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MANAGEMENT OF POLYNEUROMYOPATHY IN A CRITICALLY ILL PATIENT WITH A LEFT VENTRICULAR ASSIST DEVICE

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ABSTRACT

Critical illness polyneuromyopathy after cardiac surgery is often unrecognized and is a rarely reported clinical condition. It is characterized by more proximal than distal symmetrical flaccid muscle weakness and difficulty in weaning from a respirator. When done in a timely manner, rehabilitation prevents early complications and reduces the length of hospitalization. Rehabilitation leads to better motor outcome, improves short-term and long-term functionality, and results in a better quality of life.

Keywords: critical illness polyneuromyopathy, rehabilitation, LVAD

INTRODUCTION

With more than 15,000 pumps implanted worldwide and a postimplant survival rate of 80% in the first year and 59% in the first 5 years, the left ventricular assist device (LVAD) has become a valuable rescue therapy in terminal heart failure [1]. Critical illness polyneuromyopathy (CIPNM) in a cardiac surgical Intensive Care Unit (ICU) is a commonly underdiagnosed clinical condition. It is characterized by muscle weakness and, unless rehabilitated in time, it leaves behind a certain degree of functional disability and the need for assistance in performing daily [2]. We describe a case report of a patient with quadriparesis as a consequence of polyneuromyopathy after LVAD implantation.

CASE PRESENTATION

A 45-year-old male patient was referred for implantation of a left ventricular assist device (LVAD) under the diagnosis of advanced heart failure. The patient had a history of dilated cardiomyopathy, atrial fibrillation, and left bundle branch block. Due to an episode of ventricular tachycardia, an implantable cardioverter defibrillator was implanted for secondary prevention of life-threatening arrhythmia two years prior. He experienced two episodes of defibrillation shocks during the past two years. During the past 5 years,

he experienced increasing dyspnea upon exertion, fatigue and lower extremities edema. He gradually progressed from New York Heart Association (NYHA) functional class II to NYHA class III despite optimum medical therapy. Several episodes of cardiac decompensation occurred during the past 12 months with need for hospital admission and increasing doses of loop diuretics and inodilators. His past medical history was also positive for obesity, with a BMI of 35.8 kg/m2, chronic obstructive pulmonary disease treated with inhalatory beta-agonist and bronchodilator therapy. His diabetes mellitus type 2 was treated with oral hypoglycemic drugs. His personal history was positive for smoking and moderate alcohol consumption. The patient's 6 minute walk test was 350m.

Extensive diagnostic work-up was undertaken for assessment of suitability for durable mechanical assist device. The pulmonology assessment which included spirometry test, respiratory gas analysis and chest CT revealed partial respiratory insufficiency and diminished ventilatory function of combined, predominantly obstructive type. Right heart catheterization and transthoracic echocardiography showed severe left ventricular dysfunction with an EF of 18%, mildly reduced right ventricular systolic function, mild postcapillary pulmonary hypertension with normal systemic and pulmonary vascular resistance. The neurological examination showed no abnormalities. After careful assessment, the cardiac team decided to proceed with LVAD implantation as a bridge to heart transplant. A HeartMate III was implanted, and the procedure went without intraoperative complications. The patient was transferred in the cardiac surgical ICU with minimal inodilator and vasopressor support and was extubated on the second postoperative day. However, due to an acute hemoglobin drop and suspected acute mediastinal hemorrhage draining into the pleural cavity, surgical revision of hemostasis was performed. Unfortunately, severe hemodynamic compromise ensued, despite higher doses of inotropics and vasopressors, resulting in the development of hemorrhagic shock, severe end-organ hypoperfusion and rising lactate. Urgent venous-arterial extracorporeal membrane oxygenation (VA ECMO) was performed with arterial access via cannulation of right subclavian artery and venous access via cannulation of right femoral vein. Immediate hemodynamic stabilization was achieved, with normalization of blood lactate the following day and doses of inotropic and vasopressor drugs tapered off gradually. The patient was extubated on the fifth postoperative day and the VA ECMO was removed the following day. Shortly after extubation, generalized flaccid muscle weakness was noticed, the inability to perform active movements in the upper and lower extremities, unstable sitting, inability to stand and walk. There was also fecal and urine incontinence. A neurologist was consulted who ruled out a central motor neuron lesion. Due to the unavailability of an electrodiagnostic procedure in the ICU, a working diagnosis of CIPNM was decided upon. In consultation with a physiatrist, intensive rehabilitation was started (passive and actively assisted exercises for the upper and lower extremities, body exercises, as well as respiratory exercises). Intensive nutritional parenteral and enteral support was also initiated to meet the accumulated energy debt from the critical illness. During this period his respiratory gas exchange worsened significantly. Chest X-rays showed signs of bilateral hospital acquired pneumonia. Soon he began to develop signs of systemic infection, which we managed to resolve within 48 hours of extracorporeal blood purification with the Oxiris filter, prolonged targeted antibiotic therapy as well as non-invasive ventilation by means of high-flow oxygen and continuous positive airway pressure ventilation. The generalized muscle weakness persisted despite the patient's stable hemodynamic parameters, gradual improvement of the respiratory status and normal laboratory findings (serum electrolytes, protein and enzyme status, and normal complete blood count). After 2 months of hospitalization at the cardiac surgery clinic, and after sufficient training regarding the use of LVAD equipment, the patient was sent to a rehabilitation clinic for further rehabilitation.

