Case Report

McArdle disease causing rhabdomyolysis following vaginal delivery

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Summary

McArdle disease (glycogen storage disease type V) is a rare, autosomal recessive disorder with an incidence of roughly 1:100,000. Despite concern that labour could predispose these patients to muscle damage, there are no evidence-based guidelines for the management of labour and delivery in this population. We describe the case of a nulliparous parturient with both McArdle disease and adenosine monophosphate deaminase 1 deficiency who developed rhabdomyolysis after vaginal delivery. In the absence of common triggers, we believe that prolonged pushing efforts contributed to the increase in postpartum creatinine kinase. There are no previous cases of postpartum rhabdomyolysis after caesarean or assisted vaginal delivery within 45 min. We recommend that practitioners be alert to the possibility of rhabdomyolysis occurring with greater than 2 h of pushing efforts in parturients with McArdle disease.

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Introduction

McArdle disease (glycogen storage disease type V) is a rare, autosomal recessive disorder with an incidence of approximately 1:100,000 [1]. The genetic mutation causes a defect in muscle glycogen phosphorylase, which can lead to symptoms of muscle fatigue during exercise and cause myoglobinuria, kidney injury and rhabdomyolysis [2]. Because periods of intense activity aggravate myopathies, there is concern that active labour could predispose these parturients to muscle damage. However, there are no case reports of adverse events during vaginal delivery in parturients with McArdle disease. Here, we present the case of a nulliparous parturient with both McArdle disease and adenosine monophosphate deaminase 1 (AMPD1) deficiency who developed rhabdomyolysis after vaginal delivery.

Report

A 23-year-old gravida 1, parity 0 at 39 weeks gestation presented for scheduled induction of labour. She was 157 cm, 96 kg (body mass index 39 $kg.m^{-2}$), with a Mallampati class 1 airway. She was diagnosed with McArdle disease, confirmed by genetic testing, at 15 years of age after experiencing rhabdomyolysis from playing softball. Testing at this time was also positive for AMPD1 deficiency. She reported approximately six to seven episodes of rhabdomyolysis a year, at times without a clear trigger. Throughout this pregnancy, her renal function was within normal limits (baseline creatinine 44 $\mu mol.l^{-1}$) and her baseline creatine kinase (CK) was 3300 $units.l^{-1}$. Her self-reported baseline CK outside of pregnancy was around 10,000 $units.l^{-1}$. Her pregnancy was notable for one episode of rhabdomyolysis without a known trigger, associated with a CK increase to 33,000

units.l^{-1} at 23 weeks gestation. This was treated with intravenous (i.v.) hydration and there was no resultant kidney injury. She was on a balanced diet of carbohydrates and proteins and an exercise plan to maintain her exercise tolerance. She was evaluated by the obstetric anaesthesia team antenatally and was advised to receive early epidural analgesia during labour, adequate hydration, strict glucose monitoring and to maintain a blood glucose level above 5.6 mmol.l^{-1}. Adequate peripartum glucose management is thought to help prevent myopathy in this population by obviating the need to break down glycogen stores [1], so she was specifically counseled to continue oral intake during labour.

Cervical ripening was achieved with vaginal misoprostol followed by a Cook cervical ripening balloon (Cook Incorporated, Bloomington, IN, USA) to facilitate cervical dilatation. Blood results on admission were within normal limits (CK 2300 units.l^{-1}; Creatinine 45 μmol.l^{-1}). A lumbar epidural at L3–L4 interspace was placed uneventfully and a bilateral T10 dermatomal level was confirmed before initiation of oxytocin for augmentation of labour. During labour, her blood glucose levels were initially between 4.4 mmol.l^{-1} and 5.0 mmol.l^{-1}. She was allowed oral intake of juice, popsicles and glucose gels and was started on an i.v. Ringer's lactate solution with 5% dextrose at 150 ml.h^{-1} to maintain blood glucose > 5.6 mmol.l^{-1}. There was adequate urine output during labour (~100 ml.hr^{-1}) and CK was relatively unchanged after artificial rupture of membranes (4484 units.l^{-1}). Her labour was uneventful, with the first stage lasting 27 h and the second stage lasting 159 min. She pushed for 149 min resulting in successful vaginal delivery of a healthy baby (APGAR score: 8 and 9 at 1 min and 5 min, respectively). Her epidural catheter was removed 1 h after delivery.

