CASE REPORT

Use of Noninvasive Ventilation with Volume-Assured Pressure Support in Neuralgic Amyotrophy with Bilateral Diaphragmatic Paralysis

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Abstract: Neuralgic Amyotrophy (NA) is a rare, acute onset inflammatory brachial plexopathy that frequently presents with acute pain followed by shoulder girdle muscle weakness. Phrenic nerve involvement affecting the diaphragms occurs in 7-10% of cases. We present the case of a 52-year-old man with neuralgic amyotrophy with phrenic nerve involvement and bilateral diaphragmatic paralysis with marked respiratory symptoms and sleep hypoventilation, who was treated with non-invasive ventilation with volume assured pressure support mode. By 21 months post disease onset, the patient had experienced marked improvement in orthopnea, sleep quality and functional status. This is the first reported case of the use of this mode of noninvasive ventilation in neuralgic amyotrophy.

Keywords: Neuralgic amyotrophy, Phrenic nerve dysfunction, Diaphragmatic paralysis, Noninvasive ventilation, Sleep hypoventilation, EMG.

1. INTRODUCTION

Neuralgic Amyotrophy (NA) is a rare, acute onset inflammatory brachial plexopathy that frequently presents with acute pain followed by shoulder girdle muscle weakness. Hours to days after the onset of pain, paresis develops, and it persists after the pain goes away, usually affecting the long thoracic, suprascapular, and anterior interosseous nerves. NA can affect other peripheral nerves less often, and phrenic nerve involvement affecting the diaphragms occurs in 7-10% of cases [1 - 3].

NA may affect up to 1 per 1000 people per year and is more common in men than in women. It affects all ages, with median onset approximately 40 years of age. The exact pathophysiology is not known; an interplay between environmental factors, mechanical factors and individual susceptibility is assumed to be the cause. A typical history and exam are enough to support a clinical diagnosis. Blood tests, cerebrospinal fluid and serologies are generally unremarkable. Electromyography (EMG) may support the diagnosis and help in the differential, but there is frequently sampling error, and nerve conduction studies may fail to show abnormalities even in clinically affected nerves in NA. Mild abnormalities may be seen in some patients on magnetic resonance imaging of the cervical spine and brachial plexus. No clear treatment options are present, and therapy is mostly supportive. Although most patients recover from the most severe symptoms, persistent pain, fatigue and impairments of activities of daily living remain in a large proportion of patients with NA [1].

We present the case of a 52-year-old man with NA with phrenic nerve involvement and bilateral diaphragmatic paralysis, who was treated with Noninvasive Ventilation (NIV) with Volume Assured Pressure Support (VAPS) and experienced marked improvement in orthopnea, sleep quality and functional status.

2. CASE REPORT

A 52-year-old man woke up with intense right neck and bilateral shoulder pain followed by dyspnea with minimal exertion and bending, and severe orthopnea. There was no weakness, numbness or prodromal symptoms; the pain resolved after one week. There were no sick contacts or environmental exposures. Past medical history included hypertension, hyperlipidemia, pre-diabetes, gastroesophageal reflux disease and untreated moderate obstructive sleep apnea. Medications: amlodipine, carvedilol, metformin, losartan-hydrochlorothizide and pantoprazole. Family history included a father who had a history of diabetes type 2, myocardial infarction and hypertension.

Forced Vital Capacity (FVC) was 2.5 L (53%). Cervical spine MRI showed multi-level degenerative changes with foraminal narrowing without cord compression. Hemidiaphragms were elevated on chest computed tomography.
The patient came for evaluation to our center nine months after onset. There was a partial symptomatic improvement, but a 45-degree angle was required for sleep, with multiple awakenings, nonrestorative sleep, daily naps and persistent fatigue. Height was 1.78 m, weight was 97.1 kg, and body mass index was 30.7 kg/m². Mild respiratory accessory muscle use and decreased diaphragmatic expansion were noted. Lying supine provoked immediate dyspnea, paradoxical abdominal motion and tachypnea.

MRI of the cervical spine showed multi-level degenerative changes with foraminal narrowing, but no cord compression. Nerve conduction studies were performed 10 months following the onset of the symptoms. Motor and sensory nerve conduction studies of the right median and ulnar nerves were normal. Sensory nerve conduction studies of the right superficial radial nerve were normal. The right lateral antebrachial cutaneous nerve sensory amplitude was moderately decreased, and the sensory conduction velocity was normal. Electromyography of the right upper extremity and cervical spine showed large amplitude motor units in the right deltoid, biceps, pronator teres, extensor digitorum communis and first dorsal interosseous but with sparing of the abductor pollicis brevis and paraspinal muscles. Reduced recruitment was seen in the affected muscles. Follow-up study of the phrenic nerves 2 weeks later showed an absent response on the left and reduced amplitude on the right. Electromyography of the diaphragm showed chronic denervation but no acute denervation. Findings confirmed the diagnosis of NA with bilateral phrenic nerve dysfunction.

FVC was 2.16 L (46%), Maximal Inspiratory Pressure (MIP) 41 cmH2O (36%) and Maximal Expiratory Pressure (MEP) 77 cmH2O (36%). Arterial blood gas: pH 7.46, PaCO2 40 mmHg and PaO2 72 mmHg. Nocturnal NIV with volume-Assured Pressure Support (NIV VAPS), Phillips Respironics, (Murrayville, Pennsylvania, USA); target tidal volume (VT) 550 ml (8 ml/kg), Inspiratory Positive Airway Pressure (iPAP): 13-22 cmH2O, Expiratory Positive Airway Pressure (EPAP): 7 cmH2O, respiratory rate: 13/min, inspiratory time: 1.1 sec, rise time: 2.

