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Risk of Delayed Sepsis Diagnosis in Young Patients-Case Report of Post Anginal Sepsis (Lemierre's Syndrome)

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Abstract

Lemierre's syndrome represents a rare, but life-threatening cause of sepsis syndrome characterized by initial pharyngeal infection. Delayed diagnosis may lead to severe complications and fatal outcome. Here we report a case of a 27-year-old adult presenting in the Emergency Department with prolonged throat soreness. Manifestation of Lemierre's syndrome including pharyngeal abscesses, left internal jugular vein thrombosis and multiple septic embolic metastases could only be diagnosed by contrast enhanced computed tomography and highly elevated procalcitonin levels in this case. After initiation of antibiotic and surgical treatment the patient recovered without any complications. The case report underlines the diagnostic challenges and the importance of Lemierre's syndrome as a rare, but potentially life-threatening cause of sepsis.

Keywords: Lemierre'S Syndrome; Sepsis; SOFA Score; Fusobacterium; Necrobacillosis; Internal Jugular Vein Thrombosis

Introduction

Pharyngeal infections commonly affect healthy adolescents and young adults. Lemierre's syndrome [1] is a rare, but severe and life-threatening complication of pharyngeal infections with septic embolic metastases and internal jugular vein thrombosis caused by Fusobacterium necrophorum septicemia [2,3]. Despite its increasing incidence over the past decades, Lemierre's Syndrome, once labeled 'the forgotten disease', remains widely unknown among clinicians thus often resulting in delayed diagnoses and high mortality rates [4-6]. To demonstrate the diagnostic challenges in achieving a time-critical and accurate diagnosis for initiation of adequate treatment we here present a case of Lemierre's syndrome in a 27-year-old young adult.

Case Presentation

A 27-year-old patient without any comorbidities, but an episode of prolonged throat soreness for almost two weeks was admitted to the interdisciplinary Emergency Department (ED) at the University Medical Center Goettingen. In patient's actual history the initially right-sided symptoms were pretreated empirically with cefuroxime by a general practitioner as bacterial tonsillitis was supposed. After discontinuation the now left-sided soreness returned with intermittent ague. At the time of presentation in the ED the patient was in good general condition and seemingly minimally impaired by his symptoms.

Clinical examination revealed a mildly erythematous palate and pharynx, inconspicuous palatine tonsils, dry oral mucous membranes and upper abdominal pain. No signs of intraoral or pharyngeal asymmetry or putrid tonsils, stridor, dysphagia,

meningism or lockjaw were detected. Vital signs showed sinus tachycardia (110/min), normotension (103/62 mm Hg, MAP 76 mm Hg), eupnea (15/min, 96 % oxygen saturation) and febrile auricular temperature (38,0 °C).

Diagnostics and Treatment

Initial diagnostics included blood sampling, blood cultures, blood gas analysis, electrocardiogram and point-of-care-ultrasound. An otorhinolaryngologist consultation including laryngo-pharyngeal endoscopy and cervical ultrasound was performed revealing no further specific pathologies.

First available laboratory results included the blood cell count with thrombocytopenia (49 000/µl), elevated C-reactive protein (CRP 186 mg/l), creatinine (2.54 mg/dl, 223.52 µmol/l) and bilirubin levels (3.3 mg/dl, 56.1 µmol/l), but no leukocytosis (10.400/µl).

Common sepsis screening tools were negative or showed a low probability for systemic infection (Modified Early Warning Score MEWS = 2 points, National Early Warning Score NEWS = 3 points, Systemic Inflammatory Response Syndrome SIRS = 1 point, Quick Sepsis-related Organ Failure Assessment score qSOFA = 0 points, initial lactate = 1.1 mmol/l).

Differential diagnosis of thrombocytopenia and acute kidney injury followed immediately. We extended laboratory testing with differential blood cell count, procalcitonin (PCT), urea, lactate dehydrogenase (LDH), haptoglobin, urine status, sediment, and culture, as well as serologic immunoassays for Leptospirosis and Hantavirus infection. Differential blood cell count revealed neutrophilia (9/nl), reactive left shift, thrombocytosis and erythroanisocytosis (MPV 9,5 fl, PDW 17,3), but no signs of hemolysis such as fragmentocytes were seen. Impressively elevated PCT levels (315 µg/l) confirmed the suspected systemic infection.

Differential diagnoses such as hemolytic uremic syndrome (HUS) or thrombotic thrombocyto-penic purpura (TTP) were rendered unlikely through the absence of hemolysis and fragmentocytes. Similarly, urine microscopy without acanthocytes argued against acute glomerulonephritis due to systemic vasculitis.

Directly after first otorhinolaryngology consultation antibiotic therapy with clindamycin had already been administered. After extended laboratory results with high procalcitonin values and matching differential blood count were available the antibiotic regimen was extended by combination therapy with piperacillin, tazobactam and vancomycine.

As the focus of the severe systemic infection remained uncertain a contrast-enhanced computer tomography (CT) from neck to pelvis was performed. It confirmed primary infection of the pharynx with two small parapharyngeal submucous abscesses of each 9 x 6 mm, directly neighboring thrombosed veins extending into and occluding the left internal jugular vein (Figure 1). Further septembolic dissemination presenting by bilateral lower lobe pneumonia, inflammatory affection of intestine and colon as well as cervical and mesenterial lymphadenopathy were detected.

