

Zespół Lemierre'a

(Lemierre's syndrome)

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Streszczenie – Autorzy przedstawili opis rzadko obecnie spotykanego zespołu Lemierre'a. Zwrócono uwagę na etiologię, objawy, diagnostykę i leczenie. Niska częstotliwość występowania danej jednostki chorobowej może przyczynić się do niedostatecznej wiedzy personelu na jej temat lub jej braku. Ma to ogromny wpływ na postawienie prawidłowej diagnozy, jak i czas jej rozpoznania.

Słowa kluczowe - zespół Lemierre'a, epidemiologia, objawy, leczenie, rokowanie.

Abstract – The authors have described a condition that nowadays is encountered only rarely, namely Lemierre's syndrome. They paid attention to its aetiology, diagnostics and treatment. The low frequency of occurrence of a given disease classification can contribute to the personnel having insufficient knowledge about it or not having it at all. That has a crucial impact on coming up with a correct diagnosis as well as on the time of recognizance.

Key words - Lemierre's syndrome, epidemiology, symptoms, treatment, prognosis.

I. INTRODUCTION

Lemierre's syndrome develops most often after a sore throat with a complication of circumtonsillar abscess filled with pus and bacteria. On the bottom of the abscess, anaerobic bacteria that do not require oxygen like *Fusobacterium necrophorum* have conditions optimal to flourish. The bacteria leave the the abscess and head for the neighbouring jugular vein in the neck, where they form an infected clot. The clot is carried by the bloodstream to reach various organs, causing bacteremia.

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Parts of the clot travel to the lungs as emboli blocking branches of the pulmonary and bronchial arteries. That leads to shortness of breath, chest pain and severe pneumonia. *Fusobacteria* are a normal physiological parts of the oropharyngeal flora.

Sepsis caused by a throat infection was first described by Scottmuller in 1918. In 1936, André Lemierre published a series of 20 cases where throat infections were followed by confirmed anaerobic septicaemia. 18 of those patients died [2]. Lemierre's syndrome is a relatively rare disease unit in modern-day medicine. Its high frequency had been observed before penicillin was introduced into therapies in the early 20th century. The present tendency to avoid antibiotics in the treatment of pharyngitis may lead to increased risk of this syndrome. In 1997, 19 cases of Lemierre's syndrome were described in Great Britain, whereas in 1999 the number rose to 34. The incidence rate is 0,8 cases per million in general population, which is the reason why Lemierre's syndrome is nicknamed a "forgotten disease" [3]. The disease affects mainly young, healthy adults.

II. PATHOGENESIS

Fusobacterium necrophorum is an etiologic factor that occurs in most of the cases of Lemierre's syndrome. The *Fusobacterium necrophorum* infection leads to Lemierre's syndrome only in 1 per 400 cases [4].

Lemierre's disease is caused by *Fusobacterium necrophorum* in 81% of the cases, while the other 11% are caused by other species of *Fusobacterium*. Also, methicillin-resistant *Staphylococcus aureus* (MRSA) microbes can contribute to the development of Lemierre's syndrome. Infrequently, such bacteria as *Bacteroides fragilis*, *melaninogenicus*, *Peptostreptococcus spp*, *Streptococcus microaerophile*, *Staphylococcus ureus* and *Eikenella corrodens* were the cause of the disease [5].

Lemierre's syndrome begins with an infection of head and/or neck area. Usually it is pharyngitis, occurring in 87,1%

of the cases. The disease may also follow otitis, mastoiditis, sinusitis or mumps.

During the primary infection, *F. necrophorum* colonizes the infected area which expands in the peripharyngeal space. Then, the bacteria move to peritonsillar blood vessels, where they find their way to the internal jugular vein. There, a clot that contains the bacterial material is formed. What is more, that leads to an inflammation of that vein. As parts of the clot are separated from it, the infected embolic material is carried all over the organism, which may cause abscesses to form in distant organs and increase the risk of embolism. At first, the embolic material reaches pulmonary circulation capillaries. As a result, the most frequent location of abscess formation are lungs, followed by major joints (knee, hip, shoulder, elbow and sternoclavicular joints). In the lungs, the bacteria form abscesses, involve lymph nodes and cause necrosis in the lung tissue. Frequently, there is fluid in pleural cavities. What is more, the infected embolic material may deteriorate to muscle tissue, connective tissue, liver, spleen, kidneys and nervous system (causing, for that matter, meningitis and brain abscesses).

