Massive abscess with prolonged respiratory failure due to newly diagnosed myotonic dystrophy
A case report
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Abstract
Rationale: Myotonic dystrophy is a progressive multisystem genetic heterogeneous disorder. General anesthesia with opioids increases the risk of prolonged postanesthetic respiratory recovery in myotonic dystrophy patients.

Patient concerns: A 20-year-old previously healthy woman was transferred to our emergency department for further workup of respiratory failure, and massive ascites with abscess caused by endometriosis. Hypercapnic respiratory failure persisted under intensive care unit (ICU) management, but finally improved after cessation of fentanyl as a sedative agent.

Diagnosis: Myotonic dystrophy type 1.

Interventions: Massive ascites with abscess was accordingly managed by drainage, antibiotics, and an antifungal agent. Myotonic dystrophy type 1 was confirmed after molecular genetic testing revealed a cytosine-thymine-guanine repeat length of 400 to 450 in the DMPK gene.

Outcomes: The patient was discharged without complications on hospital day 69.

Lessons: Myotonic dystrophy should be considered when hypercapnic respiratory failure persists in sedated ICU patients. Opioids should not be used for perioperative management of patients with myotonic dystrophy.

Abbreviations: CO2 = carbon dioxide, CT = computed tomography, CTG = cytosine-thymine-guanine, ICU = intensive care unit, MD = myotonic dystrophy, MG = myasthenia gravis, MRI = magnetic resonance imaging.

Keywords: abscess, endometriosis, hypercapnic respiratory failure, massive ascites, myotonic dystrophy

1. Introduction
Myotonic dystrophy (MD) is a progressive multisystem genetic heterogeneous disorder. Both myotonic dystrophy type 1 (MD1) and myotonic dystrophy type 2 (MD2) are autosomal-dominant and characterized by skeletal muscle weakness and myotonia, abnormalities of cardiac conduction, respiratory complications, iridescent cataracts, and other abnormalities. MD1 is the most common muscular dystrophy in adults, and results from expansion of a cytosine-thymine-guanine (CTG) trinucleotide repeat in the DMPK gene on chromosome 19q 13.3. Respiratory complications are more common and severe in MD1 than in MD2. Respiratory failure and precipitating pulmonary complications are often triggered by sedatives, anesthetics, and neuromuscular blocking agents. Respiratory involvement is the leading cause of mortality. Carbon dioxide (CO2) insensitivity in MD1 patients has been reported as the main cause of alveolar hypventilation.

We managed a patient with prolonged respiratory failure during ICU management with fentanyl sedation, and MD1 was finally diagnosed. The patient and her father provided written, informed consent to publish the details of her condition.

2. Case presentation
A 20-year-old, previously healthy woman felt abdominal distension for 1 year and went to see a clinic about 7 weeks before the admission. A large volume of ascites was detected by ultrasound at the clinic, and she was referred to a hospital for workup. Her past medical history was unremarkable. She denied any allergy to medicine or food. Her mother had died of subarachnoid hemorrhage at 44 years old when the patient was 10 years old. Computed tomography (CT), magnetic resonance imaging (MRI), cytology of ascites, and culture of ascites revealed endometriosis (Fig. 1A and B). Laparoscopic biopsy and appendectomy were performed. The pathological diagnosis was endometriosis of the appendix vermiformis and endometrial...
tissue. She was discharged without complication on postoperative day 8. However, she complained of vomiting and was rehospitalized on day 11 after discharge. CT showed ileus with a large volume of ascites, and an ileus tube was inserted. The condition of the patient deteriorated with the appearance of respiratory distress and pleural effusion, and she was managed in the intensive care unit (ICU). Respiratory failure deteriorated even after insertion of bilateral chest tubes and intubation. The patient was transferred to our facility on hospital day 18 for further workup of ascites and respiratory failure. On arrival in the emergency room, vital signs were as follows: temperature, 37.0°C; heart rate, 55 beats/min with regular rhythm; respiratory rate, 16 breaths/min; blood pressure, 94/58 mm Hg; and oxygen saturation, 100% on a respirator in Synchronized Intermittent Mechanical Ventilation mode. Glasgow Coma Scale score was 5 (E1V1M3) because of sedation with propofol and fentanyl. On physical examination, the patient was thin (height, 155.8 cm; weight, 37.2 kg; body mass index, 15.3 kg/m²), coarse crackles were heard bilaterally, and the abdomen was distended. Arterial blood gas analysis revealed the following: pH, 7.52; PCO₂, 43.1 mm Hg; PO₂, 34.5 mm Hg; HCO₃⁻, 34.4 mmol/L; base excess, 10.6 mmol/L; hemoglobin, 7.2 g/dL; and lactate, 1.52 mmol/L. Contrast-enhanced CT showed a large abdominal abscess (Fig. 1C). The patient was admitted to the ICU and needed respiratory support with mechanical ventilation. Abscess (2000 mL) was drained initially and culture of the drained specimen revealed *Bacteroides fragilis* and *Prevotella oris*, both of which were beta lactamase-positive. Meropenem and vancomycin were

