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СЕКЦИЯ „ХИРУРГИЯ, АНЕСТЕЗИОЛОГИЯ И ИНТЕНЗИВНО ЛЕЧЕНИЕ“

АНЕСТЕЗИОЛОГИЧНИ И РЕАНИМАЦИОННИ ПРОБЛЕМИ ПРИ МИАСТЕНИЧЕН СИНДРОМ НА LAMBERT-EATON (LEMS) - КЛИНИЧЕН СЛУЧАЙ

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Резюме: *Въведение: Миастеничният синдром на Lambert-Eaton (LEMS) с честота 2–3 случая на 1 000 000 души, известен също като синдром на Eaton-Lambert е автоимунно заболяване на невро-мускулното съединение, при което имунната система атакува пресинаптичните волтаж-зависими калциеви канали. Това води до нарушено освобождаване на ацетилхолин, което пречи на способността на нервните клетки да изпращат сигнали към мускулните. Води до постепенна прогресия на мускулна слабост и дисфункция на автономната нервна система. В около 50–60% от случаите LEMS е паранеопластичен синдром, най-често свързан с дребноклетъчен белодробен карцином. Пациентите подложени на анестезия с LEMS имат повишена чувствителност както към деполяризиращи, така и към недеполяризиращи невромускулни блокери и са изложени на повишен риск от развитие на периоперативни респираторни усложнения с потенциална нужда от постоперативна апаратна вентилация.*
Цел: На база на реален клиничен случай преминал през интензивното отделение на МБАЛ Лайф Хоспитал да разгледаме предизвикателствата от гледна точна на анестезиологията и интензивното лечение при такива пациенти.
Материали и методи: Направен е подробен анализ на наличната медицинска информация по отношение на проблемите при пациенти със синдрома на Ламберт–Итън (LEMS) и анализ на анестезиологичните и реанимационни особености при пациенти с това заболяване. Разгледани са специфичните особености в представения от нас клиничен случай.
Заключение: Синдромът на Ламберт–Итън представлява сериозно предизвикателство за анестезиологията и интензивното лечение поради променената невро-мускулна трансмисия и високата чувствителност към миорелаксанти, както и прогресиращата във времето мускулна слабост. Успешният подход при лечението на тези пациенти изисква внимателно предоперативно планиране, мониториране и индивидуализиран анестезиологичен подход, както и прецизиране на решението за нужда от апаратна вентилация в периода на интензивни грижи.
Ключови думи: синдром на Ламберт–Итън, миастеничен синдром, анестезиология, интензивно лечение, невромускулна трансмисия.

ANESTHESIOLOGICAL AND RESUSCITATION PROBLEMS IN LAMBERT-EATON MYASTHENIC SYNDROME (LEMS)- A CLINICAL CASE

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Abstract: *Introduction: Lambert–Eaton myasthenic syndrome (LEMS), with an incidence of 2–3 cases per 1,000,000 individuals and also known as Eaton–Lambert syndrome, is an autoimmune disorder of the neuromuscular junction in which the immune system targets presynaptic voltage-gated calcium channels. This results in impaired release of acetylcholine, thereby disrupting the ability of nerve cells to transmit signals to muscle cells. The condition leads to a gradual progression of muscle weakness and autonomic nervous system dysfunction. In approximately 50–60% of cases, LEMS is a paraneoplastic syndrome, most commonly associated with small-cell lung carcinoma. Patients with LEMS undergoing anesthesia exhibit increased sensitivity to both depolarizing and non-depolarizing neuromuscular blocking agents and are at increased risk of perioperative respiratory complications, with a potential need for postoperative mechanical ventilation.*

Aim: Based on a real clinical case treated in the intensive care unit of Life Hospital, to discuss the challenges from the perspective of anesthesiology and intensive care management in patients with LEMS.

Materials and Methods: A detailed analysis of the available medical literature concerning the problems encountered in patients with Lambert–Eaton myasthenic syndrome (LEMS) was conducted, along with an analysis of the anesthetic and intensive care considerations in such patients. The specific features of the presented clinical case were also examined.

Conclusion: Lambert–Eaton myasthenic syndrome represents a significant challenge for anesthesiology and intensive care due to altered neuromuscular transmission, high sensitivity to neuromuscular blocking agents, and progressively worsening muscle weakness. Successful management of these patients requires careful preoperative planning, close monitoring, and an individualized anesthetic approach, as well as precise decision-making regarding the need for mechanical ventilation during the period of intensive care.

