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CLINICAL CASE

MYASTHENIA REVEALED FOLLOWING LAPAROTOMY. A CASE REPORT.

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ABSTRACT

Myasthenia (muscle weakness) is a rare neuromuscular disease of which respiratory failure is the main complication. The accidental discovery of such disease in the perioperative period is rare and potentially serious.

We report a case of a woman who underwent emergency operation for appendiceal peritonitis, and failed repeatedly at weaning from postoperative mechanical ventilation. The usual etiologies such as postoperative respiratory complications, ventilator-associated pneumonia, acute respiratory distress syndrome complicating the septic shock or having no impact on it, and neuromyopathy's resuscitation were considered, researched, examined or eliminated.

Faced with the diagnostic impasse and the obvious weaning failure, another interview revealed signs of muscle fatigue which led to the diagnosis of myasthenia gravis decompensated perioperatively. Once the diagnosis was confirmed by means of a neostigmine test, the specific treatment began, particularly through plasma exchange sessions, and the process of weaning resumed. The result was complete weaning. A three-month follow-up showed a stable patient with no significant muscular disability.

KEY WORDS: Myasthenia ; Ventilatory weaning ; Postoperative respiratory complications ; anesthesia ; neuromuscular disease.

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INTRODUCTION

Myasthenia is a rare neuromuscular disease related to a defect of transmission of nerve impulses between a motor neuron and skeletal muscle. Respiratory failure is the main complication of myasthenia, and can be life-threatening [1]. The anesthetic management of patients with myasthenia is unique due to drug interactions with anesthetics, and postoperative respiratory complications it can cause [2]. The accidental discovery of myasthenia in the perioperative period is rare and potentially serious given that no particular precaution is taken to anesthetize those patients.

Through a case of myasthenia revealed following an urgent exploratory laparotomy, we expose the difficulty in

giving a diagnosis after the weaning failure from postoperative mechanical ventilation.

CASE REPORT

Madam A.A., Moroccan, 49, housewife, no history of myasthenia, visited emergency room for acute abdominal syndrome due to abdominal pain and vomiting. The examination showed a conscious patient with a respiratory rate of 30 cycles per minute, heart rate 125 beats per minute, arterial pressure 105/55 mm Hg, temperature 39°C and generalized abdominal defense. Abdominal ultrasound showed abundant intraperitoneal effusion (Pouch of Douglas, paracolic gutters and pouch of Morison). The diagnosis of acute generalized peritonitis

was made and the patient was admitted to the operating room.

After filling with 2000 ml saline, induction of anesthesia was achieved with propofol 150 mg, fentanyl 350 micrograms and rocuronium 50 mg (estimated weight at 60 kg). The monitoring was standard and neuromuscular blockade was not monitored. The patient received antibiotic therapy with gentamicin (320 mg) and metronidazole (1g). Surgical exploration found acute peritonitis caused by neglected appendicitis. Therefore, Appendectomy with abundant wash-out and drainage of the peritoneal cavity were performed.

The patient was admitted to the resuscitation room to manage a septic shock due to peritonitis, for a period of 3 days. After clinical improvement (apyrexia, wake up from sedation), biological improvement (J4: urea 0.48 g/l, creatinine 12.7 mg/l, procalcitonin 1.2 ng/ml) and gasometric improvement (lactate 0.8 mmol/l, base excess at -1.8), a ventilator weaning process was started on the 6th day and has resulted in a weaning failure. A neuromyopathy's resuscitation was suggested, but this diagnosis was eliminated because of the muscle biopsy data.

Another interview with the patient's family helped raise the issue of muscle fatigue and ptosis since young age, for which the patient was consulted a month before admission. However, the possibility of neuromuscular blockade was not suggested. The diagnosis of myasthenia was suspected and confirmed by a positive neostigmine test. Antiacetylcholine receptor (AChR) antibody dosage was positive and CT scan (computed tomography) was normal. The patient received pyridostigmine at an initial dose of 360 mg six times daily. Then, respiratory weaning was started. However, the existence of swallowing disorders complicated respiratory weaning. Three sessions of plasma exchange were performed. These plasma exchange sessions resulted in the regression of pulmonary aspiration and enabled the stopping of mechanical ventilation on the 25th day of hospitalization. A three-month follow-up showed a patient with muscle tone and normal palpebral aperture with no occurrence of pulmonary aspiration during swallowing, while receiving a dose under 240 mg of pyridostigmine per day.

DISCUSSION

Myasthenia is a chronic autoimmune neuromuscular disease characterized by muscle weakness which is aggravated by exercise [1]. On the etiopathogenic level, there are three forms: the most common is characterized by the presence of antibodies directed against the acetylcholine receptor (AChR) at the post-synaptic neuromuscular junction; the second form involves antibodies directed against muscle specific tyrosine kinase (MuSK); the third form is distinguished by the absence of anti-AChR and anti-MuSK antibodies [3]. In the first form, clinical fatigue appears when destruction of postsynaptic receptors exceeds 75%. The thymus has a probable role in the genesis of autoantibodies: thymic hyperplasia was found in 65% of cases and thymoma in 15% of cases. [1] The clinical manifestations of myasthenia are related to the damage of the striated muscles. These manifestations are summed up in fatigue from exercise improved by rest. All areas can be damaged, including the extraocular muscles (often a means by which the disease is discovered), the

muscles of the face, neck, limbs and respiratory muscles. Swallowing disorders and respiratory failure are what makes myasthenia a severe disease [3]. Myasthenia is frequently associated with other autoimmune diseases: rheumatoid arthritis, lupus, and Hashimoto's thyroiditis [1]. Our patient showed, during the weeks before hospitalization, ptosis and muscle fatigue, which allowed suggesting the diagnosis. However, neither the diagnostic exploration nor treatment were performed due to the negligent behavior of the patient.

