Vascular Complication of Sore Throat: Lemierre's Syndrome: Case Report

Lemierre Sendromu: Boğaz Enfenksiyonunun Vasküler Komplikasyonu

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ABSTRACT Lemierre's syndrome is a rare disease that occurs due to an oropharyngeal infection causing internal jugular vein thrombosis and metastatic infection. It is usually seen in previously healthy adolescents and young adults. The predominant pathogen is a Gram-negative anaerobic bacillus, *Fusobacterium necrophorum*. This syndrome is routinely treated with intravenous antibiotics and anticoagulants. Herein we report a patient who presented with acute sore throat that led to a bacteremia with the left internal jugular vein thrombosis, and treated with intravenous antibiotics and anticoagulants in the light of the literature.

Key Words: Lemierre's syndrome; jugular vein thrombosis; septicaemia

ÖZET Lemierre Sendromu orofaringeal enfeksiyon sonucu internal juguler ven trombozu ve metastatik enfeksiyonlara neden olan, nadir bir hastalıktır. Genellikle öncesinde sağlıklı adolesan ve genç erişkinlerde görülür. Hakim patojen Gram-negatif anaerobik basil olan *Fusobacterium necrophorum*'dur. Bu sendrom rutin olarak intravenöz antibiyotik ve anti-koagülan ile tedavi edilir. Burada, akut boğaz ağrısının neden olduğu bakteriyemi ile birlikte sol internal juguler ven trombozu ile başvuran ve intravenöz antibiyotik ve düşük moleküler ağırlıklı heparin ile tedavi ettiğimiz hastamızı ve literatüre bakışı sunuyoruz.

Anahtar Kelimeler: Lemierre sendromu; jugular ven trombozu; septisemi

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Lemierre's syndrome or postanginal septicaemia (necrobacillosis) is the septic thrombophlebitis of the internal jugular vein (IJV) following acute oropharyngeal sepsis. *Fusobacterium necrophorum* is the most common pathogen isolated from the patients. It typically affects previously healthy adolescents and young adults and often follows an episode of pharyngotonsillitis.[1] The clinical picture tends to vary widely because of the possible involvement of a number of body systems and organs in the disease process. This potentially life-threatening syndrome can be cured by rapid initiation of antibiotherapy. The morbidity or mortality is caused mainly by lack of knowledge of the syndrome. In this article, we report a patient who presented with acute sore throat that led to a bacteremia with internal jugular vein thrombosis, review the literature.

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CASE REPORT

A previously healthy 38-year-old woman was admitted with a three-day history of sore throat, painful right-sided neck swelling, increasing dyspnea, fever and rigor. On examination, vital signs included a temperature of 38.8°C, pulse 102/min, blood pressure 100/45 mmHg and respiratory rate 24/min. There were pustular exudates on the left tonsil, and a warm and extremely painful swelling was palpable alongside the left sternocleidomastoid muscle. Auscultation of the chest revealed normal breathing sounds.

Laboratory tests showed an elevated sedimentation rate of 64 mm/hr, increased high-sensitivity C-reactive protein (hs-CRP) level of 143 mg/dL, and leukocytosis of 23,700/mm^3.

The chest radiograph was normal. Computerized tomography (CT) of the neck revealed thrombosis of the left internal jugular vein with lymphadenopathies. Color-coded Doppler sonography confirmed this diagnosis, and determined thrombosis of the left internal jugular vein (Figure 1). Magnetic resonance imaging (MRI) of the parapharyngeal region was performed to investigate this region in detail, however, there were no abscesses either in the retropharyngeal region or parapharyngeal region. MRI revealed the thrombosed left internal jugular vein.

The patient was treated with the empiricial antibiotics consisting of cefuroxime, metronidazole and gentamycine. Blood cultures yielded *Fusobacterium necrophorum*, confirming the diagnosis of Lemierre's syndrome. After identification of *Fusobacterium necrophrum*, the antibiotic regimen was switched to monotherapy with penicillin G, and continued for 6 weeks. The patient was also treated with low-molecular-weight heparin (Nadroparin calcium, 5700 IU, twice daily). After discharge, she continued oral clindamycin for 4 weeks. Complete convalescence was noted with follow up radiological studies showing resorption of the thrombus in the left jugular vein in six months (Figure 2).

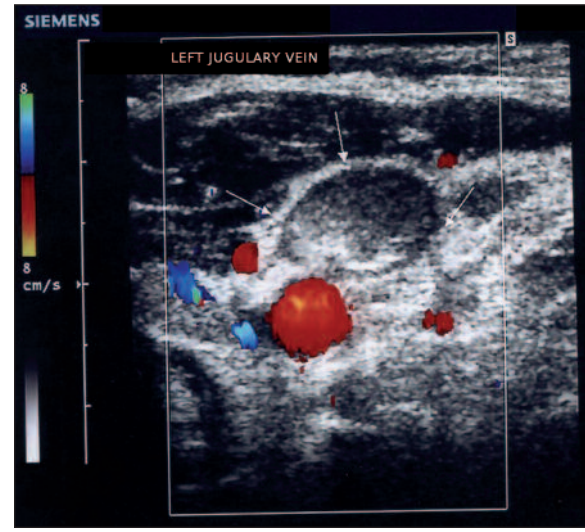

FIGURE 1: Color-coded Doppler sonography view of the left jugular vein showing a thrombus in the lumen of the vessel.

