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Acute respiratory failure in the setting of Chronic Inflammatory Demyelinating Polyneuropathy

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Abstract

Chronic inflammatory demyelinating polyneuropathy (CIDP) is symmetric, motor and sensory peripheral neuropathy with a slow, progressive course. This disorder targets the myelin sheaths of peripheral nerves, resulting in progressive weakness, fatigue and sensory neuropathy, though clinical manifestations are varied. The patient is a 61 year old male with a past medical history of coronary artery disease status post coronary artery bypass graft with subsequent percutaneous intervention, diabetes, hypertension and fatigue presenting to the Baptist Hospital of Miami Emergency Department (ED) complaining of shortness of breath and progressive weakness following physical therapy. Three years ago he began to have symptoms of weakness. At this time, he described difficulty raising the right arm above ninety degrees, progressing to involve the left arm. During this visit, he reported decreased grip strength in both arms. He had decreased pinprick sensation in a stocking and glove distribution up to the knees and just above the wrists. The patient had progressive motor sensory neuropathy. He did not exhibit physical exam signs consistent with amyotrophic lateral sclerosis. Similarly, he did not have a history to support Guillain-Barré Syndrome. The findings on exam were more consistent with a chronic inflammatory demyelinating disorder likely associated with his long-standing diabetes. This diagnosis was further supported by elevated protein in the cerebrospinal fluid (CSF) (150 mg). The patient deteriorated rapidly during his hospital stay from a respiratory perspective, becoming increasingly BiPAP dependent. After lumbar puncture findings were returned, he immediately was begun on a regimen of IV steroids and intravenous immunoglobulins (IVIg) to prevent progression of respiratory symptoms. This case was a valuable learning experience, exemplifying the pulmonary complications of neuromuscular disease in the setting of a rare disorder.

Keywords

chronic inflammatory demyelinating polyneuropathy; respiratory failure; neuromuscular respiratory failure

Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is symmetric, motor and sensory peripheral neuropathy with a slow, progressive course that is frequently underdiagnosed [1]. This disorder targets the myelin sheaths of peripheral nerves, resulting in progressive weakness, fatigue and sensory neuropathy [2]. It is hypothesized that CIDP is autoimmune in etiology [3]. Currently, the role of

circulating antibodies and the similarity in the cell-mediated and humoral mechanisms leading to the development of CIDP are areas of research and development [4]. Unfortunately, because of the varied clinical presentation and lack of clear diagnostic criteria, numerous patients with CIDP may be undiagnosed or untreated [3]. It has been suggested that the incidence of CIDP is higher in diabetic populations [1]. The differential diagnosis for CIDP may include diabetic neuropathy, hypothyroidism, and nutritional deficiencies [5]. Based on these diagnoses, work-up frequently includes electrodiagnostic testing, complete blood count, comprehensive metabolic panel, erythrocyte sedimentation rate, fasting blood glucose, vitamin B_{12} levels and thyroid stimulating hormones [5]. CIDP is characterized by electrophysiological abnormalities and elevated CSF protein with response to immunomodulation [1].

Case Presentation

A 61 year old male with a past medical history of coronary artery disease status post coronary artery bypass graft (CABG) with subsequent percutaneous intervention, diabetes, hypertension and fatigue presented to the Baptist Hospital of Miami Emergency Department (ED) complaining of shortness of breath and progressive weakness following physical therapy. The patient was initially admitted for fecal impaction. During his stay he described episodes of breathlessness with increasing frequency. While completing physical therapy in the hospital, he suffered an acute episode necessitating transfer to the ED.

Initial evaluation in the ED was exhaustive for emergent causes of dyspnea. V/Q scan suggested a low probability of pulmonary embolism. There was no evidence of sepsis nor were there physical exam signs of congestive heart failure. Complete blood count (CBC) and basic metabolic panel (BMP) did not return any electrolyte abnormalities that may explain his clinical presentation, though they did reveal an elevated hemoglobin A1c. Chest X-ray did not indicate pneumonia. Arterial blood gas revealed respiratory acidosis. The only remarkable finding was incidental bilateral adrenal adenomas visible on CT. Given his vague presentation and lack of conclusive diagnosis, the pulmonology service was consulted.

