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Case Report

Critical Illness Polyneuromyopathy-

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ABSTRACT

Critical Illness Polyneuromyopathy (CIPNM) is frequently present in critically ill as a certain degree of symmetric extremity paresis and respiratory muscle weakness. The consequences of this complication may last for months or years after severe illness. It prolongs the stay in ICU and dependence on mechanical ventilation, increases long-term disability and care costs. We report a 58-year old female patient admitted to our Intensive Care Unit for acute respiratory insufficiency due to influenza pneumonia and acute respiratory distress syndrome. Thirty-three days of mechanical ventilation and 11 days of extracorporal membrane oxygenation were complicated by severe CIPNM, tetraparesis, mental disorders, and difficulties in weaning off mechanical ventilation. No specific therapy is available for treatment of CIPNM. Preventive, supportive and rehabilitation measures are discussed in the article.

Keywords: Critical illness polyneuropathy and myopathy; Polyneuromyopathy; Mechanical ventilation; Sepsis; Multiple organ failure

INTRODUCTION

Critical Illness Myopathy (CIM) and Critical Illness Polyneuropathy (CIP), alone or in Combination - Polyneuromyopathy (CIPNM) - are frequent complications in patients in the Intensive Care Unit (ICU) [1]. The causes of axonal degeneration of sensory and motor axons in critical illness polyneuropathy, and atrophy and necrosis of myofibers in critical illness myopathy are complex and not fully understood; they possibly involve microcirculatory abnormality, metabolic derangements, reversible channelopathy, and bioenergetic dysfunction [2]. CIP and CIM are overlapping syndromes and frequently occur concomitantly involving all extremities and the diaphragm with relative sparing of the cranial nerves, but causing difficulties in weaning off mechanical ventilation [3,4].

CASE PRESENTATION

A 58-year-old female patient with arterial hypertension and hypothyroidism, was admitted to the Department of pulmonology due to pneumonia. For the last seven days at home she was febrile up to 38.9°C, with chills, shivering and dry cough. Upon arrival, chest X-ray showed interstitial and alveolar infiltrates on the left side indicative of pneumonia. Laboratory tests were as follows: RBC 4.3 x 10^{12}/ L, Hb 124 g/ L, Htc 0.39 L/ L, MCV 90.0 fL, L 5.4 x 10^{9}/ L, platelet count 188 x 10^{9}/ L, CRP 181.1 mg/ L, venous blood gas analysis: pH 7.5, pCO_2 4.0 kPa, BE 1.0 mmol/ L, pO_2 5.9 kPa, bicarbonates 23.1 mmol/ L, total CO_2 24.0 mmol/ L, O_2 saturation 85.0%, blood sugar 6.9 mmol/ L, creatinine 92 µmol/ L, urea 6.8 mmol/ L, AST 91 U/ L, ALT 66 U/ L, GGT 607 U/ L, ALP 242 U/ L, K 4.6 mmol/ L, Na 139 mmol/ L, hsTnI 12.5 ng/ L (n.v. < 15.6). Apart from oxygenotherapy and other supportive measures, triple antibiotic therapy consisted of amoxicillin-clavulanic acid, clindamycin, and azithromycin was introduced. However, 72 hours later, a severe respiratory insufficiency developed and the patient was transferred to Intensive Care Unit (ICU). Fiberoptic bronchoscopy and bronchial toilet were performed immediately, but control blood gas analysis, a great progression of bilateral lung infiltrates on chest X-ray, and pulmonary capillary wedge pressure were highly suggestive of severe Acute Respiratory Distress Syndrome (ARDS) requiring mechanical ventilation. An urgent bronchoalveolar lavage sample obtained for microbiological investigation was negative, but rapid influenza A diagnostic test was positive. The antibiotic therapy was switched to meropenem, levofloxacin, and tamoxifen in order to treat Gram-positive and Gram-negative aerobic, and some anaerobic bacteria, possible Legionella bacteria infection, and influenza A, respectively. In spite of intermittent positive pressure ventilation with FiO_2 of 90%, Positive End Expiratory Pressure (PEEP) of 15 mbar, and respiratory rate of 20/min, arterial blood-gas analysis indicated a need for veno-venous Extracorporeal Membrane Oxygenation (ECMO) which was started 24 hours later. The further course of illness was complicated by ventilator associated pneumonia which implicated a prolonged mechanical ventilation. After 11 days, ECMO was no longer needed, but the patient was still dependent on ventilator support. Neurologic examination revealed severe tetraparesis due to CIPNM which made the weaning off mechanical ventilation much more difficult. Brain imaging using multislice computed tomography as well as the lumbar puncture were within normal range. Percutaneous tracheostomy had to be performed, and continuous veno-venous hemodiafiltration had to be started because of acute renal failure.

