



Case report

Continuous Positive Airway Pressure (CPAP) for prevention of recurrent pneumonia in the Neuromyelitis Optica patient



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A B S T R A C T

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Introduction: Traumatic spinal cord injury patients with quadriplegia associated respiratory compromise are at an immediately increased risk of developing pneumonia, but the onset of pneumonia risk and use of prevention strategies in the patient with quadriplegia due to Neuromyelitis Optica has not been described.

Case report: This is a case of a Neuromyelitis Optica patient with quadriplegia, dysphagia and tracheostomy that suffered recurrent fevers due to respiratory infections. The non-specific presentation and test results led to extensive testing, while the frequent recurrence resulted in the patient residing in the acute care hospital 201 days and outside of this hospital only 118 days during the period of August 2011 to June 2012. The initiation of CPAP 10 cm while sleeping overnight for 8–10 h eliminated the recurrence of respiratory infections and thereby reduced both the frequency and duration of the patient's hospital stays.

Conclusions: Patients with Neuromyelitis Optica differ from those with traumatic spinal cord injury as they have a chronic progressive systemic illness that causes continued deterioration of their nervous system resulting in the need for routine monitoring that ensures the timely addition of CPAP for the prevention of pneumonia and its associated medical expenses.

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Case report

A 42-year-old female with quadriplegia for eight years as a result of cervical transverse myelitis due to Neuromyelitis Optica had complete paralysis of her lower extremities, no useful movement in her left upper extremity and minimal movement of her right upper extremity that enabled her to control a television remote and perform other limited functions. She had a chronic tracheostomy due to her difficulty clearing secretions and reduced respiratory muscle strength, was blind in her left eye due to the Neuromyelitis Optica, and had a suprapubic catheter for bladder dysfunction. She was on hormone replacement for pan-hypopituitarism that was due to the Neuromyelitis Optica destruction of the hypothalamus. The presence of a deep venous thrombosis resulted in chronic Coumadin therapy. She had mild dysphagia with thin liquids and a gastric feeding tube placed during one of her hospitalizations for respiratory failure, but ate routinely and was primarily using the tube for difficult to swallow

items such as medications. Similar to most quadriplegia patients, she suffered numerous infections including urinary tract infections, infected decubitus ulcers, and via direct extension from the decubitus ulcers, she required treatment for ischial osteomyelitis.

The patient was a permanent nursing home resident and completely dependent for her activities of daily living. She received all of her hospital care at our institution, and presented in [Table 1](#) are the patient's hospitalizations during the years 2010–2012. Prior to 2011, she neither had respiratory infections nor challenges in the diagnosis and treatment of infections. During the period of August 2011 to June 2012, the patient resided 201 days in the hospital and only 118 days outside of the hospital. Most notable of these hospitalizations was that she typically had a fever and acute respiratory symptoms. However, defining the cause of her fever was challenging due to a chronically abnormal chest X-ray with hypoventilation, a right hemi-diaphragm elevation and lower lobe changes that could represent atelectasis or infiltrates ([Fig. 1](#)). During these admissions the patient was treated with antibiotics and defervesced. However, shortly after discontinuation of antibiotics the patient had resumption of fever. This frequently occurred within a few days, led to prolonged hospitalizations, and resulted in the frequent admissions listed in [Table 1](#). Antibiotics were expanded and adjusted to cover the antibiotic resistant organisms

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Table 1
Hospital visits.

Year	Month	DSLH (Days)	DSLPH (Days)	LOS (Days)	Active problem	Fever	Resp Sxs	New infiltrate
2010	February	416	n/p	4	UTI, Vesico-vaginal fistula	No	No	n/a
	February	10	n/p	3	Exacerbation Neuromyelitis Optica	No	No	n/a
	May	90	n/p	2	Tracheostomy surgery	No	No	n/a
	August	82	n/p	7	Hypotension/panhypopituitarism/UTI	No	No	Possible
	September	32	n/p	8	Hypotension/panhypopituitarism/UTI	No	No	Possible
2011	August	300	n/p	11	Pneumonia/UTI	Yes	Yes	Possible
	Aug–Sept	10	10	21	Pneumonia	Yes	Yes	Possible
	Sept–Nov	14	14	54	Pneumonia	Yes	Yes	Possible
2012	January	54	54	16	Pneumonia/UTI	No	Yes	Yes
	February	8	8	20	UTI/possible pneumonia	Yes	Yes	Possible
	March	26	26	12	UTI/possible pneumonia/possible PEG site cellulitis	Yes	Yes	Possible
	April	4	4	22	Pneumonia/UTI	Yes	Yes	Yes
	April–June	2	2	45	Pneumonia/UTI	Yes	Yes	Possible
	Sept–Oct	103	103	7	Pneumonia	Yes	Yes	Possible