**Figure 1.** (A) Contrast-enhanced CT of the abdomen shows gross ascites with no other significant abnormality. (B) T2-weighted MRI shows high-intensity ascites. (C) Contrast-enhanced CT of the pelvis shows gross ascites with peripheral enhancement on the day of admission. (D) Contrast-enhanced CT of the pelvis shows decreasing ascites with peripheral enhancement on hospital day 48. (E) High-resolution CT of the chest without contrast shows bilateral GGOs on hospital day 19. (F) High-resolution CT of the chest without contrast reveals GGOs have improved by hospital day 48. CT = computed tomography, GGOs = ground glass opacities, MRI = magnetic resonance imaging.
We have presented the case of a patient who was diagnosed with MD1 and experienced prolonged respiratory failure during ICU management. The prolonged respiratory failure with hypercapnia could not be explained simply by heart failure. MD1 patients reportedly show reduced ventilatory response to hypercapnic stimulation and are prone to pulmonary complications under general anesthesia. We had not recognized the characteristic facies because she had been intubated upon arrival to the emergency room and was directly admitted to the ICU. Her mother had died of subarachnoid hemorrhage 9 years earlier and her father did not know his wife’s family history of MD1. Myopathy, brainstem lesions, and Guillain-Barré syndrome (GBS) should be taken into account for differential diagnosis of MD1 with prolonged respiratory failure. The concentration of creatine kinase was 11 IU/L on the day of admission and was always low, and thus incompatible with myopathy. Serial CT and MRI did not show any abnormalities of the brainstem, allowing us to rule out brainstem lesions. Cerebrospinal fluid did not show albuminocytological dissociation and her history was incompatible with GBS. Myasthenia gravis (MG) is yet another differential diagnosis, and is difficult to differentiate from MD1. In this case, both anti-acetylcholine receptor antibody and antibodies against muscle-specific kinase were negative. In addition, genetic testing revealed expansion of the CTG repeat in the DMPK gene, which was more compatible with MD1 than MG.

Massive ascites arose as a complication of endometriosis and caused postoperative abdominal abscess. The ascites could not be managed by antibiotic therapy, and drainage was needed. Positive results were obtained for Candida tropicalis and an antifungal agent was administered. Massive ascites related to endometriosis shows a high risk of recurrence. Non-Caucasian ethnicity, nulliparous status, and reproductive age are risk factors for endometriosis-related ascites and this patient had all of these. MD1 might not cause endometriosis, because no reports have shown an association with MD1. One report described significant, selective reduction of serum concentrations of immunoglobulin (Ig)G in MD1 patients. In this case, the normal serum IgG level of 1511 mg/dL might not have been associated with MD1.

General anesthesia has been indicated to increase the risk of prolonged postanesthetic respiratory recovery in MD1 patients. Extubation succeeded after the first operation in the previous hospital, but failed during the ICU hospitalization in our facility. The abdominal and pulmonary infections might have been reasons for the prolonged respiratory failure, but opioid use was recognized as the main cause of respiratory complications with MD1. CO2 insensitivity in MD1 patients has been reported as the main cause of alveolar hypoventilation, so we consider that opioids could interfere with and deteriorate the CO2 insensitivity and cause prolonged hypercapnic respiratory failure. Fentanyl was administered at the previous hospital and we continued this agent for 22 days, and so could represent a major reason for the persistence of hypercapnic respiratory failure. Opioid-free management is recommended during the ICU management of patients with MD1.

4. Conclusions

We managed the case of a 20-year-old MD1 patient whose respiratory failure was prolonged due to the use of opioids as sedative agents. MD1 might be considered when hypercapnic respiratory failure persisted during ICU management. Opioids should be avoided using during the perioperative or ICU management of MD1 patients.

Figure 2. Line 1 shows the DMPK gene from a normal control. Line 2 shows the DMPK gene for our patient. M, molecular size; B, BamHI; E, EcoRI. Southern blotting of EcoRI-restricted DNA of line 2 E shows a CTG repeat (black arrow) above the normal control of around 9.42 kbp (red arrow). This difference indicates expansion of the CTG repeat. CTG = cytosine-thymine-guanine.
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