Upon admission the patient was alert and oriented to person, place and time. The cranial nerves were intact. Upon physical examination, active movements in the spine could not be assessed, the patient did not sit and stand on his own. Upper extremities were correctly positioned, with severe muscle hypotrophy and hypotony. Active elevation and abduction were limited to the horizontal level in the right shoulder and below the horizontal level in the left shoulder. Active shoulder rotations were limited in full range, in the elbows, wrists and thumbs active movements were possible in full range, in the other fingers, flexion and extension were possible in full range but abduction was limited in full range. The OK sign was positive in both sides. Active movements

were performed with reduced speed and dexterity. Passive movements in the shoulders were possible in full range without pain. Positioned, the left hand quickly sank to the ground, the right could be held longer. The reflexes of the biceps brachii and triceps brachii muscles were not obtainable, reflex of the brachioradialis muscle was obtained symmetrically. Superficial and deep sensibility was normal. Rough muscle strength was reduced in both fists. He had great difficulty using his upper extremities in daily activities.

The lower extremities were positioned with hips in external rotation, knees in extension, there was hyperpigmentation and trophic changes in both lower legs and feet, with severe muscle hypotrophy and hypotony. Active movements in the hips were limited in all directions, especially abduction and flexion with an extended knee. The knee and ankles were in full range with reduced speed and dexterity. Placed in a positional test, he kept both legs long enough without tremor. Superficial and deep sensibility was preserved. Patellar and Achilles tendon reflexes were on both sights decreased. Rough muscle strength was reduced. From a supine position he sat down with help, sat stably, with one-person support he was able to rise from bed, stood unstable with a tendency to flex his knees, walked short distances by dragging his feet and with the help of a walker and the support of two physiotherapists.

Urinary and fecal incontinence were present. The following tests were performed:

Manual muscle test: Upper extremities: shoulder muscles: flexors, abductors and extensors = 2, elbow flexor and extensor muscles = 3, wrist flexor and extensor muscles = 3, thumb abductor = 2, hand interossei and lumbrical muscles = 3. Lower extremities: hip muscles: flexors and abductors = 2-3, hip extensors = 2, knee extensors = 2-3, knee flexors = 3, ankle flexors and extensors = 3.

The 6-minute walk test on admission was interrupted because the patient was verticalized with the help of two physiotherapists and could walk a maximum of 10 meters with the help of a walker and the support of two.

The Barthel index score for evaluation of the degree of functional independence upon admission was 17, meaning total dependence.

The rehabilitation program lasted 3 weeks and included: manual massage of the upper and lower extremities, respiratory gymnastics, respiratory resistance exercises, actively assisted exercises for the shoulders and hips, active exercises for the elbows, wrists, knees and ankles without resistance. During the rehabilitation this progressed to active resistance exercises, functional occupational therapy for the upper and lower extremities, neck exercises, isometric exercises for the abdominal, paravertebral and gluteal muscles, aerobic exercises, walking exercises with a walker with support from two therapists. This later progressed to walking exercises with one crutch without support.

