

Lemierre's Syndrome: A Rare Clinical Condition Diagnosed, Exceptionally, by Imaging

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In 1936 Lemierre described a then invariably fatal syndrome consisting of internal jugular vein thrombosis, septicemia and metastatic lung infections following oropharyngeal infection [1]. Today, the widespread use of antibiotics in upper respiratory tract infections has led to a decrease in the incidence of the syndrome [2] but also in a failure to sometimes recognize it, and fatalities continue to be reported [3,4]. Traditionally, the diagnosis has been based on a typical clinical picture coupled with laboratory evidence of an anaerobic septicemia and evidence pointing to internal jugular venous thrombosis. Nowadays an implicative clinical history and examination with unequivocal imaging findings probably suffice. We report here a case of the condition.

Patient Description

A 20 year old usually healthy male consulted his family physician after 3 days of sore throat and fever. On examination his tonsils were bilaterally inflamed and covered with exudate. The cervical lymph nodes were also enlarged but physical examination was otherwise unremarkable. A clinical diagnosis of bacterial pharyngitis was made and the patient received 2.4 million units of benzathine penicillin G intramuscularly followed by thrice daily oral 500 mg amoxycylav.

Nevertheless, his condition continued to deteriorate with the development of dyspnea and, self-referred to a hospital emergency room, he was admitted to the otolaryngology department with a diagnosis of peritonsillar abscess. No chest X-ray

was performed at this stage. Despite treatment with intravenous penicillin no improvement occurred over the next 72 hours, by which time the symptoms increased to include chest pain and a dry cough with bloody mucous. At this stage the patient's blood pressure was 115/70 mmHg, pulse regular at 100/minutes, temperature 38.4°C, oxygen saturation 94% on breathing room air, and respiratory rate 20/min with reduced breath sounds over the right lung. The heart sounds were normal with no murmurs. The abdomen was soft, although the liver was palpable 2 cm below the costal margin and a spleen tip could be felt.

Laboratory investigation revealed hemoglobin 9.5 g/dl, white blood cell count 14,000/mm³, erythrocyte sedimentation rate 88 in the first hour, and total bilirubin 1.7 mg/dl. Blood samples were sent for laboratory culture but were reported as sterile, presumably due to previous antibiotic therapy. However, chest radiology revealed disseminated findings in both lungs with infiltrates in the lower lobe of

the right lung [Figure]. The clinical diagnoses under consideration included infectious mononucleosis, secondary pneumonia, cytomegalovirus, human papilloma virus and human immunodeficiency virus infection, but all relevant serologic investigations were negative. Trans-thoracic and trans-esophageal echocardiography to exclude bacterial endocarditis were normal, as was a bone scan performed to exclude osteomyelitis. To cover the possibility of an atypical pneumonia, amoxycylav and roxithromycin were substituted for penicillin on the 11th day of the illness. Computerized tomography of the chest revealed a large right-sided pleural effusion, multiple infiltrates in both lungs and nodular markings with evidence of cavitation, compatible with septic emboli [1,3] or vasculitis. To quantify the cervical lymphadenopathy a cervical CT was also performed. This delineated thrombophlebitis in a branch of the left internal jugular vein, which finally confirmed the illness as Lemierre's syndrome.

On the 15th day of the illness the patient began to improve clinically, with the chest pain and dyspnea subsiding and the white blood cells falling to 10,000/mm³. The fever, however, remained elevated. As this was thought to be related to the high amoxycylav dosage, intravenous clindamycin was substituted and the fever promptly fell. Concurrent repeat cervical and thoracic CT examination showed the thrombophlebitis to be resolving along with progressive clearing of the lung findings. The patient was discharged home on continued antibiotic treatment with clindamycin for a further 3 weeks as recommended in previous reports [3,5]. Subsequent follow-up was uneventful.



Chest X-ray showing disseminated findings in both lungs and infiltrates in the lower lobe of the right lung.

Comment

Lemierre's syndrome is usually seen in previously healthy young individuals infected with the Gram-negative obligate anaerobic bacterium *Fusobacterium necroforum* following an episode of throat or dental infection [1]. The clinical manifestations are so characteristic that, according to Lemierre himself, "a mistake is almost impossible" [1]. Nevertheless, local evidence of pharyngitis may have disappeared by the time the full syndrome has developed. The development of multiple metastatic abscesses, most frequently involving the lungs and large joints, may lead to diagnostic confusion with right-sided endocarditis or commoner entities such as pneumonia, pulmonary embolism, osteomyelitis or an acute arthritis. Today, however, Lemierre's syndrome must still be considered in any previously healthy febrile young patient with acute oropharyngeal disease in whom pulmonary symptoms and the likelihood of internal jugular vein thrombophlebitis arise [1]. Suspicion of the latter can be subsequently verified by appropriate imaging studies even when, as in this case, proof of an anaerobic septic-

mia is lacking. Chest radiology should also be performed to detect septic emboli or other pulmonary pathology [3].

As indicated, *F. necroforum* can usually, but not invariably, be cultured from the blood and metastatic sites of infection [5]. However, because of its relatively long 4 day incubation period, antimicrobial therapy should not be withheld pending blood culture results or more definitive localizing signs. Such therapy should initially comprise antibiotics with an effective anti-anaerobic spectrum of activity such as penicillin, clindamycin, metronidazole and chloramphenicol, and prolonged high dose treatment is recommended in view of the endovascular nature of the syndrome. Ligation and resection of the thrombosed internal jugular vein is usually unnecessary and should only be considered in patients with refractory sepsis or severe respiratory compromise from repeated pulmonary emboli. The use of anticoagulation remains controversial and no controlled trials have been performed on its efficacy in septic thrombosis [3].

Rare as it is in the post-antimicrobial era, the possibility of Lemierre's syndrome

is often overlooked by the physician. Thus, the overall mortality of the syndrome is still about 15%, which may rise to 80% among patients who fail to receive appropriate and timely antibiotic therapy [5].

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