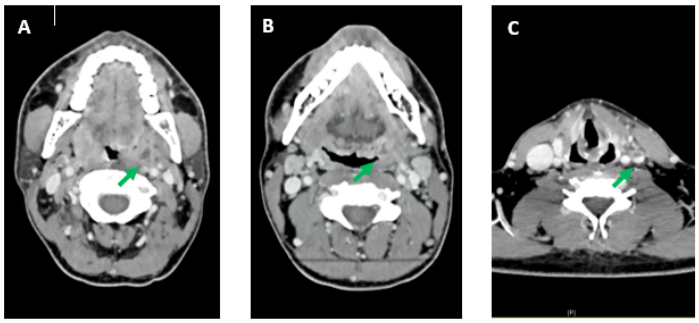

Figure 1: Contrast-enhanced computed tomography demonstrating parapharyngeal abscesses (contrast enhanced and thickened wall) (A, B) and a lack of contrast medium in the left internal jugular vein (C).

Findings of postanginal abscesses, thrombosis of the internal jugular vein, and septic dissemination thus led to the diagnosis of Lemierre's syndrome. Later blood as well as intraoperative cultures revealed Fusobacterium necrophorum as causative infective organism.

The patient was immediately transferred to surgical tonsillectomy for the left-sided abscess formations. Due to the detection of internal jugular vein thrombosis a PTT-controlled continuous anticoagulation with heparin was initiated.

Follow-up and outcome

After surgery, the patient was transferred to the intensive care unit and received antibiotic triple treatment along with continuous therapeutic anticoagulation. According to the Fusobacterium necrophorum detection in blood cultures, the antibiotic treatment was deescalated to monotherapy with piperacillin and tazobactam after four days and continued for a further 10-day period. After one week of therapy, the patient's condition warranted transfer to the general ward. Anticoagulation was switched to apixaban. Throughout the entire clinical stay, the patient experienced no complications and could be discharged after 14 days of hospital stay. Subsequent follow-up appointments revealed no long-term impairment despite the persistence of the internal jugular vein thrombosis.

Discussion

Lemierre's syndrome is a rare, but severe complication of

common pharyngeal infections characterized by septic metastasis and internal jugular vein thrombosis due to Fusobacterium necrophorum septicemia [2,3]. Most cases have been described in adolescents and young adults without severe comorbidities thus showing the high pathogenic potential of Fusobacterium necrophorum [6].

Clinical manifestation may also include other primary infection sites (e.g. lower respiratory tract), thrombotic vessels (e.g. cavernous sinus) and different bacterial species may be responsible for septicaemia (e.g. Fusobacterium spp., Streptococcus spp. or Staphylococcus spp.) [2,5,6].

Although the mortality rates dropped from 90% at the time of first description [1] to below 10% in more recent years [2,3], it remains a life-threatening disease due to sepsis syndrome and often delayed diagnosis.

The delay in timecritical diagnosis of this syndrome is mainly due to the predominant occurrence in healthy and immunocompetent young individuals and its rare incidence in relation to the frequency of common pharyngeal infections. Lemierre's syndrome thus represents a challenging diagnosis. As seen in the present case there is a high risk of underestimating the patients' life-threatening condition due to mild symptomatology. However, the transition from a localized pharyngeal infection to severe bacteremia can occur rapidly within days. This poses a high risk for fatal outcomes of sepsis syndromes, especially if out-of-hospital care is thought to be sufficient as seen in this case. Furthermore, clinical sepsis scores such as MEWS or NEWS commonly used as screening tools in the ED almost failed to alert the clinicians in this case [7]. A solution to detect SIRS or sepsis syndromes in ED patients as soon as possible might be the use of the so-called SOFA score which also integrates laboratory results. Even if initial screening is negative, application of this SOFA score as soon as laboratory results are available might increase sensitivity.

Definitive diagnosis of Lemierre's syndrome is usually confirmed by blood cultures positive for Fusobacterium necrophorum. However, as anaerobic blood culture results may require multiple days, microbiological testing of blood cultures may lead to delayed diagnosis as well. Only in a minority of cases Lemierre's syndrome is confirmed radiographically [3,8]. In the presented case the CT scan revealed two small fluid collections and contrast media recess within the internal jugular vein. These findings previously undectected by clinical and endoscopic examinations in this case represent typical abscess formations and internal jugular vein thrombosis in Lemierre's syndrome [9] and led to the final diagnosis.

Thus, only the combination of patient's history, highly elevated PCT levels and CT scan findings allowed for the detection of systemic infection and correct diagnosis of severe bacteremia in this case. For rapid detection and diagnosis of potentially life-threatening early stage infectious diseases high resources and interdisciplinary cooperation are thus required, especially in case of rare etiologies.

Conclusions

Lemierre's syndrome is a rare cause of sepsis particularly affecting young immunocompetent patients. Due to mild symptomatology in early stages of bacteremia it poses a high risk of undertriage and treatment delay. Prolonged throat soreness in combination with slightest signs of systemic inflammation should follow further thorough diagnostic investigations for this syndrome in case of pharyngeal infections. After having lost its title "forgotten disease" Lemierre's syndrome thus needs to gain awareness among clinicians to optimize outcomes and reduce mortality.

Ethical Guidelines: The patient gave written informed consent for the case report.

Conflicts of Interest: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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