Keywords: Lambert–Eaton myasthenic syndrome, myasthenic syndrome, anesthesiology, intensive care, neuromuscular transmission.

Introduction

Lambert-Eaton syndrome is also known as Lambert-Eaton myasthenic syndrome(LEMS). It is autoimmune disorder in which the immune system attacks the neuromuscular junctions. Lambert-Eaton myasthenic syndrome is a rare disease. More specifically, voltage-gated calcium channels in the presynaptic neuronal cell membrane are attacked by antibodies, and with fewer calcium channels, acetylcholine release from the presynaptic nerve terminals is reduced. Worldwide, it affects about 2.8 million people. In the U.S., about 400 people have LEMS.

Types and clinical characteristics of lems

There are two types of LEMS:

1. Paraneoplastic LEMS is often associated (50-60%) with small-cell lung cancer. Treat underlying cancer as this may improve symptoms from this condition. Characterized by an older age of onset (average 60 years). Caused by an accidental attack by the immune system as it attempts to fight the cancer
2. LEMS may also be associated with:

Endocrine diseases such as hypothyroidism or diabetes mellitus type 1. It may have a genetic component linked to autoimmunity . Younger age of onset (on average 35 years).

Myasthenia gravis has very similar symptoms to Lambert-Eaton syndrome. In contrast to myasthenia gravis, the weakness in Eaton–Lambert syndrome usually improves with exercise.

Primary clinical manifestation: muscle weakness and muscle fatigue, Trouble walking, Muscle pain or stiffness, Tingling sensation in your hands or feet, Droopy eyelids (ptosis), Double vision (diplopia), Dry mouth and dry eyes, Constipation, Decreased sweating, Weight loss, Difficulty peeing, Erectile dysfunction, Dysarthria and dysphagia are late-stage symptoms, Dyspnea and respiratory failure.

Disease progression and pathophysiology

How does Lambert-Eaton myasthenic syndrome typically appear and progress?

Lambert-Eaton myasthenic syndrome typically affects leg muscle strength (upper leg) first, followed by shoulder muscles, muscles of your hands and feet, muscles affecting your speech and swallowing, and eye muscles.Early in the disorder, you may have trouble getting up from a chair, walking up stairs or a steep walkway, or doing anything strenuous with your legs. Symptoms usually develop slowly over weeks to many months. Symptoms develop more quickly if you have cancer and LEMS.

Etiology and immunopathogenesis

What causes Lambert-Eaton myasthenic syndrome (LEMS)?

Lambert-Eaton myasthenic syndrome is an autoimmune disorder. This means your body's natural defenders (antibodies) attack your own healthy tissue. In LEMS, your antibodies attack where nerve cells meet muscle fibers (neuromuscular junction); more specifically, at calcium channels that are critical in signaling at this contact point.

Like a lineup of dominoes, the attack causes a sequence of events:

1. Antibodies bind to and block calcium channels at the end of nerve cells.
2. Fewer calcium channels mean less of the neurotransmitter acetylcholine is released. Acetylcholine is critical to signal your muscles to activate.
3. As there isn't enough acetylcholine released, not enough "messages" reach muscle fibers, and then your muscles can't be fully activated. Your muscles won't function as they normally should, which causes weakness.

LEMS happens in people who have small-cell lung cancer. In this case, your body's antibodies recognize their "call to action" when there's cancer in your body. However, instead of attacking the cancer cells (specifically, the calcium channel on cancer cells), it attacks the calcium channel at the end of nerve cells. And the sequence listed above happens. When this happens, your muscle cells can't pick up the message to contract to make your muscle move.

Diagnostic approach

The first step in the diagnosis of LEMS is a detailed history of your symptoms, as well as a review of your medical and medication history. A detailed neurological examination is often done to look for patterns of weakness and signs consistent with a diagnosis of LEMS.

The next step in the evaluation is testing. Testing for LEMS includes:

- **Blood test.** A blood test can reveal if you have anti-calcium channel antibodies. About 85% of people with LEMS have these antibodies in their blood.
- **Electromyography.** This test shows how well your muscles and nerves are working together. LEMS produces very specific findings on electromyography that can help confirm a diagnosis.
- **Lung X-ray or computed tomography (CT) scan or magnetic resonance imaging (MRI) of your chest.** These imaging tests check for signs of lung cancer, as LEMS can happen in people who have small-cell lung cancer.

LEMS can appear from months to up to six years before the cancer does. The typical lung cancer screening schedule is every three to six months for at least two years after your diagnosis. Your healthcare provider will recommend a screening schedule that's appropriate for you.

