THE EUROPEAN SOCIETY OF MEDICINE
Medical Research Archives, Volume 12 Issue 8

CASE REPORT

Long-term mechanical ventilation in a myasthenic patient with suspected obstructive sleep apnea

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OPEN ACCESS

PUBLISHED
31 August 2024

CITATION
Bahaddi, A., Tougar, S., et al., 2024. Long-term mechanical ventilation in a myasthenic patient with suspected obstructive sleep apnea. Medical Research Archives, [online] 12(8).
https://doi.org/10.18103/mra.v12i8. 5712

DOI
https://doi.org/10.18103/mra.v12i8. 5712

ISSN
2375-1924

ABSTRACT

Myasthenic crisis is a medical emergency with a very poor prognosis if not treated early and appropriately. In the absence of mechanical ventilation, the respiratory impairment is the most serious aspect of the condition, leading the intensive care anaesthetist to resort to intubation. The association of this disease with sleep apnea syndrome is frequent but little-known, and can make it difficult to disconnect the patient from the ventilator. Making the diagnosis of sleep apnea syndrome is difficult because of polysomnography, which is laborious to set up in intensive care, but can be facilitated by the use of a simple questionnaire when there is a strong clinical suspicion. Weaning a patient suffering from neuromuscular disease, whether or not associated with sleep apnea syndrome from mechanical ventilation can be a complex task. When extubation proves impossible, tracheostomy remains the most desirable alternative. The introduction of long-term mechanical ventilation at home is an interesting solution for shortening the long hospital stay of patients in intensive care units, and significantly reduces the risk of infection, morbidity and mortality.

Introduction:

Myasthenia is a disease of autoimmune origin, caused by dysfunction of neuromuscular transmission, and clinically manifested by muscle weakness accentuated by exertion. It is a chronic pathology, the evolutionary risk of which is the onset of myasthenic relapses and crises that can lead to the patient being admitted to intensive care. Obstructive sleep apnea (OSA) is defined by significant interruptions (apneas) or reductions in ventilation (hypopneas) during sleep. The prevalence of sleep apnea is estimated at 36% in myasthenic patients, compared with 15-20% in the general population[1-2]. When it comes to making a diagnosis of sleep apnea in intensive care, polysomnography, despite being the reference test, remains difficult to perform in an intensive care unit, and the use of a questionnaire may be an alternative for making the diagnosis in cases of strong clinical suspicion. Besides knowing how to make the diagnosis, this sleep disturbance, which is added to the underlying pathology, will have an impact on the duration of ventilation and weaning, and therefore on the length of the stay in intensive care, which could be wrongly blamed on several other more frequent aetiologies. Long-term mechanical ventilation (or mechanical ventilation at home) can be proposed to accelerate patient discharge and reduce the risk of infection associated with a stay in intensive care. In order to shed light on the weaning difficulties that anaesthetists may encounter in such situations, we report the clinical case of a myasthenic patient admitted to the resuscitation medical service of the Ibn Rochd University Hospital in Casablanca, and who was diagnosed with sleep apnea only at a very late stage, using a method that is uncommon and unknown in the literature, enabling him to return home after a very long stay in intensive care.

Case Report:

A 38-year-old man, 95 kg (BMI 30), with a history of diabetes and oral antidiabetic medication discontinued 1 year previously, presented to the neurology department with a 9-month history of diplopia, ptosis of the right eye and fatigue on exertion, complicated by swallowing and phonation difficulties. On admission, neurological examination revealed a conscious patient, symmetrical reactive pupils, blood glucose 2.48, apyretic. He was polypneic, with SpO2 at 60% in the open air. Pleuropulmonary auscultation revealed bilateral snoring. Hemodynamically, he was tachycardic at 130 bpm with a BP of 15/8. Physical and general examination revealed no particularities apart from moderate android obesity. The evolution was marked by a rapid worsening of his respiratory distress (sudden desaturation to 30%, appearance of signs of respiratory struggles and respiratory pauses) with signs of hypercapnia and consciousness disorders. The patient was rapidly intubated and placed on mechanical ventilation (volume-controlled mode, FiO2 100%, PEEP 5, Vt 450 mL). Saturation rose to 98%. Echocardiography showed good overall cardiac contractility, preserved ejection fraction, undilated right ventricle and compliant IVC at 17. Emergency ECG revealed no particular abnormalities apart from sinus tachycardia. Gasometry showed pH 7.33, Po2 59, PCo2 47, HCO3- 19, ratio 147 and lactatemia 4.2. Chest imaging was normal. No biological abnormalities were noted. Lumbar puncture yielded a clear fluid, proteinorachy 0.28, glycorachy 1.46 (ratio 1.2), WBC < 3 elements, direct examination negative. Therapeutically, the patient received a 5-day course of polyvalent immunoglobulin IV at a dose of 0.4g/Kg/d, followed by Pyridostigmine (Mestinon*) at a dose of 3cp/d, with gradual increase in dosage until optimal dose was reached, adapted to the patient and signs of overdosage. An early tracheotomy was proposed and performed at Day 5. His hospitalization in the intensive care unit was marked by the occurrence of two documented septic shocks due to Serratia marcescens and Klebsiella pneumoniae, which were controlled with urgent and then directed antibiotic therapy, while respecting the absolute contraindications to his neuromuscular pathology. Thromboembolic disorders were prevented by repeated echodoppler examinations of the lower limbs, preventive anticoagulation, mechanical compression stockings, motor and respiratory physiotherapy and nursings several times a day. Psychological and psychiatric follow-up was initiated from the 2nd month of hospitalization. Ventilatory weaning was long and difficult. During the day, ventilatory assistance was manually reduced (pressure-controlled mode with AI), with intermittent disconnection windows, more laborious at the end of the day, and impossible to disconnect at night. Respiratory and cardiovascular causes were ruled out by clinical examinations, normal gas levels, laboratory tests, X-rays and ultrasound scans.

