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Case Report

Myotonic dystrophy complicated by Clostridium Perfringens sepsis resulting in uterine rupture

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Abstract

Myotonic dystrophy is a rare autosomal dominant degenerative neuromuscular and neuroendocrine disease which can also affect the uterine muscles. In pregnancy, issues can arise with abnormalities in all three stages of labour. We report a unique case in which a woman with classic myotonic dystrophy suffered an occult uterine rupture following post-term induction of labour and an apparently uncomplicated forceps delivery. She was diagnosed with fulminating Clostridium perfringens infection two days postpartum and required an emergency hysterectomy. We postulate that abnormal myotonic uterine contractions in labour caused focal uterine ischemic necrosis. Subsequent infection with the commensal organism Clostridium perfringens resulted in uterine infection and fulminating retroperitoneal sepsis through the disruption of the uterine wall. Despite the atypical presentation of uterine rupture and sepsis, timely recognition, judicious use of intravenous antibiotics, multidisciplinary involvement, early imaging, and undelayed surgery averted maternal mortality.

Key Words:

Myotonic dystrophy, uterine rupture, Clostridium perfringens infection, postpartum sepsis

Introduction

Myotonic dystrophy is a rare autosomal dominant degenerative neuromuscular and neuroendocrine disease which can affect the uterine muscles. In pregnancy, issues can arise with abnormalities in all three stages of labour. Infection with Clostridium perfringens may further increase the risk of uterine rupture. Literature is sparse linking myotonic dystrophy with Clostridium perfringens infection and uterine rupture. A Medline search combining these terms in various combinations resulted in two or less case reports. In fact, the only association between myotonic dystrophy with Clostridium perfringens infection is in animal medicine [1].

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Case Presentation

The patient was a 32-year-old woman known to have myotonic dystrophy. The condition was diagnosed after cataract surgery in her mid 20's with inability to release muscle movements especially in her hands, mouth and tongue. Genetic testing confirmed the diagnosis with 163 to 233 cystosine-thymine-guanine (CTG) repeats, which is in classic range. She was counselled by the genetics services, seen by the cardiologists and had pre-conceptual counselling. She fell pregnant spontaneously. She was otherwise low risk with a body mass index (BMI) of 25 and a non-smoker. All antenatal booking investigations were within normal limits. In addition to standard antenatal care, there was a consultation with a maternal fetal medicine (MFM) specialist and an obstetric anaesthesia review. All routine tests continued to be within normal range. Fetal growth on ultrasound was monitored regularly and was within normal growth parameters with no evidence of polyhydramnios.

She attended Delivery Suite at Wellington Hospital at 41+3 for a post-term induction. Two doses of 2mg prostaglandin E2 (Pfizer®) were administered over two days. On the afternoon of the second day, an artificial rupture of membranes was performed and an oxytocin infusion commenced. An epidural was placed by a consultant anaesthetist because of the known risks of general anaesthesia with myotonic dystrophy. After a total of 12 and half hours of labour, including 90 minutes of maternal pushing, a low-cavity forceps delivery was performed due to inadequate progress in second stage. Pathological changes on the cardiotocography were noted once that decision had been made. There was no intrapartum bleeding. The baby weighed 3602 grams with first and fifth minute Apgar scores of 5 and 9 respectively. A high second-degree tear had extended from the right mediolateral episiotomy and this was repaired in routine fashion. A per rectal (PR) examination was performed with no sutures felt and paracetamol and diclofenac suppositories given. No excessive uterine bleeding was observed. An oxytocin infusion was started prophylactically to avoid a post-partum haemorrhage. Over the next 48 hours the patient developed constant fundal pain associated with nausea and vomiting. Her observations remained normal. On examination there was generalised abdominal tenderness particularly over the uterine fundus. Lochia was noted to be normal. Empiric intravenous amoxicillin/clavulanate was started for possible early endometritis on day 1 postpartum when C-reactive protein (CRP) was noted to be 300 mg/dL (normal range <3 mg/dL) despite a normal white cell count (WCC). She then developed numbness of her right leg, weakness of left hip flexion and faecal incontinence. The only new clinical finding was absent anal sphincter tone. Initial investigations included a magnetic resonance imaging (MRI) of the spine, which excluded an epidural haematoma or abscess. The patient rapidly deteriorated with severe back pain, pallor and an observable rapidly spreading and deep red discolouration over her left hip, buttock and left flank which was exquisitely tender. The uterine fundus remained subinvoluted and tender though she did not have heavy vaginal bleeding. An urgent computerised tomography (CT) scan showed a heterogeneous collection of air and fluid in the soft tissue of the left parametrial region. Antibiotics were changed to intravenous Meropenem and Clindamycin on advice from the infectious disease team. Urgent multidisciplinary consultation ensued with obstetric, infectious diseases, general surgery, radiology and the anaesthetic services. She was taken to the operating theatre for an examination under anaesthesia and exploratory laparotomy was performed with two obstetric and gynaecology specialists and a general surgeon. The uterus was grossly inflamed and necrotic. A 4 cm full thickness defect was noted in the lower segment associated with a large area of necrosis and bubbling gas under the peritoneal reflection. The gas tracked retroperitoneally adjacent to the left ureter. No rectal injury was identified. Due to the findings, a consensus was reached to proceed with a subtotal hysterectomy. This was then performed in conjunction with extensive intrapelvic and retroperitoneal lavage. A three-way catheter was left in situ as a drain and a vacuum-assisted wound closure dressing applied. She was transferred to the intensive care unit (ICU) for post-operative care, was commenced on total parenteral nutrition (TPN) and continued to receive intravenous antibiotics. She returned to the operating theatre two days later as planned. At repeat laparotomy there was no active infection. Extensive lavage was again performed, the drain removed and the abdominal wound closed. She returned to ICU for immediate post-operative care and transferred to the postnatal ward later that day. The following day, the pre-operative high vaginal swab and intra-operative swab from peri-uterine fluid grew Clostridium perfringens. The intravenous antibiotic was changed to Tazocin® (Pfizer). Histology of the uterus reported inflammatory necrosis of the endometrium which extended to the serosal surface. She remained as an inpatient for a further week and made a satisfactory recovery. She had a total of eight days of TPN and nine days of intravenous antibiotics. Genetic testing of her baby daughter has been declined and follow-up to date shows no signs of myotonic dystrophy.