During rehabilitation, the patient's general condition was stable. Mean blood pressure was around 75 mmHg.

After 3 weeks, the patient's active movements in the extremities were significantly improved and his condition was better. In the upper and lower extremities, he performed active movements in all joints at full range with better strength, speed, and dexterity. The patient walked on discharge, with a crutch without assistance. This was stable, over long distances on flat terrain. The results of the manual muscle test on discharge: shoulders flexors, abductors and extensors = 3, elbows flexors and extensor = 3-4, wrists and hands flexors and extensors = 3-4, hips muscles = 3-4, knees flexors = 4, ankles muscles 4. After 3 weeks his 6-minute walk test was 140 meters. He walked with a forearm crutch.

The Borg scale for perceived effort was used. At the beginning of the test, the grade was 13 (somewhat hard-considerable effort, feels tired, but can continue), at the end of the test 15 (hard-speaking is a challenge, answers short, intermittent with one, two words). SpO2 at the beginning of the test was 99% without oxygen support, at the end of the test 98% without oxygen support. The Barthel index score upon discharge was 54 points - severe dependence.

In terms of incontinence, he had urges for miction and defecation and could control his sphincters. These achievements were also maintained one month after the completion of the rehabilitation.

Three months after LVAD implantation, an attempt for electromyography testing was done. However, it was technically inadequate and with inconclusive findings.

The patient continued to practice the learned exercises at home. Three months after completing his rehabilitation, he walked alone without assis-

tance. The manual muscle test for the muscles of both upper and lower extremities was 4. The 6-minute walk test was 450 meters. The Borg rating at the beginning of the test was 7 (extremely light), at the end of the test, 13 (somewhat hard). The Barthel index score was 98 points – low dependence. He later controlled his sphincter completely.

DISCUSSION

The term CIPNM was initially described by Bolton et al. in 1983 and has been sporadically reported during the management of critically ill ICU patients [3]. This self-limited acute axonal neuropathy is the most common cause of muscle weakness in ICU and not uncommon cause of failure to wean from mechanical ventilation [4].

The incidence of CIPNM ranges from 25% – 83%, depending on the underlying critical illness. [2] It presents in 50-70% of patients, with sepsis lasting longer than 2 weeks [5]. Of all patients who are at least 4 to 7 days on mechanical ventilation, 25% – 33% develop myopathy, and in 58% to 68% it is confirmed by electrodiagnostic testing. Up to 62% of patients failing to wean from respirator develop certain form of neuromuscular weakness [6].

CIPNM after cardiac surgery is a widely underdiagnosed clinical condition. Moreover, these patients had normal neuromuscular status preoperatively. Risk factors include corticosteroid administration, aminoglycoside antibiotics, high doses of catecholamines, hypotensive shock and multiorgan failure [7–11].

Myopathy as dominant component of CIPNM is characterized by more proximal rather than distal symmetric flaccid muscle weakness of variable degree-ranging from paresis to paralysis. Weaning off from a respirator is the consequence of respiratory muscle weakness. Superficial and deep sensibility is preserved. Tendon reflexes are initially preserved, but gradually disappear as muscle weakness progresses. Despite some cases of quadriplegia, the autonomic nervous system is intact, and the bulbar musculature is preserved [2].

Electromyography is a diagnostic tool of choice, but it poses a technical challenge in a typical ICU setting. The unavailability of electro diagnostical procedures, the presence of other electronic devices, the clinical status of the patient, and difficult patient cooperation are some of the many factors that lead to misinterpretation and the difficulty in setting a diagnosis [12]. Having in mind the limitations of electrodiagnostic procedures, muscle biopsy with typical finding of myosin and type 2 muscle fibers loss would help in setting a definite diagnosis [13].