Neuromuscular causes were the most likely in view of the context, but diaphragmatic paralysis and resuscitation neuromyopathy were ruled out by an electromyogram that came back negative. A non-organic cause was ruled out following a psychiatric interview, which returned normal. The patient was totally unplugged during the day from the 7th month onwards, but still needed to be plugged in at night, with sleep disturbances such as insomnia on falling asleep. Following psychiatric advice, a low-dose hydroxyzine antihistamine (Taraxet*) was prescribed, and close monitoring of the patient during the night after disconnection noted the existence of several periods of apnea > 10s with respiratory pauses observed by the doctors on duty.

Excessive daytime somnolence, morning headaches and difficulty concentrating were often reported by the patient, but were often attributed to his asthenia caused by his long stay in the intensive care unit. In the absence of polysomnography in the intensive care setting, the diagnosis of OSA was made on the basis of criteria A and B and a low score on the Richards-Campbell sleep questionnaire. Weight loss (over 30 kg in 6 months) made the presumption of diagnosis difficult. In consultation with the pulmonologists, home mechanical ventilation was proposed using a mobile device with an interface adapted to his tracheostomy cannula. Continuous positive airway pressure with a low level of support was the optimal mode. After a period of close monitoring and adaptation to the new device, the patient was transferred to the neurology ward. His total length of stay in intensive care was 9 months. Suspension laryngoscopy performed cold came back without any notable anomaly. Final decanulation took place a week later, with home ventilation now via a bucconaral mask.

Discussion:

Management of a myasthenic crisis is primarily symptomatic. Swallowing disorders, which put the VAS at risk and prevent patients from taking their medication, mean that a nasogastric tube must first be systematically inserted. In addition to the usual vitals and state of consciousness, daily monitoring in the ICU includes reinforced monitoring of muscle deficit and, above all, respiratory impairment. Acute respiratory failure[3] is described as hypoxemia below 60 mmHg and/or hypercapnia above 45 mmHg. This biological definition is unsuitable for acutely evolving neuromuscular diseases such as Guillain-Barré or myasthenic crisis, since they are considered to be respiratory insufficiencies with normal gas exchange. It has also been shown that gasometric parameters are the last to deteriorate before the onset of acute respiratory distress and endotracheal intubation[4]. This explains why gasometry monitoring is a poor tool in this type of pathology. Similarly, care must be taken when oxygen therapy is required, as the risk is to mask alveolar hypoventilation, or even aggravate it by suppressing the hypoxic stimulus. The challenge is therefore to identify subjects at risk of respiratory decompensation who should first be monitored in the intensive care unit, and then to determine which of these will require invasive ventilation. The indication for mechanical ventilation is based on clinical criteria (polypnoea, orthopnoea, difficulty or inability to speak, ineffective cough, severe congestion, rapid desaturation, signs of hypercapnia, etc.). There are no specific recommendations as to the type of ventilation to use. The use of non-invasive ventilation is not recommended in this type of pathology, and cannot currently be recommended for these patients, due to the risk of exhaustion, swallowing disorders and ineffective coughing, which can hamper the prescription of NIV.