Discussion

Myotonic dystrophy is a rare degenerative neuromuscular and neuroendocrine disease. It is an autosomal dominant tri-nucleotide repeat disorder with the affected gene located on chromosome 19. Individuals with more repeats have an earlier onset and more severe form of the disease [1]. Pregnancy may be associated with marked exacerbations of myotonia and muscle weakness, or symptoms may be unchanged. Deterioration may occur early in pregnancy but is most severe in the third trimester with rapid improvement after delivery [1].

There is an increased risk of first and second trimester miscarriage, stillbirth, polyhydramnios, preterm delivery and placenta praevia. Abnormalities of all three stages of labour have been described, due to weakness and uncoordinated action of the uterine musculature, although there are no reported adverse effects of specific induction agents in patients with myotonic dystrophy. The baby may be affected with congenital myotonic dystrophy [2]. It is unclear whether myotonic dystrophy increases the risk of uterine rupture. A Medline search combining these two conditions only gave one result. The case was of a woman with placenta accreta with the baby being affected with congenital myotonic dystrophy which resulted in a 33 week stillbirth. It is also unclear whether Clostridium perfringens infection increases risk of uterine rupture. A Medline search combining these two conditions yielded two results, one of which was of an ectopic pregnancy [2,3]. Clostridium perfringens is a gram-positive rod-shaped anaerobic spore-forming bacterium [4]. It is commonly found throughout nature but has been isolated from human colonic flora, skin and vagina. It is the most common bacterial agent implicated in gas gangrene, and the mortality rate with treatment is 20-30%. Myonecrosis can spread as fast as 2cm per hour [5]. Uterine infection with this organism is a potentially fatal disease infrequently seen in obstetric practice. The manifestations of Clostridium perfringens uterine infection are variable, ranging from endometritis to gas gangrene and fulminant septicaemia [6]. There is a case similar to ours describing postpartum uterine gas gangrene [7]. In this case, Clostridium perfringens infection was noted and treated but there was also co-infection with Toxoplasma gondii. Presence of both organisms can explain severe myonecrosis but this woman did not require a hysterectomy despite infection with two organisms; unlike ours who required a hysterectomy with only one pathogen. There is also another case of a woman requiring hysterectomy after ruptured tuboovarian abscess with Clostridium perfringens infection [8]. However, this woman was postmenopausal and the final histology revealed endometrial carcinoma. In our case, we postulate that Clostridium perfringens was a commensal organism which within abnormal musculature, under stress from labour, formed an ischemic zone resulting in a rupture without bleeding. Regardless of the causal organism, sepsis is a leading contributor to all maternal mortality [9]. In this case, despite an atypical presentation of sepsis and rupture, timely recognition, judicious use of intravenous antibiotics and a multidisciplinary team approach helped save her life. Imaging was performed early and the source of infection was removed surgically in a timely manner as necrotic tissue has poor perfusion and consequently tissue concentration of antibiotics may not reach the target tissue adequately.

Acknowledgement
None

Declaration of Interest
None

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