During our interview, he was using accessory muscles of respiration and became breathless after each sentence. He was visibly weak. Beginning five years ago, he had numbness in the feet with began in the right foot, progressed to the left, and then involved his right hand. At this time he was diagnosed with carpal tunnel syndrome and had a surgical release on the right hand. Three years ago he began to have symptoms of weakness. He first described difficulty raising the right arm above ninety degrees, progressing to involve the left arm. He reported decreased grip strength in both arms. In addition to these symptoms, he described a history of labile blood pressure and obstructive sleep apnea. Over the "past few months" he had shortness of breath, which worsened after his CABG two months ago. The patient denied alcohol and substance use. He was a former smoker with a 15 pack/year history, but had not smoked in 30 years.

The patient's blood pressure fluctuated from readings consistent with hypertensive urgency (220/100 mmHg) to readings that are below the normal range, as was the case with present reading (98/57 mmHg). He was persistently tachycardic. There were no fasciculation of the tongue. Sensory examination of the extremities revealed decreased pinprick sensation in a stocking and glove

distribution up to the knees and just above the wrists. His history and exam were consistent with progressive motor sensory neuropathy.

His history of bilateral adrenal adenomas suggested an underlying endocrine etiology of these symptoms. Therefore, the patient was evaluated for Cushing's syndrome prior to admission. Further, pheochromocytoma, hyperaldosteronism and hypogonadism were determined unlikely following extensive endocrine evaluation. Physical exam did not reveal signs consistent with amyotrophic lateral sclerosis. Similarly, he did not have a history of recent infection to support Guillain-Barré Syndrome (GBS). His presentation was more consistent with a chronic inflammatory demyelinating disorder likely associated with his long-standing diabetes. The patient's MRI was non-revelatory. This diagnosis is further supported by elevated protein in the CSF (150 mg). However, the CSF findings did not support albumin cytological dissociation, further decreasing the likelihood of acute phase GBS. The patient deteriorated rapidly from a respiratory perspective, becoming increasingly reliant on BiPAP ventilator support. After lumbar puncture findings were returned, he immediately began a regimen of IV steroids and IVIg to prevent progression of his respiratory failure.

Discussion

Due to the number of comorbidities with which this patient presented, an exhaustive workup was conducted before diagnosis of CIDP. Arriving at this conclusion was further complicated by the varied clinical presentations of the condition and lack of true diagnostic criteria for CIDP [2]. In the work-up of this patient, magnetic resonance imaging (MRI) assisted in ruling out other neurological disorders, including multiple sclerosis (MS) and GBS in the evaluation of this patient, though it did not point to a particular etiology. As in the case presentation, the patient received the standard of care of corticosteroids and IVIg with the possible addition of plasmapheresis [6]. Currently, maintenance therapy with IVIg has demonstrated improved quality of life and sustained remission [6].

The pathophysiology of CIDP remains incompletely understood. In rodent models, inoculation of animals with peripheral nerve myelin produces symptoms similar to GBS and in some cases CIDP through induction of immune processes by molecular mimicry mechanisms [7]. Further, the similarity in the pathophysiology of MS and CIPD is leading to clinical studies on the use of therapies applied to multiple sclerosis for CIDP [5].

CIDP is often misperceived as a chronic disease that persists for life. For this reason, patients may receive IVIg though their disease state may no longer necessitate therapy. This is to say that CIDP can wax and wane, and remit with maintenance therapy. Current standard of care for patients with CIDP is corticosteroids, IVIg and plasmapheresis [6]. In the Intravenous CIDP Efficacy (ICE) trial, the largest double-blind, placebo-controlled, crossover study to evaluate CIPD treatment, improvement in disability status that sustained through all twenty-four weeks of the study was observed in 54% of those who received IVIg and only 21% of those who received placebo [8]. These three methods are currently the only therapies that have demonstrated benefits to treat CIDP, though these regimens are often insufficient because 25% of patients respond inadequately [9].

Current trials evaluating alternative tactics of immunosuppression such as cyclosporine, cyclophosphamide or monoclonal antibodies, do not demonstrate significant benefit in CIDP to date [10].

Though axonal degeneration is likely irreversible, patients can remain in stable conditions without further progression of these disease process [10]. It is hypothesized that nearly 40% of patients who receive IVIg for CIDP may safely discontinue therapy and maintain remission [8].

This patient provided an excellent example of the pulmonary complications associated with CIDP, a relatively rare disorder. His exhaustive work-up and treatment design required intensive research and planning. Ultimately, his regimen is ideal for him and his care team is confident in his ability to maintain remission.

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