Bacterial toxins such as lipopolysaccharide (LPS) may disable cytokinogenesis and facilitate the development of sepsis. *F. Necrophorum* also produces hemagglutinin initiating platelet aggregation, which may lead to disseminated intravascular coagulation (DIC) with subsequent consumption thrombocytopenia.

III. DIAGNOSIS

Symptoms differ depending on each case, yet most of the time the illness begins with pharyngitis, fever and generalized weakness. Then, serious exhaustion is experienced and septic fevers occur. What follows is lymphadenopathy; also, the neck becomes numb, swollen and sensitive. Usually, abdominal pain, nausea, diarrhea and vomiting also occur. These symptoms can be observed for as long as between a few days and two weeks since the first symptoms occur [5].

The symptoms which point to the involvement of respiratory system are airlessness, coughing and pleurodynia. Haemoptysis is observed as well, yet rather rarely. In case joints are involved, also pain and other symptoms of arthritis.

The symptoms of septic shock are hypotension, tachycardia, oliguria and tachypnoe. Numbness of the neck, headache and photophobia may suggest meningitis. Also, hepatomegaly and splenomegaly may occur, not necessarily caused by abscesses in liver and spleen, respectively [6].

Other symptoms include: headaches (unrelated to meningitis), muscle pains (general pain in the whole body), jaundice, lock-

jaw, lung field crackles (rales), friction rub as a symptom of pericarditis (rarely), cranial nerve palsy, Horner's syndrome (myosis, ptosis, endophthalmus – occurring rarely).

Diagnosics

The range of laboratory and imaging examinations are determined depending on the patient's general condition. If pharyngitis persists and the aforementioned symptoms occur, health screening should be performed with regard to Lemierre's syndrome.

Laboratory tests indicate bacterial infection with higher acute-phase protein level (CRP), increased erythrocyte sedimentation rate (ESR) and leukocytes, especially neutrophils. The level of platelets is usually higher than normal, although it can be too low as well. Liver and kidney parameters are generally abnormal.

Thrombosis of the jugular vein may be exposed in ultrasound examination. Nevertheless, a newly formed clot may not be visible in the USG of jugular veins as its echogenicity is low. In such cases, a CT scan or MRI may prove to be more effective [3].

The X-ray or tomography may indicate fluid in pleural cavities, parenchymal densification within lung fields and abscesses.

Bacterial cultures from blood, synovial fluid and other locations may enable the identification of the etiologic factor. The following disease units should be taken into account in differential diagnosis: Q fever, tuberculosis and pneumonia of some other aetiology.

IV. TREATMENT AND PROGNOSIS

Lemierre's syndrome is usually treated by antibiotics applied intravenously. However, because a frequent cause of pharyngitis is a virus infection, antibiotics are not obligatory in the first line treatment [7].

Fusobacterium necrophorum is highly susceptible to beta-lactam antibiotics, metronidazole, clindamycin and third-generation cephalosporins, whereas other *Fusobacterium* strains show different level of susceptibility to beta-lactams and clindamycin. Moreover, a co-infection by other microbes may occur in a given single case of the disease. Therefore, monotherapy is not recommended for treating Lemierre's syndrome. Usually, a penicillin antibiotic along with a beta-lactamase inhibitor (like clavulonic acid) and metronidazole are applied. Clindamycin can be applied in monotherapy [8].

When antibiotics fail, the formed abscesses need to be removed surgically. Also, the clots must be removed from the internal

jugular vein and antibiotics applied to the area of the infected clot.

There are no unequivocal indications as far as anticoagulant therapy for Lemierre's syndrome is concerned. The decisions are made depending on each patient's condition. In case there are complications (meningitis, severe pneumonia), the disease may be resistant to treatment [6].

Mortality rate of Lemierre's syndrome before the antibiotic era was very high, amounting to as much as 90%. Presently, as it can be timely diagnosed and the treatment can be quickly implemented, the mortality rate is between 6,4 and 15% [6]. Nevertheless, in the cases when the diagnosis is delayed, the mortality rate may be significantly higher [9].

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