Therapeutic strategies in lems

Symptom treatment

- Amifampridine (Firdapse®), Guanidine.
- Pyridostigmine (Mestinon®).

Immune-modulating treatment

- Immunosuppressants.
- Plasma exchange.

- Immunoglobulins

Anesthesiological and intensive care challenges

Lambert–Eaton syndrome poses a serious challenge to anesthesiology and intensive care because of:

1. Respiratory failure: although in many patients respiratory function is relatively preserved, cases of acute or progressive respiratory compromise, including the need for mechanical ventilation, have been described. The risk is higher in the presence of bulbar/fascial involvement, progressive general weakness, concomitant pulmonary infections or restrictions (pneumonia, abscess), paraneoplastic activity and therapeutic interactions.
2. Changes in physiological parameters and functions: reduced vital capacity (VC), reduced spontaneous volumes (VT), ineffective cough, and risk of aspiration in bulbar involvement — all predispose to pulmonary complications and a longer ventilatory period.
3. Hypersensitivity to muscle relaxants. LEMS alters neuromuscular transmission, leading to a pronounced sensitivity to the drugs used in intubation.

Problems: Depolarizing muscle relaxants (e.g., suxamethonium). Patients may show prolonged block and disproportionately long paralysis. Nondepolarizing muscle relaxants: Even usual doses can cause prolonged and profound muscle relaxation, making extubation difficult, recovery of spontaneous breathing difficult, and stable neuromuscular function difficult. Both groups (depolarizing and nondepolarizing) can cause unpredictable hypersensitivity and prolonged paralysis at lower doses than expected. This increases the likelihood that the patient will remain dependent on mechanical ventilation for longer than expected and may make extubation difficult.

4. Autonomic dysfunction — impact on response to intubation and ventilation. LEMS often presents with signs of autonomic dysfunction (dry mouth, orthostatic hypotension, anhidrosis, other autonomic disturbances). In a clinical context, this means: labile blood pressure and variable hemodynamics during intubation/seizure/recovery; increased risk of arrhythmias or hemodynamic reactions with certain medications; changes in bronchial and salivary secretions, which affect the risk of aspiration and the need for targeted secretion management.

Airway management and neuromuscular blockade

Principles and issues in necessary intubation. 1. Pre-procedural assessment: clinical assessment of respiratory function (VC, spontaneous VT, cough response) and assessment for bulbar involvement and autonomic dysfunction are key. In patients with progressive weakness or pulmonary comorbidities, the risk of postoperative ventilation is increased. 2. Use of neuromuscular blockers: according to literature data, there is an increased sensitivity: practical implications include a preference for minimal or no use of NMBAs if intubation can be performed with adequate intubation technique (Rapid Sequence Induction if necessary) and/or the use of short-acting agents with the possibility of monitoring and a short duration of action of the agent used;

When NMBAs are absolutely necessary, dose adjustment and continuous monitoring with a neuromuscular stimulator should be considered 3. Monitoring: intraoperative monitoring of neuromuscular block (TOF), capnography, arterial blood gases, and infusion/hemodynamic monitoring are critical for timely detection of respiratory dysfunction and avoidance of prolonged ventilation.

Mechanical ventilation and weaning considerations

Mechanical ventilation - features and challenges Indications: acute respiratory failure, decompensation due to infection, operations with risk of postoperative respiratory failure, ineffective airway protection (bulbar symptoms). Modes and goals: ventilation principles follow general intensive are standards (ensuring adequate oxygenation, control of $PaCO_2$ and volutrauma/atelectrauma). Specifically in LEMS, the following should be considered: low reserve for spontaneous breathing → need for more conservative airflow titration and maintenance regimens; • possibility of using non-invasive ventilation (NIV) to assist in weaning attempts when

the bulbar reflex and protective cough are adequate. Secretion and aspiration: increased risk of aspiration requires attention to secretion control and, if necessary, bronchial hygiene; bulbar disorders require increased caution during extubation. (!).

Extubation and weaning from the ventilator in patients with LEMS is often complicated by: • lability of muscle strength (possible short-term improvement after activation followed by exhaustion), • prolonged sensitivity to NMBAs and a level of anesthesia that may suppress respiratory control, • weakness of the cough reflex and risk of aspiration

As a result, successful extubation often requires: adequate neuromuscular monitoring (TOF > defined values according to clinical protocol), assessment of VC and cough force, possibility of transition to NIV, and readiness for re-intubation

Clinical case

The patient was admitted from the intensive care unit with severe respiratory failure based on right-sided bronchopneumonia, visible on a chest X-ray. The patient had a history of Eaton Lambert syndrome for about 4 years and was on therapy with Firdapse and Pyridostigmine. During the day, she visited a medical center, where antibiotic therapy was started, corticosteroids, oxygen therapy were administered and the patient was transported to the Life Hospital on 26.08.2025.