Blood samples were taken approximately 45 min after delivery at which time her CK was 24,090 units.l^{-1} and her creatinine 57 μmol.l^{-1}. She received 1l i.v. bolus of Ringer's lactate solution and her dextrose-containing maintenance infusion was increased to 200 ml.hr^{-1}. She noted shoulder pain after delivery that resolved spontaneously and had no symptoms of muscle weakness. A nephrology consultation was requested due to myoglobinuria and CK elevation, with recommendations to continue supportive care, maintain urine output greater than 200 ml.hr^{-1} and alkalinise the urine if urine pH decreased below 6.5 (no alkalinisation was required). Her CK peaked at 28,500 units.l^{-1} on postpartum day two and eventually decreased to 9708 units.l^{-1} by postpartum day five. No symptoms of muscle pain or weakness were reported. Creatinine remained at baseline throughout the rest of her hospital admission. She was discharged home on postpartum day five with scheduled follow-up.

Discussion

McArdle disease results from a genetic mutation that limits the ability of skeletal muscle cells to access glycogen stores during exercise, leading to increased muscle catabolism [2]. Uterine activity is typically normal in patients with McArdle disease due to functional smooth muscle phosphorylase [2]. It has previously been suggested that parturients with McArdle disease require no special precautions apart from glucose administration to help prevent muscle fatigue [2]. Previous reports show no adverse events from either vaginal or caesarean delivery in parturients with McArdle disease [1–3]. Cochrane et al. have reported forceps-assisted delivery after 45 min of stage two labour without adverse events [3].

To our knowledge, there are no documented cases of rhabdomyolysis following labour and delivery in parturients with McArdle disease. A prolonged second stage of labour in a nulliparous parturient is defined as greater than 3 h with epidural analgesia [4]. This parturient actively pushed for 2.5 h, and while not defined as prolonged, this could have contributed to an increase in postpartum CK. Because we pre-emptively addressed potential dehydration with i.v. fluid administration and by allowing oral intake during labour, we speculate that prolonged pushing efforts were the most likely trigger. This assumption is supported by near-normal baseline plasma CK level during the first stage of labour. Skeletal muscle exertion during prolonged pushing efforts, along with possible injury to pelvic floor muscles, might have been contributory. Additionally, this parturient's AMPD1 deficiency—characterised by an inability to break down adenosine monophosphate to inosine monophosphate—may have been contributory. Although it is unlikely that the AMPD1 deficiency alone impaired muscle performance [5], the combination of two disorders could have increased her susceptibility to rhabdomyolysis.

Rhabdomyolysis can be triggered by a wide range of exercise demands in patients with McArdle disease and it is difficult to predict a safe cut-off for exertion. In our case, there was minimal CK elevation in the first stage of labour, but a large CK increase after the second stage of labour. This led us to conclude that 149 min of pushing during the second stage of labour was the most likely cause for rhabdomyolysis. With an incidence of approximately 1:100,000 [1], little clinical information is available to guide management of labour. Three case reports show that the risk of developing rhabdomyolysis from either caesarean or forceps-assisted deliveries (within 45 min of pushing efforts) is low in this population [1–3]. Although caesarean delivery is a more convenient option, it is not without risks. For example, caesarean delivery is associated with an increased risk of maternal morbidity, including postpartum haemorrhage, infection, sepsis and maternal death compared with vaginal delivery [6]. In

contrast, a forceps-assisted vaginal delivery, commonly recommended to shorten the duration of second stage of labour, can be associated with an increased risk of perineal tears and anal sphincter injury [7]. Our case suggests that even vaginal delivery is not without risk even if the second stage of labour is not prolonged. Therefore, we recommend that practitioners be alert to the possibility of rhabdomyolysis with greater than 2 h of pushing efforts in parturients with McArdle disease, and to consider an operative vaginal delivery when appropriate. Regardless of the mode of delivery, we also recommend continued biochemical vigilance in the postpartum period to ensure early detection and appropriate management of rhabdomyolysis.

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