Once the patient's respiratory status has been stabilized, the treatment of a myasthenic crisis should systematically include a search for a triggering factor. In 30 to 40% of cases, this is an infection (viral, bacterial or fungal), most often of pulmonary origin[5]. The second cause is a drug interaction, since many drugs can aggravate myasthenia through their action on the neuromuscular junction. Specific treatment of relapses is based on plasma exchange (PE) and intravenous immunoglobulin (IVIG). Both treatments have been shown to be effective, with net improvement in 70-75% of cases after the second or third exchange for PE, and in 76% of cases with treatment at a dose of 0.4 g/kg per day for five days for IVIG[6-7]. The choice of treatment depends essentially on the availability of these therapies, and compliance with their respective contraindications (absence of sufficient vascular access and evolving infection for PE, renal insufficiency and theoretical risk of pathogen transmission for IVIG). The response to specific therapies varies from one subject to another, and the expected improvement can sometimes take several weeks or months. Nursing care therefore plays a central role in management, and requires a nursing team trained to deal with this type of patient[8].

Studies[1] suggest that the prevalence of obstructive sleep apnea (OSA) in the general population is 15-20%. According to a cross-sectional study[2], the prevalence is higher among myasthenic patients, at 36%. The association is therefore frequent and should be evoked in the presence of sleep disorders in a patient suffering from myasthenia. The reasons for the low rate of screening for sleep disorders in patients admitted to the intensive care unit are partly due to a lack of awareness of these pathologies among anaesthetists, but difficult access to tests may also play a role. Various study methods and recording techniques can be used to analyze sleep. Sleep in intensive care can be analyzed by self- or hetero-assessment[9], actimetry[10] or bispectral index[11], but the gold standard remains polysomnography[12]. Nevertheless, its use in ICU patients is complex, and its analysis remains to be perfected. Indeed, systematic sleep assessment by polysomnography (PSG) is costly and not routinely feasible due to the need for equipment, technicians and expert interpretation. The alternative is self-assessment using a questionnaire, which has been shown to correlate well with polysomnography results. To date, there is no screening questionnaire specifically developed for sleep assessment in myasthenic patients, but the Richards-Campbell Sleep Questionnaire (RCSQ) is a simple five-item visual analog scale, validated for measuring sleep quality in cooperative ICU patients[13]. Given the difficulty of accessing instrumental sleep assessment in such a clinical setting, the RCSQ has been proposed as a consistent alternative for sleep assessment in intensive care[14]. A recent systematic review and meta-analysis evaluating sleep assessment tools in critically ill patients revealed a good correlation between the results of the RCSQ and equivalent PSG sleep variables, establishing the questionnaire as a reliable tool for assessing sleep disturbance in intensive care[15]. Therefore, subjective survey instruments are needed as practical tools for rapid screening of sleep disturbance in the ICU setting, without replacing PSG, which remains the gold standard.

Failed weaning usually leads to discussion of tracheostomy, taking into account the risk/benefit ratio of prolonged tracheal intubation, the degree of comfort of the intubated patient, the presence of absolute or relative contraindications to tracheostomy, and the wishes of the patient or those close to him/her, particularly when the prospect of long-term ventilatory assistance is in prospect. However, in myasthenic patients expected to require ventilatory assistance for more than 14 days, early tracheostomy is proposed from the outset. The latter is well received by patients. A recent ten-year study showed that 90% of patients and 80% of their carers would again be in favour of a tracheostomy[16]. Once a patient has had a tracheotomy, a home ventilator should be used to prepare the patient and his or her family for a return home as quickly as possible. There are three levels of home ventilator, and the choice of ventilator is made taking into account the patient's clinical condition, and in particular the patient's level of dependence on the ventilator:

- Level 1: ventilators without battery for patients ventilating only at night and less than 8 h/24
- Level 2: ventilators with internal battery for patients who ventilate more than at night and between 8 a.m. and 4 p.m./24 a.m.
- Level 3 or "life-support ventilators" with internal battery and multiple alarms, for patients ventilating more than 16 h/24 or dependent on ventilation.

The resuscitator is therefore responsible for prescribing the home ventilator when the patient is discharged, and must constantly check that the choice of ventilator is perfectly suited to the severity of the patient's condition. As far as ventilatory modes are concerned, pressure modes are currently the most widely used[17]. This trend is due to the emergence of non-invasive ventilation, where pressure modes have the advantage of compensating for any leaks. Some home ventilators feature mixed modes that theoretically combine the advantages and disadvantages of volumetric and barometric modes, but they have yet to demonstrate their superiority over conventional modes.

Conclusion:

Myasthenia-SAOS is a frequent but often unrecognized association, and the diagnosis should be made in the event of weaning difficulties in the intensive care unit, after eliminating obvious causes. Polysomnography remains the gold standard for confirming the diagnosis of OSA, but it is complex to perform in the intensive care unit. The use of a questionnaire may be an alternative for making the diagnosis when there is a strong clinical suspicion. Management includes ventilatory support, early tracheostomy, progressive weaning, prevention of thromboembolic disorders, nursing and long-term mechanical ventilation at home.

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