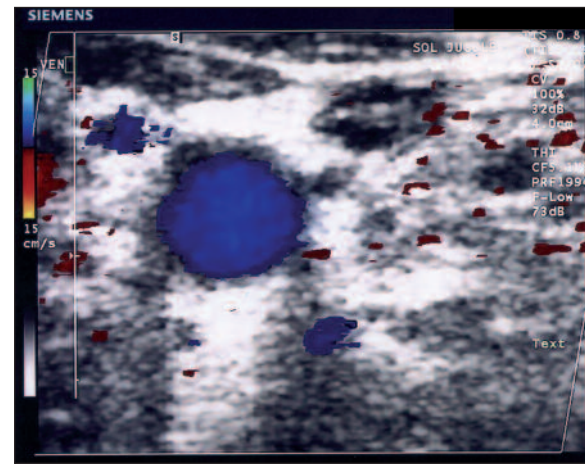

FIGURE 2: Color-coded Doppler sonography showing resorption of the thrombus in the left jugular vein after six months.

DISCUSSION

Lemierre syndrome, also known as postanginal sepsis, is an illness characterized by the development of fusobacterial septicaemia with thrombophlebitis of the internal jugular vein, and metastatic infections predominantly in the lungs and joints following an attack of acute tonsillitis.[1] It is usually seen in previously healthy adolescents and young adults. In the literature, 73.4% of patients were between the ages of 16 and 40 years.[1]

Our patient was a previously healthy 38-year-old woman.

In Lemierre Syndrome, the oropharynx is the primary site of infection in most of the cases. The interval between the oropharyngeal infection and the onset of the septicaemia is usually one week or less.[2] However, the signs and symptoms related to oropharyngeal infection may have cleared by the time internal jugular vein thrombosis or metastatic infection develop, even without a prior antibiotic therapy.[3,4] Clinical signs and symptoms include fever, painful swelling and tenderness or induration at the angle of the mandible and along the sternocleidomastoid muscle, representing septic thrombophlebitis of the internal jugular vein.[5] Lemierre Syndrome usually occurs at the ipsilateral internal jugular vein. Bilateral thrombophlebitis of the internal jugular vein can rarely occur; in the literature there are two reported cases of Lemierre's syndrome in which internal jugular vein thrombosis occurred bilaterally. The first case was reported by Moore et al. in 2002, and Benhayoun et al. reported the second case in 2003.[6,7]

The suspicion of Lemierre's syndrome should prompt investigation for internal jugular vein thrombosis, that can be revealed by ultrasonography.[8] A CT scan or MRI can also be used to assess the extent of internal jugular vein thrombosis and the presence of micro-abscess formation.[9,10] Contrast-enhanced CT has been considered as the imaging method of choice because it is more sensitive, and allows discovery of additional pathologies such as pulmonary emboli, soft tissue abscesses, osteomyelitis, and septic arthritis.[11] The Color-coded Doppler sonography, as the imaging method in follow-up of the lesions, was used in our case.

Metastatic infection is another feature of Lemierre's syndrome. The most common site of metastatic involvement is the lungs, with 97% incidence reported in the literature.[2] Multiple metastatic abscesses may follow, and septic arthritis is another consistent finding. Large joints, including knee, hip, shoulder, ankle, and sternoclavicular joint, are often involved, and multifocal osteomyelitis may occur.[8] Metastatic infections of the central nervous system, spleen, liver, and soft tissue were also reported.[2,12,13] There were not any clinical and diagnostic evidence for abscesses in our patient, and we followed her closely for complications.

The causative microorganism is frequently *Fusobacterium necrophorum*, a Gram-negative anaerobic organism that normally inhabits the oropharynx. It is presumed that the extracellular products of the bacteria play an important role in its spread into the tissues and bloodstream.[2] The treatment is mainly prolonged, high-dose antibiotics.[8] Generally, Fusobacterium flora are susceptible to penicillin. Some strains are penicillin resistant, through production of beta-lactamase.[14] Clindamycin, metronidazole, antipseudomonal penicillins, and ampicillin-sulbactam offer good coverage.[15] The role of anticoagulation in Lemierre's disease is controversial. Bach et al. found that treatment with heparin was associated with a more rapid resolution of the septic emboli, but there have not been any controlled studies examining the role of anticoagulation.[16] It is generally not recommended, except in the case of retrograde extension of the thrombus to the cavernous sinus.[1,17] Yücel et al. suggested that patients with genetic thrombophilic mutations and/or antiphospholipid syndrome should use anticoagulants for lifelong.[18] In our case, we preferred to use anticoagulation in order to avoid septic emboli.

In conclusion, although rare, Lemierre's syndrome is a potentially lethal but treatable complication of acute oropharyngeal sepsis. Early diagnosis and treatment with anticoagulants and parenteral antibiotics is life-saving.

Conflict of Interest

Authors declared no conflict of interest or financial support.

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