Notes: DSLH = Days Since Last Hospitalization. DSLPH = Days Since Last Pneumonia Hospitalization – only reported if current hospitalization is for pneumonia. LOS = Length of Stay. Resp Sxs = Respiratory Symptoms = increased sputum/respiratory secretions, hypoxia, respiratory failure. Active Problem = primary problem that was the cause for admission and addressed during the hospital stay. New infiltrate = new infiltrate seen on Chest X-ray or chest CT scan. n/p = no prior episode of pneumonia. n/a = data not available because test not performed.

identified in various cultures. Despite adjustments and lengthened antibiotic treatment durations, the patient's fever returned shortly after discontinuation of each antibiotic course. Alternative sources of fever were pursued with tests and actions that frequently failed to identify a fever source.

Based on the reliable defervescence with antibiotic treatment, the clinicians believed the patient was suffering from either an infection that was being inadequately treated, or one that had a frequent relapse due to her physiology. After an initial approach of testing for alternative sources of fever along with prolonged and adjusted antibiotic coverage, the clinicians believed that recurrent infection was likely, and the patient's hypoventilation, dysphagia and inadequate cough increased the risk of recurrent pneumonia. As a strategy to improve her lung physiology, nocturnal Continuous Positive Airway Pressure (CPAP) of 10 cm was initiated in the beginning of June 2012. Following the initiation of this treatment, the patient did not have recurrent fevers when the antibiotics were discontinued. There were no recurrent episodes of pneumonia while using CPAP, and the recurrence of fever and respiratory symptoms consistent with pneumonia occurred within 12 days of the patient refusing to use CPAP at the nursing home during late September 2012. Three months following this single episode of non-compliance, our patient ultimately passed away after

developing a seizure disorder but prior to any further episodes of pneumonia or fever.

Discussion

Our patient had frequent and prolonged hospital stays due to pneumonia, a pattern that was immediately changed upon the initiation of CPAP 10 cm while the patient was sleeping at night for a period of 8–10 h. Prior to initiating nocturnal CPAP, days between episodes of pneumonia ranged from 2 to 54 days, while this was extended to 103 days with the use nocturnal CPAP. The occurrence of pneumonia at 103 days was during a period of non-compliance with the CPAP. Because our patient did not previously suffer frequent pulmonary infections, the diagnosis was delayed. While traumatic spinal cord injury patients have a pulmonary function and associated infection risk that remains stable for many years from the time of their injury, Neuromyelitis Optica patients have progressive disease and care needs that escalate over a period of years.

Patients with spinal cord injury suffer recurrent infections that are most commonly urinary tract and soft tissue in origin [1]. While occurring in paraplegia, pulmonary infections are more common in tetraplegia patients during a 1, 2 and 5-year follow-up period [2,3]. Among newly acquired upper cervical spine injury patients, 17% have been found to develop pulmonary infections, among which there is a 31% mortality rate [4]. Respiratory infections are among the three most common causes of rehospitalization within one year of spinal cord injury [5]. As well, longitudinal studies have identified an association of diseases of the respiratory tract as the cause of rehospitalization in tetraplegia during 20 years following acute traumatic spinal cord injury [6]. The occurrence of pneumonia in the acute severe traumatic spinal cord injury patient has been associated with worse neurologic outcome one year after injury and consideration of the use of preventative measures has been suggested [7]. Of similar concern is the identified rise in the rate of death due to pneumonia after the first anniversary following a spinal cord injury [8]. While subject reviews supporting the use of Noninvasive ventilation for the management of the high level spinal cord injury patient have been published, herein is the first detailed case report describing the diagnostic and management challenges that may lead to delays in the institution of this technology and its use in a patient with Neuromyelitis Optica [9,10].

