



# Remembering the Forgotten Disease

Francis Phan MD, Melissa Rae LeBlanc MD, Alan J. Hunter, MD, FACP  
Oregon Health & Science University, Portland OR

## Introduction

- Lemierre syndrome, or jugular vein suppurative thrombophlebitis, was common before the discovery of antibiotics. <sup>(1)</sup>
- The incidence of Lemierre syndrome decreased after use of penicillin and was nearly forgotten for 30 years, hence, also referred to as the forgotten disease. <sup>(1)</sup>
- Although still rare overall, the incidence is now starting to rise. <sup>(2)</sup>

## Case Description

- A healthy 36-year-old woman is evaluated at urgent care for acute pharyngitis and dyspnea then sent home with supportive treatment.
- Symptoms progress over a week to include right neck pain, fever and chills.
- Presented to ED with respiratory distress, found to be febrile, hypoxic with leukocytosis and thrombocytopenia.
- CXR notable for RLL airspace disease, CT neck w/ non-occlusive in R internal jugular vein (Figure A).
- Admitted to MICU for hypoxic respiratory failure requiring intubation, broad spectrum antibiotics and anticoagulation started.
- Blood cultures grew *Fusobacterium necrophorum*.

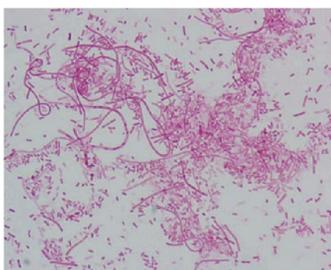


Figure E. *Fusobacterium necrophorum*. Staining negative on gram stain. Long & short rods that curl & tangle. (3)

- Developed pleural effusions requiring bilateral chest tubes (Figure C).
- Fevers persisted despite clinical improvement, CT C/A/P negative for occult abscess.
- Discharged to rehab after antibiotics course. Represented 3 weeks later with severe lower back pain found to have L4-S1 osteomyelitis w/ epidural phlegmon on MRI (Figure D).
- After extended antibiotic course she recovers and returns to work.

## Imaging

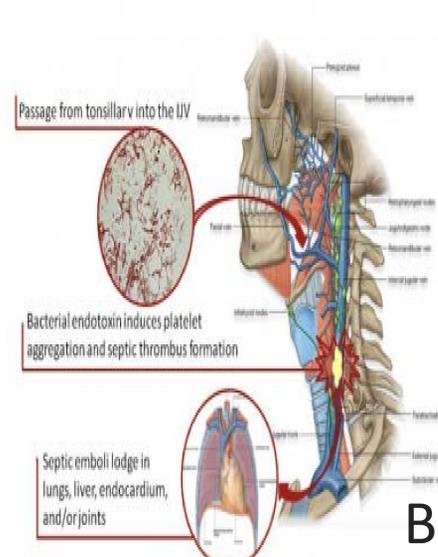


Figure A: CT Neck showing non occlusive Right IJ Thrombus. Figure B: Graphic demonstrating pathogenesis of bacteria induced thrombus formation <sup>(4)</sup>

## Lemierre Syndrome Illness Script

### Presentation

- Young healthy adolescents
- High fever, Chills, Sore throat, neck pain
- Respiratory distress

### Diagnostics

- Internal jugular Vein thrombophlebitis on CT Neck w/ Contrast
- Anaerobic bacteremia (e.g. *Fusobacterium spp*)
- Septic emboli

### Management

- Empiric Antibiotics
- Source control
- Anticoagulation (Debatable)

## Imaging



Figure C: CT Chest showing R empyema from septic emboli. Figure D: MRI showing L5 osteomyelitis and adjacent epidural phlegmon

## Discussion

- Lemierre Syndrome (also termed **The Forgotten Disease**), is defined as post-pharyngitic suppurative jugular vein thrombophlebitis, with metastatic spread of disease (bone, lung, brain, joints)(Figure B).
- Up to 80% of cases are caused by the gram negative anaerobic bacteria *Fusobacterium necrophorum*, with the remainder caused by a variety of organisms. <sup>(5)</sup>
- *Fusobacterium spp.* are common microbiologic isolates in non-streptococcal pharyngitis and peritonsillar abscesses, accounting for about 20% of community onset pharyngitis, (Table 2), and result in more morbidity & mortality (**up to 5%**) than streptococcal pharyngitis<sup>(5)</sup>

Table 2. Summary of Bacterial Species Identified in Patients and Asymptomatic Students\*

Species	Patients (n = 312)	Asymptomatic Students (n = 180)
<i>Fusobacterium necrophorum</i>	64 (20.5)	17 (9.4)
<i>F. necrophorum</i> alone	47 (15.1)	16 (8.9)
<i>F. necrophorum</i> and GAS	9 (2.9)	-
<i>F. necrophorum</i> and GCS/GGS	7 (2.2)	1 (0.6)
<i>F. necrophorum</i> , GAS, and <i>Mycoplasma pneumoniae</i>	1 (0.3)	-

- Lemierre syndrome should be suspected in young healthy adults with preceding pharyngitis that rapidly progresses to fever, rigors, chills, neck pain and respiratory distress.
- Additionally counseling in 'return precautions' should focus on these symptoms.
- The incidence of this "forgotten disease" may continue to rise in the era of increasing antimicrobial resistance and stewardship. <sup>(1)(2)</sup>

## Teaching Points

- In the era of increasing antibiotic stewardship clinicians should be vigilant about the potential of developing Lemierre syndrome.
- Remember **The Forgotten Disease**, a morbid & mortal complication of pharyngitis in young healthy individuals with rapidly progressive systemic syndromes preceded by upper respiratory symptoms.

## References

1. Riordan T. Human infection with *Fusobacterium necrophorum* (Necrobacillosis), with a focus on Lemierre's syndrome. Clin Microbiol Rev. 2007;20(4):622-59.
2. Karkos PD, Asrani S, Karkos CD, et al. Lemierre's syndrome: A systematic review. Laryngoscope. 2009;119(8):1552-9.
3. Bank S, Nielsen HM, Hoyer Mathiasen B, Christiansen Leth D, Hagelskjær Kristensen L, Prag J. *Fusobacterium necrophorum* - detection and identification on a selective agar. APMIS 2010; 118: 994-9
4. <https://pedclerk.bsd.uchicago.edu/page/lemierre%E2%80%99s-syndrome>
5. Centor RM, Atkinson TP, Ratliff AE, Xiao L, Crabb DM, Estrada CA, et al. The Clinical Presentation of *Fusobacterium*-Positive and Streptococcal-Positive Pharyngitis in a University Health Clinic: A Cross-sectional Study. Ann Intern Med. 2015;162 (4):241-247. doi: 10.7326/M14-1305