# Isolated Facial Vein Thrombophlebitis: A Variant of Lemierre Syndrome

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**Lemierre syndrome is a rare complication of acute tonsillitis. It is caused by the anaerobic bacterium *Fusobacterium necrophorum* and is characterized by bacteremia and septic thrombosis of the internal jugular vein. Dissemination of septic emboli may occur. The diagnosis can be difficult since different organs can be involved. We discuss a case of Lemierre syndrome in a 35-year-old woman with isolated thrombophlebitis of the facial vein and fusobacteria growth in blood culture. This case emphasizes the need for awareness of the condition.**

***Keywords.*** Lemierre syndrome; facial vein thrombosis; *Fusobacterium necrophorum*; anaerobic postanginal sepsis.

Lemierre syndrome (LS) is a rare complication of acute tonsillitis, which often affects young adults. In Denmark, the incidence is 14.4/1 000 000 [1]. The syndrome is caused by a pharyngitis with the anaerobic bacterium *Fusobacterium necrophorum* (FN) and is characterized by bacteremia and septic embolism of the internal jugular vein (IJV). Dissemination of septic emboli can occur, most commonly pulmonary, as well as peritonsillar, parapharyngeal, and pulmonary abscess [1–7].

Received 18 April 2014; accepted 01 June 2014.

The case-report must be published from Department of Otorhinolaryngology, Holstebro Hospital, Holstebro, Denmark.

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**Open Forum Infectious Diseases**



DOI: 10.1093/ofid/ofu053

The clinical picture varies between the clinical relatively unaffected patient and the septic patient with multiple organ failure. The diagnosis can be difficult. Mortality is 6%–22%, [4, 7] which underlines the importance of early diagnosis and proper treatment. Most patients are initially admitted to an internal medical department (79%), [1, 7] for which reason knowledge of the syndrome is important for physicians who treats emergency patients. We present a history of atypical clinical presentation, which emphasizes the need for awareness of the condition.

## CASE

A previously healthy 35-year-old woman was admitted to the internal medicine department with a history of 6 days of sore throat and increasing poor general condition. On day 2, she was seen by her general practitioner. At that time, the rapid streptococcal test was negative, thus no antibiotics had been prescribed. On day 3, her symptoms progressed, and she developed a tender left swelling of her face and neck.

When she was admitted to a tertiary care center, her temperature was 37.6, the blood pressure was 90/46 mmHg, the heart rate was 103 beats per min, respiratory rate was 30 breaths per min, and oxygen saturation of 95% on room air. Laboratory tests showed increased C-reactive protein (CRP; 185 mg/L) and leukocytosis (17.9 × 109/L). The streptococcus A antigen rapid test was positive. Antibiotic treatment was started using intravenous (IV) penicillin and metronidazole. The patient was transferred to our ear, nose, and throat department. The physical examination showed left facial swelling (Figure 1). The patient had trismus, the tonsils were slightly equally enlarged, and there was pain on palpation of the neck, but otherwise there were no signs of peritonsillar abscess or sialadenitis. She had bilateral cervical lymphadenopathy. On the second day of hospitalization and despite antibiotics, the symptoms progressed; just like the facial swelling increased, CRP increased to 229 mg/L and leukocytes to 26 × 109/L.

Contrast-enhanced computed tomography (CT) scan of the neck and sinuses showed isolated thrombosis of the left facial vein (Figure 1) and bilateral pulmonary infiltrates. The patient started to receive anticoagulation treatment (tinzaparin 13.500IE). By day 8, the