Dysphagia has been found in 30% of acute cervical spinal cord injury patients and these individuals are found to have higher

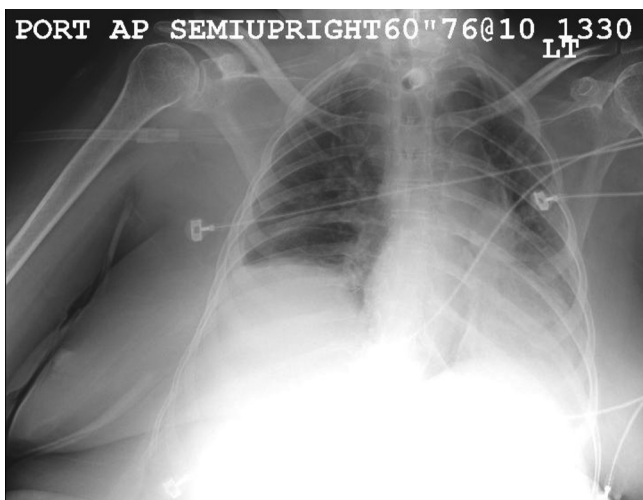


Fig. 1. Typical Patient Chest X-ray.

occurrences of pneumonia and longer length of hospital stays compared with patient without dysphagia [11]. Dysphagia is found at higher rates in patients with tracheostomies and associated with an inability to handle oral secretions, particularly at times of rest. Exemplified by the need for a tracheostomy and a feeding tube, our patient had significant dysphagia that placed her at high risk for poor self-management of oral secretions. With the premise that the CPAP reduced chronic aspiration of saliva, the use of CPAP to reduce the frequency of hospitalization due to respiratory disease was found to be successful in children with tracheostomy for treatment of airway obstruction [12]. It may indeed be that our patient benefited from nocturnal CPAP administered via her tracheostomy due to its prevention of nocturnal oral secretion aspiration.

The increased frequency of lower respiratory tract infections in the tetraplegia patient relative to the paraplegia patient is likely indicative of its dependency on the degree of respiratory failure and the associated inability to ventilate, avoid atelectasis and clear secretions. Post-operative abdominal surgery patients suffer from hypoventilation and studies have demonstrated the ability of CPAP to prevent atelectasis and pneumonia in this population [13]. As another potential mechanism of action, the CPAP may have reduced the occurrence pulmonary infections in our patient by its demonstrated ability to reduce the degree of atelectasis or its assistance in the mobilization of secretions in patients prone to hypoventilation.

While we do not hesitate to implement the long-term use of nocturnal CPAP in patients suffering a common ailment such as sleep apnea, the initiation of CPAP of any duration in a patient with a chronically progressive and ultimately terminal disease such as Neuromyelitis Optica must be balanced with the patient's condition and desire to utilize the treatment. Once it appeared that CPAP was successful at eliminating the nearly continuous hospitalizations and use of antibiotics, our patient was required to choose between life-long nocturnal CPAP and hospice care. Other than the one episode of a 12-day break from the CPAP, our patient found nocturnal CPAP a desirable alternative to accepting end-of-life focused care. However, when her disease progressed to cause frequent and difficult to control seizures, the patient's family members choose to limit her care to end-of-life comfort measures.

Conclusions

Neuromyelitis Optica patients may develop chronic hypoventilation and dysphagia due to injury to the cervical spinal cord and other affected areas of the nervous system. When these patients develop recurrent fevers, clinicians may not immediately consider that regardless of their prior status the patient may now be at risk for recurrent respiratory infections. In contrary to traumatic spinal cord injury patients, Neuromyelitis Optica patients may develop an increased susceptibility to respiratory infections at a delayed time due to the chronic progressive course of their disease. Our case report suggests that monitoring these patients for signs of progression and early use of nocturnal CPAP to reduce the frequency of these infections may reduce the patient morbidity and expense associated with frequent and prolonged hospital stays.

Learning points

1. CPAP can be used to prevent pulmonary infections for Neuromyelitis Optica patients.
2. Neuromyelitis Optica is different from other causes of spinal cord injury in that it is a chronic progressive disease that leads to a continuously increasing state of disability and risk for new complications that previously did not exist for the patient.
3. Clinicians should monitor Neuromyelitis Optica patients for the development of dysphagia, hypoventilation, atelectasis, inadequate cough and inability to manage oral secretions as an indication of an increased pulmonary infection risk.
4. Early implementation of preventative therapies such CPAP can reduce patient morbidity and healthcare expenditures.
5. Compared with paraplegia patients, pneumonia is a more common source of fever and infection in the quadriplegia patient, is a common cause of hospitalization and is an indicator of a higher mortality rate.

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