The pathophysiology of the condition has not yet been clarified. It is characterized by distal axonal degeneration of sensory and motor nerves.9. Muscles and nerves are not isolated from the microcirculatory, cellular, and metabolic disorders that occur during critical illness. Microvascular alteration includes vasodilation, increased permeability, leukocyte extravasation, and hypoxemia. Metabolic alteration includes hyperglycemia, catabolic activation, mitochondrial insufficiency, free radical generation and hypoalbuminemia. Cellular electric alteration implies sodium and calcium channel dysfunction and lost myocyte irritability [14, 15].

Critical illness myopathy is a heterogeneous entity with different pathological subtypes: traditional myopathy after a critical illness is characterized by myosin loss, fulminant acute necrotizing myopathy and cachectic myopathy. Acute necrotizing myopathy is usually characterized by elevated creatinine kinase and myoglobinuria [16]. Cachectic myopathy is characterized by marked hypotrophy similar to inactivity and loss of type 2 muscle fibers [17]. Traditional critical illness myopathy is characterized by myosin loss, derangements of the contractile apparatus and normal or slightly elevated creatinine kinase values [16].

Electrophysiological studies are recommended to be performed when this is possible in order to rule out other causes of neuromuscular weakness, especially in uncooperative and sedated patients. It has been shown that despite a normal rate of nerve conduction, there is an inability of the sarcolemma to transmit signals [17].

Patients with CIPNM have prolonged hospitalization, prolonged weaning from mechanical ventilation, and experience significant chronic complications.18,19 At ICU discharge, all patients with CIPNM are dependent on outpatient care and require inpatient rehabilitation. They are usually transferred to rehabilitation centers where they stay for 4 to 16 weeks [11].

The prognosis for patients with CIPNM ranges from complete recovery within a few months with the disappearance of neurophysiological signs to chronic weakness with signs of

denervation up to 5 years after ICU discharge. The best results are achieved in the first 6 to 12 months. Electrodiagnostic evaluation indicates that recovery can be prolonged up to 24 months, and rehabilitation helps in faster regeneration of the nervous and muscular structures [20–23]. The results of the clinical examination of our patient at the end of the rehabilitation program proved beneficial. Owing to the rehabilitation program, he gained better muscle strength and dexterity in the upper and lower extremities and greater independence in performing daily activities.

In the absence of appropriate specific treatment, it remains unclear to what extent recovery can occur spontaneously.

CONCLUSION

Early recognition of CIPNM is imperative for conducting early rehabilitation. It prevents complications due to immobilization such as deep venous thrombosis, contractures, decubitus ulcers and reduces the length of hospital stay. Early rehabilitation leads to a better motor outcome, improves short-term and long-term functionality and leads to a better quality of life.

Given that this is the only rehabilitated patient and the first reported case in our country so far, it is not possible to compare the results and outcome of rehabilitation. Based on our own experience and review of the literature, we conclude that rehabilitation is necessary and long-lasting in patients with CIPNM.

Further case collection and research addressing this complex pathophysiology of the disorder will shed light on the prevention and effective treatment of the disease.

