CASE REPORT

A Lemierre syndrome variant caused by *Klebsiella pneumoniae*

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Lemierre syndrome is an extremely rare disease characterized by oropharyngeal infection, septicemia, internal jugular vein thrombosis, and skip lesions. The most common causative pathogen is *Fusobacterium necrophorum*. We reported a 45-year-old woman who presented with left neck painful swelling and septicemia. Magnetic resonance imaging of the head and neck demonstrated venous thrombosis extending from the left internal jugular vein to the sigmoid sinus. During admission we discovered that the patient had uncontrolled diabetes mellitus. We also found a metastatic lesion through chest radiography. *Klebsiella pneumoniae* was cultivated from both blood samples and pus from deep neck spaces. Surgical drainage, early and adequate antibiotic treatment, anticoagulation, and strict control of blood glucose led to the patient’s complete recovery. Because Lemierre syndrome is a forgotten disease in the era of antibiotics, awareness of the signs and symptoms of this disease is important because of its associated high mortality rate. This case illustrated that the presence of *K pneumoniae* can lead to Lemierre syndrome.

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**Introduction**

Lemierre syndrome is a rare but fatal disease with an incidence rate between 0.6 and 2.3 per million people and a mortality rate ranging from 4% to 18%. It was first described in 1900 by Courmant Cade and in 1936 was named after the French microbiologist Andre Lemierre, who reported 20 cases; 18 patients died. Lemierre syndrome is characterized...
by septicemia documented by at least one positive blood culture, with primary infection in the oropharynx and clinical or radiographic evidence of internal jugular vein thrombosis. Other metastatic foci and embolization at various sites may also be found, especially in the lung.

The "classic" pathogen of Lemierre syndrome is *Fusobacterium necrophorum*, a normal flora of oropharynx that only becomes pathogenic when normal host defense mechanisms do not function. Other microorganisms have also been implicated alone or in combination with *F. necrophorum*. We present what we believe is the first reported case of a variant of Lemierre syndrome originating from *Klebsiella pneumoniae* infection.

**Case report**

A 45-year-old woman was hospitalized due to fever and chills for 3 days. Other symptoms at admission were sore throat, odynophagia, dysphagia, productive cough, and painful left neck swelling. She denied any history of dental procedure, oral ulcer, or upper airway infection in the previous 1 month. Computed tomography of the head and neck with and without contrast was performed. Results revealed left neck cellulitic change and fluid accumulation over the retropharyngeal space, an enlarged lymph node with central abscess formation between the left sternocleidomastoid muscle and the ramus of the mandible, and thrombosis of the left internal jugular vein (Fig. 1). Painful swelling of the neck and fever persisted even after 5 days of antimicrobial therapy with gentamycin and 2 days with ceftriaxone. The patient was transferred to our hospital at her family's request. The white blood cell count was 11,200/μL at admission with levels of the inflammatory marker C-reactive protein exceeding normal limits (11 ng/mL). Blood culture showed *K. pneumoniae* as the only infectious agent. Fiberoptic laryngoscopy showed bulging of the left lateral pharyngeal wall extending from the nasopharynx down to the hypopharynx, but the airway was patent. A suspected hazy density lesion in the right upper lung was noted on chest radiography (Fig. 2). Laboratory data revealed glucose 206 mg/dL, HbA1c 11.6%, hypokalemia (K 2.3 meq/L), hyponatremia (Na 131 meq/L) and thrombocytopenia (Platelet 94,000/μL). Incision and drainage were performed with caution on the second day of admission. The parapharyngeal, retropharyngeal, and parotid spaces were opened by drainage of profuse amounts of pus. Two Penrose drains were inserted in the parapharyngeal and retropharyngeal spaces, respectively. The patient was transferred to the intensive care unit for additional care after the surgery. A pus culture grew *K. pneumoniae*, which was in agreement with the blood culture result. Postoperative magnetic resonance imaging of the head and neck showed that the left internal jugular vein thrombosis extended from the clavicle to the left sigmoid sinus (Fig. 3), which might explain the persistent symptom of left unilateral headache. Flomoxef was administered intravenously for infection and low-molecular-weight heparin was added for the intravenous thrombosis. Meanwhile, blood glucose was strictly controlled in range between 100 to 150 mg/dL. On the 10th day of hospitalization, follow-up laboratory data showed a white blood cell count of 9500/μL and a C-reactive protein level of 0.897 mg/dL. The hazy density of the right upper lung appeared to have regressed on the follow-up chest radiograph. The patient improved with treatment and was sent home 15 days after admission. The patient has had regular follow-up visits with no recurrence or other complications noted.

**Discussion**

Over the past 25 years, Lemierre syndrome has only been reported in approximately 150 cases, partly due to antibiotic...
therapy preventing disease progression and a general unawareness of the syndrome. The term “forgotten disease” has been used to describe the rarity of Lemierre syndrome in the antibiotic era. It typically occurs in adolescents and young adults, although in our case the patient was 45 years old.

_F. necrophorum_ is the most commonly reported causative pathogen. Other variant causes include infection with group A streptococci, _Streptococcus pyogenes_, and _Leptotrichia buccalis_. Virus infection may also serve as a predisposing factor, including Epstein-Barr virus or cytomegalovirus infection. In the current case, _K. pneumoniae_ was identified in both blood culture and pus culture, adding to the diversity of causative agents.

In the current case, the disease progressed rapidly from the first stage of primary infection. Unspecific symptoms included fever, rigor, dyspnea, sore throat, cough, swollen neck, or gastrointestinal problems and local invasion of the lateral pharyngeal space and internal jugular vein septic thrombophlebitis via direct extension through the fascia plane between the tonsils and the parapharyngeal space by hematogenous or lymphatic space spread from peritonsillar vessels. Metastasis is the final stage of this disease.

Internal jugular vein thrombosis is the major complication of Lemierre syndrome. It has been reported that Horner syndrome can be caused by jugular vein thrombosis. In our case, thrombosis from the clavicle through the internal jugular vein to the sigmoid sinus was noted. The patient only presented with headache and dizziness, without other neurologic symptoms.

The most common sites of metastasis are the lungs and joints. In the current case, pulmonary abscess formation was noted in the right upper lung. The patient presented with productive cough. Sputum culture identified _Pseudomonas aeruginosa_, which was different from the pus and blood culture results. In our review of cases, only coinfection with _Mycoplasma pneumoniae_ has ever been reported.

There are also hematologic complications of Lemierre syndrome, such as thrombocytopenia and disseminated intravascular coagulation. The patient’s platelet count was only 94,000/µL at admission. Hypokalemia, which has been reported, was also present in this case.

The treatment of Lemierre syndrome must include antibioteic therapy. Three to 6 weeks of antimicrobial treatment is usually recommended. Anticoagulation therapy is more controversial due to the risk of hemorrhage and extension of the disease. Surgical ligation of the internal jugular vein was the only recourse for thrombosis during the preantibiotic era, but is currently reserved for use with uncontrolled sepsis or ongoing evidence of septic emboli. In our experience, with the exception of the antibiotic treatment, the strict blood glucose control and timely surgical drainage, especially to the diabetic patient with Lemierre syndrome, is critical.

References


Figure 3  Axial T2 Weighted MRI of the head. The white arrow indicates the sigmoid sinus thrombosis.