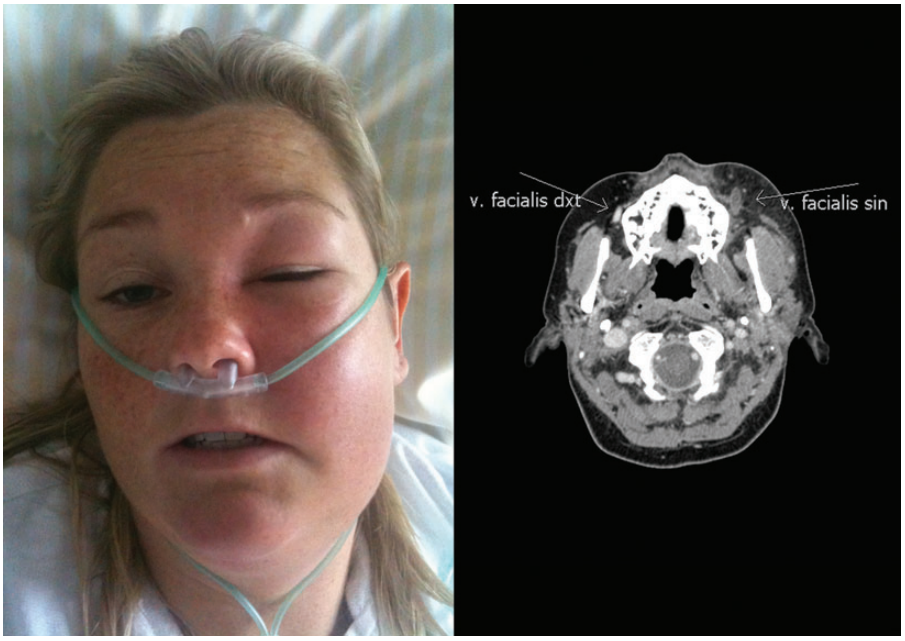


**Figure 1.** Facial swelling on the presentation of the patient. Contrast-enhanced CT-scan of head and neck demonstration the occluded facial vein on the left side and normal facial vein on the right side. Abbreviation: CT, computed tomography. Reproduced with permission from the patient.

blood culture grew positive for FN. The combination of pharyngitis, CT demonstration of facial vein thrombosis, and FN sepsis confirmed the diagnosis of LS. After conference with the microbiological department the antibiotics were changed to penicillin and clindamycin. The patient was transferred to the infectious diseases ward, where she received IV antibiotics for a month and was discharged after 3 months oral anticoagulant therapy (warfarin).

## DISCUSSION

In 1936, André Lemierre described the clinical findings of "anaerobic postanginal sepsis" today known as LS [6].

Previously known as "the forgotten disease," [4, 5, 7] the incidence of LS cases is reported to be increasing in recent years, and this is probably due to a combination of more restricted use of antibiotics to treat pharyngitis, but it may also be an indication of publication bias or increased awareness of the disease [4, 7]. There is a need for increased awareness of the common sore throat with protracted course in the form of lack of response to treatment or complications of the disease. The exact pathogenetic mechanism remains unexplained but is caused by several factors including hemolysin, lipase, and leukotoxin [1, 8]. FN can activate the intrinsic pathway of coagulation that leads to the thrombocytopenia and disseminated intravascular coagulation [8, 9]. Extension of the infection to the cervical region results in thrombosis of the internal jugular vein (IJV) or one of it tributaries, hence explaining the isolated facial vein thrombosis in our case [8, 10]. The LS diagnosis is made clinically and by detection of the FN in blood culture, but it should also be taken into account in case of negative blood culture results [1]. Occasionally cases of LS have been associated with other bacteria than FN, for example, *Streptococcus pyrogenes, Klebsiella pneumonie, Proteus* [4, 7, 11]. Contrast-enhanced CT of the neck and thorax can detect thrombosis of IJV and other complications, for example, pulmonary septic emboli, parapharyngeal- and lung abscesses [7, 8]. FN is often associated with LS, but more often it causes isolated infection in the form of peritonsillar abscess and is perhaps the most frequent pathogen in PTA patients [2]. Some FN strains can be β-lactamase-producing and the recommended treatment is IV penicillin and metronidazole for 2 weeks, followed by oral treatment until the infection parameters have been normalized for at least 2 weeks [1, 7]. The role of therapeutic anticoagulation in LS remains controversial due to the lack of clinical trials [1, 4, 5, 7, 12]. Surgical treatment may be necessary, for example, drainage of empyema or abscess [1, 2, 4, 7]. Surgery for LS previously included ligation of IJV; now this procedure is reserved for patients with persistence of septic emboli, despite appropriate antibiotic treatment [7, 8, 13, 14]. Isolated thrombosis of the facial vein was previously described by Lemierre but otherwise only a few times in the literature [3, 5, 6]. This case underlines the difficulty of the diagnosis and the clinical variance.

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