REFERENCES

1. Holley CT, Harvey L, John R. Left ventricular assist devices as a bridge to cardiac transplantation. J Thorac Dis. 2014 Aug; 6(8): 1110–1119.
2. Linos K, Foot C, Ziegenfuss M, et al. Critical illness weakness: common questions. Curr Anesth Crit Care. 2007; 18: 252–260.
3. Bolton CF, Laverty DA, Brown JD, et al. Critically ill polyneuropathy: electrophysiological studies and differentiation from Guillain-Barré syndrome. J Neurol Neurosurg Psychiatry. 1986; 49: 563–73.
4. van Mook W, Hulsewé-Evers R. Critical illness polyneuropathy. Curr Opin Crit Care. 2002;8:302-310.
5. Bolton C, Young B, Zochodne D. The neurological complications of sepsis. Ann Neurol. 1993; 33: 94–100.
6. National Heart, Lung and Blood Institute (NHLBI) workshop on respiratory muscle fatigue: a report of the respiratory muscle fatigue workshop group. Am Rev Resp Dis. 1990; 142(2): 474–480.
7. Alhan HC, Cakalağaoğlu C, Hanci M, et al. Critical-illness polyneuropathy complicating cardiac operation. Ann Thorac Surg. 1996; 61: 1237–9.
8. Thiele RI, Jakob H, Hund E, et al. Critical illness polyneuropathy: a new iatrogenically induced syndrome after cardiac surgery? Eur J Cardiothorac Surg. 1997; 12: 826–35.
9. Piper SN, Koetter KP, Triem JG, et al. Critical illness polyneuropathy following cardiac surgery. Scand Cardiovasc J. 1998; 32: 309–12.
10. Keaveney AM. Critical illness polyneuropathy in adults after cardiac surgery: a case study. Am J Crit Care. 2004; 13: 421–4.
11. Van der schaaf M , Beelen A., De Groot I.J.M. CLINICAL COMMENTARY Critical illness polyneuropathy: a summary of the literature on rehabilitation outcome. Disability and rehabilitation, 2000 ; vol. 22, no. 17, 808±810
12. Bolton CF. Neuromuscular manifestation of critical illness. Muscle Nerve. 2005; 32(2): 140–163.
13. Di Giovanni S, Mirabella M, D'Amico A, et al. Apoptotic feature accompany acute quadriplegic myopathy. Neurology. 2000; 55(6): 854–858.
14. Hermans G, De Jonghe B, Bruyninckx F, et al. Clinical review: critical illness polyneuropathy and myopathy. Crit Care. 2008; 12(6): 238–244.
15. Zhou C., Wu L., Ni F., et al. Critical illness polyneuropathy and myopathy: a systematic review. Neural Regen Res. 2014; 9(1): 101–110.
16. Waclawik AJ, Sufit RL, Beinlich BR, et al. Acute myopathy with selective degeneration of myosin filaments following status asthmaticus treated with methylprednisolone and vecuronium. Neuromuscul Disord. 1992; 2(1): 19–26.
17. Hund E. Neurological complications of sepsis: critical illness polyneuropathy and myopathy. J Neurol. 2001; 248(11): 929–934.
18. Visser LH. Critical illness polyneuropathy and myopathy clinical features, risk factors and prognosis. Eur J Neurol. 2006;13(11):1203–1212.
19. Guarneri B, Bertolini G, Latronico N. Long-term outcome in patients with critical illness myopathy or neuropathy: the Italian multicentre CRIMYNE study. J Neurol Neurosurg Psychiatry. 2008; 79(7): 838–841.

20. Zifko UA. Long-term outcome of critical illness polyneuropathy. Muscle Nerve Suppl. 2000; 9: S49–S52.
21. Mehrholz J, Phol M, Kugler J, et al. Physical rehabilitation for critical illness myopathy and neuropathy. Cochrane Database Syst Rev. 2015; (3): CD010942.
22. Morris PE, Goad A, Thompson C, et al. Early intensive care unit mobility therapy in the treatment of acute respiratory failure. Crit Care Med. 2008; 36(8): 2238–2243.
23. Fletcher SN, Kennedy DD, Ghosh IR, et al. Persistent neuromuscular and neurophysiologic abnormalities in long-term survivors of prolonged critical illness. Crit Care Med. 2003; 31: 1012–6.

Резиме

МЕНАЏМЕНТ НА ПОЛИНЕВРОМИОПАТИЈА КАЈ КРИТИЧНО БОЛЕН ПАЦИЕНТ ПО ИМПЛАНТАЦИЈА НА УРЕД ЗА МЕХАНИЧКА ЦИРКУЛАТОРНА ПОТПОРА НА ЛЕВАТА КОМОРА

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Полиневромиопатијата кај критично болен пациент по кардиохируршка интервенција е често непрепознаена и ретко пријавувана клиничка состојба. Се карактеризира со повеќе проксимална отколку дистална симетрична флакцидна мускулна слабост од различен степен и тешкотија при одвикнувањето од респиратор. Навремената рехабилитацијата ги спречува раните компликации и ја намалува должината на хоспитализацијата. Рехабилитацијата води кон подобар моторен исход, ја подобрува краткорочната и долгорочната функционалност и води кон подобар квалитет на живот.

Клучни зборови: полиневромиопатија, критично болен пациент, рехабилитација, уред за механичка циркулаторна потпора на левата комора