The patient experienced immediate improvement with NIV; sleep was restful and uninterrupted, with less fatigue and no naps. At 11, 15, 21 and 28 months post onset there was slow continued overall improvement during each visit. At 28 months post onset the patient had significant improvement in his ability to breathe and activity tolerance and FVC had improved to 3.31 L (71%). He was able to walk up to two miles and was able to exercise slightly. There was still dyspnea with more than usual exertion, but not with light to moderate regular activity. He could now bend over and tie his shoes, although this was still not fully comfortable. No orthopnea was present with NIV; he was able to lie in bed for a few minutes without NIV but had not tried sleeping without it. Data downloaded 19 months after NIV showed 100% daily use for 9.1 h and apnea-hypopnea index 2.0 events/h.

3. DISCUSSION

In this patient with NA with phrenic nerve involvement and bilateral diaphragmatic paralysis, NIV VAPS led to marked improvement in orthopnea, sleep quality, fatigue and overall wellbeing. There was moderate recovery in exercise capacity with partial improvement in pulmonary function.

NA is a rare, acute onset inflammatory brachial plexopathy. Patients typically wake up with severe pain in the shoulders, upper arms or neck. Weakness, sensory loss and atrophy of the shoulder girdle muscles develop while the pain fades. Phrenic nerve involvement with diaphragmatic paralasis/paralysis occurs in 7-10% of cases, with male to female ratio 3:1, causing dyspnea, orthopnea, sleep disturbances and fatigue [1 - 3]. Diaphragmatic dysfunction often goes undiagnosed for months [3]. Recovery of pulmonary function is varied. In a study of 16 subjects (15 men) with mean follow-up 4 years [2], 31% recovered to a normal FVC, 38% recovered partially and 31% did not recover. When FVC improved, it was rapid initially followed by slowing and plateau. By modeling, it was calculated that recovery mid-point was 22 months and full recovery may take > 3 years. In another series of 79 patients with 1.6 years median follow-up, 24% had no recovery, 44% had some recovery and 32% reported good recovery. In a subset of 36 patients with 5.2 years median follow-up, 28% still used NIV, 83% had exertional dyspnea, 69% had orthopnea and 31% had sleep disturbances [3]. The current patient experienced partial recovery, similar to the 38% and the 44% of patients in the above-mentioned series. No predictors of recovery, neither complete nor partial, have been determined [2, 3].

The presentation of diaphragmatic dysfunction varies from asymptomatic to dyspnea to respiratory failure [4]. Bilateral diaphragmatic paralysis of any cause is symptomatic and can be challenging to diagnose. FVC is around 45% and MIP is greater than-60 cmH2O. FVC may decrease > 40% further while supine, leading to intense orthopnea [5], and is the most sensitive test to diagnose diaphragmatic dysfunction [3, 4]. While fluoroscopy is 90% sensitive for hemidiaphragm paralysis, with bilateral paralysis upward movements of the ribs by the accessory respiratory muscles often gives the false impression of the diaphragm moving downward with respiratory effort [5].

Treatment of diaphragm dysfunction due to phrenic neuropathy primarily consists of supportive measures such as sleep in an elevated position, but NIV is indicated with sleep

![Figure 1](https://example.com/figure1.png)
hypoventilation, orthopnea, sleep deprivation, or respiratory failure. NIV is well tolerated and has a positive effect in most patients with NA and diaphragm dysfunction [3].

To our knowledge, this is the first report of NIV with average VAPS in NA. In this mode, a target VT, fixed EPAP and backup respiratory rate are set, along with IPAP that self-adjusts within a prescribed range to maintain the target VT despite physiologic changes in ventilatory control, respiratory mechanics and respiratory muscle recruitment that occur with different sleep stages and changes in the position [6].

Since NA patients who improve can sometimes discontinue NIV [7], NIV VAPS might offer the advantage of automatically decreasing IPAP during recovery. While patients with amyotrophic lateral sclerosis on traditional NIV bilevel PAP often require serial IPAP increases for declining respiratory muscle strength [8], NIV VAPS can maintain stable minute ventilation in these patients with few adjustments despite marked disease progression [9].

Patients with NA should be screened for diaphragmatic dysfunction [3], and when present, a low threshold for starting NIV is important given the potential benefits of therapy [2, 3]. Studies are needed to determine if different NIV modes offer a distinct benefit in the management of this disease.

**AUTHORS’ CONTRIBUTIONS**

MDA: conceptualized and drafted the initial manuscript and contributed to acquisition and interpretation of data. NP and LZ: contributed to acquisition and interpretation of data. NT: contributed to conception and design and contributed to acquisition and interpretation of data. All authors critically reviewed and revised the manuscript and approved the final version.

**ETHICS APPROVAL AND CONSENT TO PARTICIPATE**

Not applicable.

**HUMAN AND ANIMAL RIGHTS**

Not applicable.

**CONSENT FOR PUBLICATION**

Informed consent was taken from all the patients when they were enrolled.

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**CONFLICT OF INTEREST**

The authors declare no conflict of interest, financial or otherwise.

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Declared none.

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