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Rehabilitation of a patient with critical illness polyneuropathy (CIP) following acute respiratory failure: a case report and review of literature

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Abstract

Critical illness polyneuropathy (CIP), a neurologic complication that may occur secondary to cardio-respiratory distress, surgery, trauma and coma, is associated with sepsis or multiple organ failure. CIP is characterized by an axonal distal degeneration of sensory and motor fibres. The patients will often become neurologically conspicuous when weaning from mechanical ventilation is unexpectedly difficult. There are just a few cases reported with description of the functional outcome and rehabilitation issues of this condition. An additional CIP case of a 62-year old man complicated with anoxic brain damage during the respiratory distress is reported here. He was referred for rehabilitation, made a remarkable recovery (FIM gain 45!) and returned home after 79 days of treatment in the ward. A review of the pertinent literature is provided. Rehabilitation specialists and other professionals working within ICU's should be aware of this condition and be able to recognize and treat CIP at early possible stage.

Introduction

Improving survival rates have prompted clinicians to pay more attention to the complications that can hinder or jeopardize recovery from critical illness. Neuromuscular dysfunction is now well recognized as a common complication of critical illness, which may result in extreme weakness and is responsible for considerable morbidity, as well as being associated with increased mortality.[1]

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Critically ill patients may during intensive care therapy develop pathological conditions within the neuromuscular system resulting in generalized muscle weakness that involves patients' limbs and sometimes becomes complete. This tetraplegic condition may be caused by impairment in nerve conduction, neuromuscular transmission or muscle.

The dominating symptoms are the absence of voluntary contractions, arreflexia and sometimes-reduced sensory function. Thus, the diagnoses are frequently considered first when sedation is stopped and the patients are ready for weaning from mechanical ventilation. Of great importance for the medical care providers, are the facts that these symptoms are to be considered even if central neuronal function may be intact. For correct evaluation and prognostic information to the patient, routine electrophysiological evaluation is required; including electromyography (EMG), electroneurography (ENeG) and in selected cases also a muscle biopsy for detailed hystopathological evaluation.[2]

In the literature minimal attention was given to the rehabilitation aspects and long-term outcome of patients with CIP. The specific course and long-term outcome remains unclear. Research on the course and long-term functional outcome in CIP is necessary in order to identify rehabilitation problems and to formulate treatment strategies specifically directed towards the outcome of this disease.[3]

In reviewing the pertinent literature, just two papers were found describing the functional outcome and rehabilitation issues of six patients with this condition.[4, 5] An additional case of rehabilitation of a CIP patient is reported here.

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Case report

A 62-year-old man was admitted to the Department of Neurological Rehabilitation 'C' of the Loewenstein Hospital Rehabilitation Centre in Raanana, a free-standing 240 bed facility of the Clalit Health Services organization, affiliated to the Sackler School of Medicine of the Tel Aviv University, with asymmetric hypotonic and arreflexic tetra paresis, distal hypoesthesia, muscular atrophy, with tracheotomy, lesion of the left vocal cord and with a big and deep decubitus ulcer in the sacral region.

Per history he suffered from ischemic heart disease, myocardial infarct (6 years ago) congestive heart failure, treated with CAGB (5 years ago) with good outcome. He received Prednisone (10 mg/d) during several months due to rheumatoid arthritis till and during the present disease.

Prior to the present acute episode he was functionally independent and was living with his family and worked until the onset of the disease.

Two months before admission to our ward, he was hospitalized in a general hospital due to high fever and respiratory difficulties. Intubation was performed and the patient was admitted in the Respiratory ICU with diagnoses of right upper lobe pneumonia and acute respiratory failure. A tracheotomy was performed and he was placed under mechanical ventilatory assistance thorough tracheotomy and was given antibiotic therapy during 3 weeks.

The use of high doses of corticosteroids is reported but not of neuromuscular blocking agents during this period of time.

When he awoke, he was not able to move his four limbs and trunk.

CT scan of the brain showed lacunar infarcts in the left frontal region and right basal nucleus without mass effect, which reveals possible hypoxia of the brain during the acute respiratory distress. He was then transferred for rehabilitation to our facilities with diagnosis of bilateral hemiparesis due to anoxic stroke. The tracheotomy was closed immediately after admission.

He began his rehabilitation programme with the following goals:

- treatment of the pressure wound of sacrum;
- treatment of dysphonia;
- recovery of muscle force, trophism and functional skills of the 4 limbs; and
- independency in ADL and mobility.

FIM score at time of hospitalization was 33 with good performances in communicative and cognitive skills. Modified Rankin scale level, 5 (complete dependence).

Nerve conduction velocity (NCV) tests and needle EMG were performed. Motor and sensory nerve conduction studies revealed neurogenic damage with axonal and myelinic impairment in the upper limbs and predominantly axonal in the lower limbs, without signals of myopatic impairment and without denervation activity. Median nerve NCV was 44 m/sec, Ulnar nerve NCV was 47.9 m/sec, Tibial nerve NCV = 33 m/s and common peroneal NCV 40.5 m/sec, all with very low amplitude. Sensory conduction tests showed delayed sensory distal latencies (SDL) in the median (4.4 msec.) and ulnar (3.6 msec) nerves. The Sural nerve was unexcitable (no response elicited).

