstetrics 267: 247-249.

6. Moriarty KT, McFarland R, Whittaker R, Burch J, Turnbull HE, et al. (2008) Pre-eclampsia and magnesium toxicity with therapeutic plasma level in a woman with m.3243A>G melas mutation. J Obstet Gynaecol 28: 349.
7. Rosaeg OP, Morrison S, MacLeod JP (1996) Anaesthetic management of labour and delivery in the parturient with mitochondrial myopathy. Can J Anaesth 43: 403-407.
8. Löwik MM, Hol FA, Steenbergen EJ (2005) Mitochondrial tRNALeu(UUR) mutation in a patient with steroid-resistant nephrotic syndrome and focal segmental glomerulosclerosis. Nephrol Dial Transplant 20: 336-341.
9. Levi M, Ten Cate H (1999) Disseminated intravascular coagulation. N Engl J Med 341: 586-592.
10. Nagasue N, Iwaki A, Yukaya H, Koyanagi N, Kobayashi M (1977) Disseminated intravascular coagulation and refractory shock induced by splanchnic metabolic acidosis. Surg Gynecol Obstet 144: 519-524.
11. Martini WZ, Holcomb JB (2007) Acidosis and coagulopathy: the differential effects on fibrinogen synthesis and breakdown in pigs. Ann Surg 246: 831-835.
12. Engstrom M, Schött U, Romner B, Reinstrup P (2006) Acidosis impairs the coagulation: A thromboelastographic study. J Trauma 61: 624-628.
13. Martini WZ (2009) Coagulopathy by hypothermia and acidosis: mechanisms of thrombin generation and fibrinogen availability. J Trauma 67: 202-208.
14. Darlington DN, Kheirabadi BS, Delgado AV, Scherer MR, Martini WZ, et al. (2011) Coagulation changes to systemic acidosis and bicarbonate correction in swine. J Trauma 71: 1271-1277.