Repeated bronchoalveolar lavage samples obtained for microbiological investigation revealed nosocomial strains (MRSA, *Pseudomonas sp.*, and *Acinetobacter baumanni*). Antibiotic regimen had to be changed into vancomycin, piperacillin-tazobactam, and colistin. After 37 days, antibiotic treatment and ventilatory support were finally discontinued. Intensive physical therapy was carrying out all the time but with only moderate success. The patient remained permanently dependent on hemodialysis. At the end of treatment, clinical and laboratory findings were satisfactory, which allowed the closing of tracheostomy and the transfer to the hospital for chronic diseases until maximum recovery.

DISCUSSION

CIP affects more than one third of critically ill patients in ICU-s, and reaches up to 100 percent when multiple organ failure occurs [5]. CIM is present in 36 percent of mechanically ventilated patients for asthma, and approximately 70 percent of patients hospitalized in the ICU for at least 7 days [6,7]. Patients with acute respiratory distress syndrome on mechanical ventilation, and patients with systemic inflammatory response syndrome, sepsis, or multiple-organ dysfunction syndrome have a high risk for the development of polyneuromyopathy [8,9]. Although widely present in Intensive Care Units, these complications, in their mild form, usually stay underdiagnosed [10].

Electrophysiologic studies such as electromyography, electroneurography, and muscle and nerve biopsies, are considered the gold standard for aiding in the diagnosis of CIPNM, but are not always possible or easy to perform in critically ill patients [9].

Clinical and ancillary test results should be carefully interpreted to differentiate critical illness polyneuropathy/myopathy from similar weaknesses in this patient population [2].

Differential diagnosis of acute neuromuscular weakness includes Guillain-Barré syndrome, metabolic neuropathies, toxic neuropathies, and neuropathies due to nutritional deficiencies.

Unlike CIP, which develops during a critical illness in ICU,

Guillain-Barré syndrome is usually the cause of admission to ICU [11].

Until present hospitalization, our patient was a healthy, active person with normal neuromuscular status.

A prospective multicenter study of Guarneri et al., showed that severity of muscle weakness was not correlated with the clinical and electrophysiological diagnosis, but the rapidity and completeness of recovery were [12]. At 3 months, three of the five survivors with CIM had complete clinical and electrophysiological recovery and two patients recovered within 6 months. Conversely, patients with CIP or CIP/CIM had a slower recovery, or did not recover at all. Patients with a definite diagnosis of CIM recovered earlier and better than those with CIP, the majority of whom remained severely disabled 1 year after hospital discharge.

There is no specific therapy in the management of CIP and CIM, only preventive and supportive measures such as: intensive treatment of sepsis, reduction of dose and duration of therapy with neuromuscular blocking agents and corticosteroids, early rehabilitation, careful nutritional regimens, anti-oxidant therapy, testosterone derivates, growth hormones, and immunoglobulins [13]. Insulin therapy in diabetic patients improves blood glucose control, and independently reduces the incidence of CIP and CIM. Blood glucosae level between 8,0 and 10,0 mmol/ L is recommended since more intensive regulation carries the risk of hypoglycaemia and the consecutive increase of mortality rate [14].

CONCLUSION

Based on available reference data and our experience, successful treatment of basic disease, intensive control of risk factors, quality supportive therapy, and intensive physiotherapy is the only way to reduce the incidence and the intensity of CIP and CIM. Optimal regulation of blood sugar in diabetic patients may generally also be helpful. A long term rehabilitation following discharge from the hospital is usually necessary.

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