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Salima Moez Meherali
Aga Khan University

Yasmin Parpio
Aga Khan University

Tazeen Ali
Aga Khan University

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Case Report

Critical illness myopathy

Salima Moez Meherali, Yasmin Parpio, Tazeen Saeed Ali

School of Nursing, Department of Community Health Sciences, The Aga Khan University, Karachi.

Abstract

Critical illness myopathy (CIM) is a syndrome of widespread muscle weakness and neurological dysfunction which can develop in critically ill patients receiving intensive care. CIM are often distinguished largely on the basis of specialized electrophysiologic testing or muscle and nerve biopsy and its causes are unknown, though they are thought to be a possible neurological manifestation of systemic inflammatory response syndrome usually developing in

patients after a brief period of stay in the Intensive Care Unit (ICU). This case report aims to analyze the Clinical feature, diagnosis and treatment of CIM of 60 years old male case with Chronic Obstructive Lung disease (COPD) admitted to the intensive care. Health professionals working at critical care unit should be aware that any ICU patient may develop CIM.

Introduction

Until recently, in most of the critical care units, survival to hospital discharge was the primary outcome.

However, the body of knowledge is now being developed to focus more on health-related quality of life. It is widely recognized that patients admitted to the intensive care unit often develop subsequent difficulties with neuromuscular function,¹ whereby Critical illness Myopathy (CIM) is the most common one. In CIM, myopathies developed to critical patients even after a brief period of stay in the intensive care unit.¹ It occurs in 25-63% of patients who have been mechanically ventilated for \geq one week. Recent studies have revealed that myopathies are in fact more prevalent than neuropathies and early recognition and interventions would have better patient outcome.²

Case Report

A 60 year old male with past history of COPD presented in Emergency Department (ED) with tachypnoea, cyanosis, hypoxaemia, hypercapnia and acidosis. The physicians in the ED diagnosed status asthmaticus. Patient was intubated, and started on intravenous methylprednisolone, intravenous theophylline, and aerosol bronchodilators. A chest radiograph showed a right perihilar infiltrate suggestive of pneumonia. The patient was admitted to ICU. He received multiple antibiotics along with lorazepam and midazolam intravenously for sedation and was kept on sliding scale of insulin. No neuromuscular blocking agents were used during the hospital course.

The hospital course was complicated by the development of ARDS and later tension pneumothorax requiring a chest tube. During this time period, the patient's mental status changed indicating multiple conditions like: hypoxia, hypercapnia, and sedating drugs and hence, his length of stay in ICU increased. Later, patient's mental status gradually improved, ARDS got resolved and gradually lung compliance improved. After spending a month in ICU, he was extubated. At that time, it was noted that patient has marked muscle wasting and diffuse weakness of his extremities more distally than proximally. His muscle strength was graded 2 to 3 on a scale of 5 proximally and 0 to 2 distally. His sensory systems and reflexes were normal. The muscles of the neck and respiratory system were not weak, and there were no other significant findings on neurological assessment.

Discussion

This case was subjected to a number of differential diagnoses. The most likely diagnoses at that time was Electrolyte disturbance (e.g. hypokalaemia, hypercalcaemia, hypermagnesaemia, hypophosphataemia) and other metabolic derangements. Guillain-Barré syndrome was unlikely, even though the patient had symmetric weakness, his reflexes were normal and he did not have sensory disturbances, autonomic dysfunction, or involvement of the

extraocular or facial muscles. Stroke typically presents as an acute focal neurologic deficit with an upper motor neuron pattern of weakness. This patient had generalized weakness of the lower motor neuron type without significant cerebral or brain stem dysfunction. Hopkins syndrome, sometimes called acute post-asthmatic amyotrophy, is an acute attack of muscle weakness that arises in a small number of patients with asthma, not necessarily occurring in ventilated patient.²

Critical illness myopathy (CIM) can present with acute-onset weakness in the critical care setting and has been reported in patients with COPD, status asthmaticus, ARDS and sepsis who have received steroids in high doses and non-depolarizing neuromuscular blocking agents.³

To confirm the diagnosis of CIM and differentiate it from other illnesses, electromyogram (EMG) and nerve conduction (NC) were done which showed reduction of amplitude and duration of muscle compound action potentials and \pm decreased amplitude of sensory nerve action potentials which is common in both CIM and critical illness polyneuropathy (CIP).⁴

It is not easy to classify as strictly CIP or CIM. Therefore, differentiating between both disorders, the needle EMG was done which shows fibrillation potentials and positive sharp waves of low amplitude, particularly in distal muscles. Creatine phosphokinase (CPK) levels were relatively normal (170units/L), consistent with a myopathy. Finally, muscle biopsy was done which demonstrated myopathy as there was myosin loss. The physical examination also revealed that muscle weakness and wasting was more prominent in the proximal than the distal extremities. Initially, reflexes were present but disappeared during the course of the diseases. The presentation of this patient was consistent with critical illness myopathy.