Current status upon admission - it concerns a woman of apparent age corresponding to the actual one, with pronounced obesity, impaired general condition. Psychomotor excited, with pronounced tachypnea. Against the background of 6l O2 with a mask, she maintains peripheral saturation around 92%. Hemodynamically stable. Skin - warm and important. Auscultation - bronchial breathing in the right lung base, dry wheezing rales bilaterally, prolonged expiration. After a consultation with a pulmonologist, the therapeutic behavior was optimized.

Therapy: Ceftriaxone 2 x 2 grams i.v., Klacid 2x500mg i.v., Cortisone 2 x 40mg i.v., Inhalations with Pulmicort and Ventolin - three times a day, Pyridostigmine 60mg - per os - every 4 hours, Firdapse - per os - 2t. every four hours, Fraxiparine - 0.8 s.c., Ringer 500ml - infusion, Omeprazole 40mg i.v.

On 27.08.24, a consultation with a neurologist was held and the following neurological status was described:

This is a patient with a history of Ethan Lambert's syndrome of about 4 years, she is being treated with Kalimin 4x60 mg, she was admitted with respiratory failure, fever with X-ray data for bronchopneumonia on the right. Objective condition - contact, adequate Cmn-eyeballs mobile in all directions, no diplopia and dysphagia, tests for latent paresis are not positive. Comparatively preserved muscle strength for all four limbs

Objective condition as of 27.08.25: Impaired general condition. Against the background of 5l O2 with a face mask, good oxygenation is maintained, from peripheral CAS persistent hypercapnia. In the right lung base - bronchial breathing, prolonged expiration, dry wheezing wheezing. When coughing, purulent secretions are secreted. Awake, communicative, oriented. Skin - dry and warm. Effective diuresis. Afebrile.

The blood gas analyses performed showed pronounced respiratory acidosis, with evidence of hypercapnia and carbon dioxide retention despite the non-invasive pulmonary ventilation applied:

Blood gas analysis data: 27.08.2024 CAS (ACR)• ph: 7.306; • pCO2: 71.00; • pO2: 71.00; • HCO3: 35.4; • Beb: 6.2; • O2Sat: 91.4; • tCO2: 37.6 27.08.2024 » CAS (ACR) • ph: 7.275; • pCO2: 78.70; • pO2: 70.00; • HCO3: 36.6; • Beb: 6.4; • O2Sat: 90.1; • tCO2: 39.0; » CAS (ACR) 27.08• ph: 7.313; • pCO2: 62.50; • pO2: 121.00; HCO3: 31.7; • Beb: 3.4; • O2Sat: 98.2; • tCO2: 33.6;

On 28.08.25 there was no significant change in the condition. Hypercarbia persisted. Severe oxygen dependence. Sat02 88-90% at 7-8 l/02. Data from the blood gas analysis: 28.08.2024» KAS (AKR)• ph: 7.306; • pCO2: 76.10; • pO2: 88.00; • HCO3: 38.0; • Beb: 11.5; • O2Sat: 95.1; • tCO2: 40.3;. With stable hemodynamics and a relatively cooperative patient. Within the day, the patient was transported by ambulance to Israel.

Discussion

The main challenge in the described clinical case is the decision for or against starting mechanical ventilation. When analyzing the blood gas analyses performed, there is evidence of acute respiratory failure with pronounced respiratory acidosis and carbon dioxide retention on the background of bilateral pneumonia. However, with preserved consciousness and adequate reactions, a decision was made to resort to invasive ventilation only in case of quantitative disturbances in consciousness and/or instability of the main vital signs, in order to avoid the risk of protracted weaning from the device and dependence on the respirator. Gradually, the patient's clinical condition was stabilized at this critical level without dynamics towards deterioration, which allowed for transportation without the need for mechanical ventilation, which we consider to be the correct therapeutic approach in this case.

Conclusion

In conclusion, it can be said that patients with this syndrome represent a serious challenge for developing an adequate therapeutic strategy and balancing the benefits and risks when deciding on the degree of invasiveness of respiratory support.

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