CPK serum levels were within normal range.

During his hospitalization he participated in an intensive multidisciplinary rehabilitation programme composed by physical occupational and speech therapy, nursing and medical assistance.

He was discharged home 79 days after his hospitalization, the pressure wound improved with good granulation tissue and without need of surgical flapping.

At this time distal hypotrophy, weakness and hypoesthesia, specially in the interosseous muscles persisted in the four limbs, he walked with a four points cane for short distances with fatigue and he was independent for grooming and he needed mild assistance for bathing and dressing. FIM score at discharge 78 (FIM gain = 45); modified Rankin scale level, 3 (partially dependent). He continued an ambulatory rehabilitation treatment in the centre's day hospital after discharge.

Discussion

Complications are common in patients treated for sepsis and multiple organ dysfunctions in critical care units. Failure to wean from the ventilator, due to involvement of the respiratory system, and severe muscular weakness are typical symptoms. Patients with severe infectious (sepsis) could suffer multiple organ dysfunction and what is named systemic inflammatory response syndrome – SIRS.[6]

One third of these patients may develop acute myopathy, and about 13%, acute axonal sensorimotor polyneuropathy.[7,8] This acute polyneuropathy, predominantly is axonal and motor.[9–11]

Acute myopathy occurs in critically ill patients, receiving neuromuscular blocking agents or corticoster-

oids during intensive care hospitalization but may occur in patients with severe systemic illness without exposure to corticosteroids or neuromuscular blocking agents.[21] The mechanisms apparently are at the cellular and molecular levels.[6, 22]

Critical illness polyneuropathy and myopathy, either as separate or combined entities (CRIMYNE) or 'neuromyopathie de reanimation' are common causes of muscular weakness during treatment of critical illness. These disorders are often difficult to distinguish from each other, as the clinical and electrophysiological findings may overlap.[6, 8, 23, 24]

As for the possible pathogenesis, hyperosmolality, parenteral nutrition, non-depolarizing neuromuscular blockers and neurological failure can favour CIP development.[9] In fact, many factors have been claimed to be involved in the pathogenesis of CIP, including malnutrition, hyper alimentation, vitamin depletion and hyperglycemia, but none has been definitely proven so far. However, many authors suggest that the factors mediating the systemic effects of sepsis, i.e. the systemic inflammatory response syndrome (SIRS), are responsible for the axonal degeneration of CIP.[12]

It therefore appears that CIP is just another organ system that fails during sepsis or SIRS. The prevalence of SIRS is high. In surgical ICUs, it can affect more than 80% of patients. Most of these patients, however, do not have infection documented.[13]

A disturbance in the microcirculation of peripheral nerves has also been suggested as disturbed microcirculation is a crucial feature in sepsis.[14] Other clinical and experimental research works support the hypothesis of a multi-factorial aetiopathogenesis of CIP, with possible involvement of a so far unknown, low-molecular-weight neurotoxic agent.[15, 16]

The mortality in unselected patients with multiple organ failure approximates 50% and may reach 98% in those with three or more failed organ systems. If patients survive, those with mild polyneuropathy may recover from neuropathy within weeks and those with more severe forms within months.[17] Unusually severe forms may be associated with incomplete recovery, resulting in persistent motor handicaps.[18] Patients with significant slowing of nerve conduction may have a particularly poor prognosis.[19]

A study addressing the quality of life after CIP showed marked impairment in all patients.[20] These findings are in contrast to the earlier belief that overall outcome of CIP is generally favourable.[10]

In summary, the concept of CIP as a self-limited acute axonal neuropathy developing during ICU treatment, unknown in clinical practice just two decades ago, is now well established. Clinical manifestations include delayed weaning from the respirator, muscle weakness and prolongation of the mobilization phase. Recovery is rapid and mostly complete in patients with mild to moderate neuropathy; survivors of severe sepsis with prolonged stay on the ICU may show slow and incomplete recovery and reduced quality-of-life scores. A thorough differential diagnosis should be made before CIP is diagnosed. Bedside electrophysiological studies will aid early recognition of CIP and monitoring of its course. The systemic inflammatory response evoked by sepsis is regarded to cause axonal injury to peripheral nerves. Deeper insight into these mechanisms is urgently needed to develop pathogenetically based therapies. For the present, treatment of CIP is symptomatic. Stabilization of the underlying critical condition and elimination of sepsis appear to be of major importance for prevention and reversal of ICU neuropathy. Awareness of adverse effect of steroids and pancuronium, the use of passive mobilization, shortening the use of nondepolarizing neuromuscular blockers if necessary and early rehabilitation would minimize disability due to this phenomenon.[22]

Neuromuscular rehabilitation should be instituted as early as possible. The case presented indeed encourages these policies of early referral and active interdisciplinary rehabilitation in appropriate CIP cases. Compared with the reported cases[4, 5] our case was relatively severely impaired but, nevertheless, improvement was remarkable and the patient returned home with some degree of independence in self-care and ambulation that significantly influenced patient's quality of life. Rehabilitation specialists and other professionals working within ICU's should be aware of this condition and be able to recognize and treat CIP at early possible stage.

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