The diagnostic was confirmed based on a positive anticholinesterase test (neostigmine or edrophonium) [4, 5]. The presence of anti-acetylcholine receptor antibodies is an additional element, but not necessary for diagnosis, since 10 to 15% of myasthenia cases are seronegative [1, 2]. The presence of anti-acetylcholine receptor was sometimes used as the only paraclinical element of diagnostic value [6, 7]. A thoraco-mediastinal assessment must be undertaken in the search for thymoma.

The evolution of myasthenia and the majority of myasthenic syndromes evolve through relapsingremitting. These outbreaks can be spontaneous, cyclical, or secondary to a trigger factor. Most often, the case is due to medication non-adherence, infection, physical or surgical aggression, drug administration where active ingredients interfere with or affect the neuromuscular junction (antibiotics in general practice) [8].

The discovery of an unnoticed myasthenia following emergency surgery is rare. The discovery of myasthenia was demonstrated following cardiac surgery [4], locoregional anesthesia for trauma surgery [6], or cesarean section [5, 7]. It is necessary to consider this disease when faced with a difficulty at weaning from mechanical ventilation or unexpected respiratory distress.

In our case, several conditions were involved: the ignorance of the disease, severe deep-seated abdominal infection, surgical intervention, and stress of hospitalization in resuscitation.

The myasthenic patient receives anesthesia to undergo Thymectomy as a specific surgical treatment of myasthenia. The critical points in the perioperative period are postoperative respiratory complications, the use of neuromuscular blocking agents, and the use of drugs affecting the neuromuscular junction. Pre-anesthetic consultation should allow the assessment of the disease and the patient according to the classification scale of Myasthenia Gravis Foundation of America [9] to optimize preoperative treatment. Our patient did not benefit from this step. General anesthesia for a myasthenic patient should follow an established protocol specific for each team. Currently, two techniques are recommended: inhalation anesthesia without neuromuscular blocking agents, or total intravenous anesthesia with or without neuromuscular blocking agents [2, 10].

No neuromuscular blocking agent is contraindicated. Their use must be rational (avoiding long-acting neuromuscular blocking agents and saving succinylcholine for anesthesia on a full stomach), the dose should be adjusted (always reduced by 25-75% depending on the molecule and other defects), and their activity should be systematically monitored [2, 8]. However Skedros et al [5] and Cheung et al [8] each reported a case of postoperative decompensation of myasthenia without prior administration of neuromuscular blocking agent during induction of anesthesia. Other drugs that can be used in the perioperative period are the barbiturates, etomidate, morphine, and nitrous oxide. It is necessary to avoid ketamine (postsynaptic destabilizing effect), benzodiazepines (central and peripheral muscle relaxant effects), and halogenated (potentiating the activity of neuromuscular blocking agents, but not contraindicated). It was not known that our patient was myasthenic, thus the anesthetic protocol used was in conformity with the uses of anesthesia in a situation of full stomach for an emergency visceral surgery.

Even if it was known that the patient had a history of myasthenia, the anesthetic method would not change because the absence of succinylcholine in Moroccan public hospitals would require the use of rocuronium as was the case. The dose could hardly have been reduced given the context of a full stomach. Monitoring of neuromuscular blockade could have been performed to optimize the dose and monitor the reversal in the morning. The therapeutic management involves four components. Firstly, patient education to recognize warning signs of an outbreak, the eviction of trigger factors, and rational use of drugs. Secondly, the use of cholinesterase inhibitors (pyridostigmine and ambenonium) that must be initiated in the hospital. Thirdly, the selection of patients requiring thymectomy [10].

Finally, the management of severe cases undergoing resuscitation for a treatment specific to immunosuppressants, corticosteroids or plasma exchange. [3] Once the diagnosis was made, our operated patient underwent an anticholinesterase treatment allowing recovery of muscle strength and achievement of respiratory weaning.

CONCLUSION

Although rare, clinical signs that may suggest myasthenia should be researched systematically during the preanesthetic visit interview, even in the case of emergency. In the intraoperative period, monitoring of neuromuscular blockade is essential. In the postoperative period, myasthenia must be considered in the case of difficult ventilatory weaning. These precautions will help adapt the anesthetic protocol to these patients, and will help avoid prolonged ventilation leading to many complications.

PATIENT CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

COMPETING INTERESTS

The authors declare no competing interests.

AUTHORS' CONTRIBUTIONS

The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the <u>Recommendations for the Conduct, Reporting, Editing,</u> and <u>Publication of Scholarly work in Medical Journals</u> of the <u>International Committee of Medical Journal Editors</u>. Indeed, all the authors have actively participated in the redaction, the revision of the manuscript and provided approval for this final revised version.

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