This case illustrates a common problem in critical care setting that is usually unrecognized. The term critical illness myopathy describes a group of myopathies, which commonly occur in critically ill patients.^{2,5} The risk factors of CIM are sepsis and systemic inflammatory response syndrome (SIRS) which disturbs the microcirculation throughout the body.³ The release of inflammatory mediators during sepsis allows the passage of toxic substances (e.g. neuromuscular-blocking agents, and corticosteroids).³ Multi-organ dysfunction syndrome is also identified as a significant risk factor of CIM,^{6,7} but its mechanism remains unknown.⁶ The use of neuromuscular-blocking agents such as vecuronium has been associated with CIM development.² Furthermore the use of Aminoglycosides have been cited as a risk factor,^{1,2} but not been shown as an independent risk factor,² but may adversely affect "axons" in the hyper permeable state along with sepsis.²

Today it is agreed that the SIRS is responsible for the release of a number of proinflammatory cytokines and other metabolites into the blood accounts for the muscle organ failure of CIM that is similar to other organ failures seen in patients with sepsis.² External factors trigger the onset of CIM are high-dose steroids, administered to critically ill patients with respiratory distress syndrome or asthma muscle protein depletion is much more rapid and extensive in the critically ill patients coupled with inappropriate nutritional support lead to the development of myopathies.⁶

The clinical features of CIM are quite nonspecific, the most important of which are generalized muscle weakness and atrophy more distally than proximally,⁶ which are more severe than expected from immobilization alone, and delayed weaning from the respirator⁷ as occurred in the patient presented above. Both factors inevitably result in a prolongation not only of ICU treatment. In addition, CIM causes secondary complications such as pneumonia, deep vein thrombosis and lung embolism.⁷

The diagnosis of CIM is challenging and it is usually diagnosed during the process of delayed weaning from mechanical ventilation and progressive muscle weakness.⁵ The other diagnostic tests which would be useful in the patient are Serum electrolytes, cerebrospinal fluid analysis, imaging studies of the spinal cord, electro diagnostic examination and muscle and nerve biopsy.

In treatment clinicians should minimize the use of steroids and neuromuscular blocking agents, particularly vecuronium.⁷ Earlier therapeutic intervention in physical therapy may prove to be of benefit, to prevent the disuse atrophy.⁶ Lack of physical activity, in fact, may amplify the catabolic effects of cortisol on muscle protein degradation.^{3,8} Nutritional and supplemental therapies include protein and amino acid supplementation, antioxidant therapy, and hormonal therapy are also found helpful in treating CIM.⁸ Novak and colleagues identified that glutamine supplementation were associated with decreased mortality; shorter hospital length of stay and a lower incidence of ICU acquired myopathies.⁹

Further, testosterone hormonal therapies are also beneficial in the treatment of myopathies in male ICU patients.¹⁰ The testosterone derivative oxandrolone may improve muscle strength in the case of muscular dystrophy.¹⁰ Finally, tightly controlling glucose levels with I/V, insulin infusions are also recognized useful in the treatment and

prevention of CIM.¹¹

Critical-illness myopathy is fairly neglected in critical care setting. Standardized screening for weakness is uncommon, and persistent weakness as a sequela of critical illness is usually not recognized by physicians in the ICU. Mostly patients develop this condition after they are moving out from ICU, either in the hospital or as an outpatient which may lead to the development of complications and affect the quality of life of the survivor.

The management should be aimed to optimize the care available for CIM patients in the short- and long-term by having multidisciplinary approach.

Critical care nurse and physicians should assess the patients who are on high doses of steroids and neuromuscular blocking agents in every shift to minimize the steroid doses according to patients' condition. In addition, balanced nutritional requirement adjusted with protein intake and range of motion exercises is essential for the prevention of muscle atrophy. Health professionals should be aware about CIM, to provide interventions to prevent early detect and manage for the quality of life of critically ill patients.

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