Lemierre’s syndrome: case of a patient with pulmonary embolism and cavernous sinus thrombosis complicating a septic internal jugular vein thrombus

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ABSTRACT
Lemierre’s syndrome is a complex and unusual clinical entity, characterized by septic thrombophlebitis of the internal jugular vein. We present the case of a patient with Lemierre’s syndrome, pulmonary embolism and propagation of an internal jugular vein thrombus retrograde to the cavernous sinus. The patient was treated with antibiotics and heparin. The importance of rapid diagnosis of the extent of the disease, in directing the otherwise somewhat controversial treatment with heparin in patients with Lemierre’s syndrome, is stressed.

Key words: Lemierre’s syndrome, internal jugular vein, septic venous thrombosis, anticoagulation treatment

Introduction
In 1936, Andre Lemierre described 20 cases of internal jugular vein thrombophlebitis (IJVT), with postanginal sepsis that would later become known as Lemierre’s syndrome. (1) Septicemia was mostly associated with anaerobic oropharyngeal infection extending to the neck, with disseminated (mostly pulmonary) abscesses. (1,2) The mortality rate in Lemierre’s series was 90%. (1) The syndrome is usually characterized by a history of recent oropharyngeal infection, clinical or radiological evidence of IJVT, including remote septic emboli, and isolation of anaerobic pathogens, mainly Fusobacterium necrophorum, although other pathogens have been reported. (2-7) Reports of Lemierre’s syndrome significantly declined after the widespread use of antibiotics and it was thought to be a rare and forgotten disease with a suggested incidence of approximately one per million. (5) However, an increase in frequency over the past years is thought to be due to changes in antibiotic usage. (8) Lemierre’s syndrome occurs mainly in previously healthy children and young adults, although it spares no age group. (3,6) We present a case in which an adult patient suffered from Lemierre’s syndrome as a complication of an untreated oropharyngeal infection.

Case report
A 56-year old previously healthy male patient presented to our emergency department with a 5-day history of fever and one day history of a swollen neck. The patient had been in good health until 5 days earlier, when he began to have a sore throat and fever. He reported that he was examined by his physician who told him that it was probably a viral infection. On the sixth day he developed painful swelling in the right anterior side of his neck. He reported no previous history of trauma, malignant disease, infection, operation or IV catheterization, and had never consumed tobacco or alcohol.

Physical examination revealed a non-tender, soft and immobile mass in the right anterior side of his neck. The mass was not associated with hyperemia or local heat. Oropharyngeal examination was significant for pharyngeal erythema with no exudates. Examination of other systems was normal. Ultrasonography revealed a hyperechogenic mass around the right internal jugular vein (IJV), which was suggestive of a thrombus. Blood flow was not demonstrated by Doppler ultrasonography and the vein was not compressible. The patient was diagnosed with IJVT and admitted to hospital. Computed tomography (CT) images demonstrated complete thrombosis of the
right internal jugular vein, thrombosis of the cavernous sinus, multiple septic pulmonary emboli, and bilateral necrotizing pneumonia. His vital signs were: temperature 38.6°C, pulse 125 beats/min, respiration 20 breaths/min, blood pressure 100/80 mmHg, and room air pulse oximetry was 100%. Laboratory investigation revealed a white blood cell count of 18,600/µL; hemoglobin 12.8 g/dL; red blood cell count, 4.2x10⁶/mL; platelet count, 282x10³/mL; prothrombin time, 18.1; partial thromboplastin time (PTT), 46.5; international normalized ratio (INR), 1.45; C-reactive protein, 128 mg/L. Within one hour of the patient’s arrival he reported increased difficulty breathing. The patient underwent intubation and was admitted to the Intensive Care Unit with a diagnosis of Lemierre’s syndrome. Empirical antibiotic treatment was started with imipenem and metronidazole. Low molecular weight heparin in therapeutic doses was given. The blood culture obtained at the time of hospitalization revealed no organisms. A follow-up CT scan of the neck and chest showed a decrease in the size of thrombi. He was extubated on day 7 of hospitalization. He underwent six weeks of vancomycin therapy with good recovery. Anticoagulation treatment with a vitamin K antagonist was prolonged for 6 months.

Discussion

The two leading causes of IJVT are iatrogenic trauma secondary to jugular vein catheterization, and repeated IV injections by drug users. (9) Lemierre’s syndrome was thought to be a rare and forgotten disease with a suggested incidence of approximately one per million. (5) However, an increase in frequency over the past years has been attributed to changes in antibiotic usage. (8) Unfortunately, widespread antibiotic usage has also changed the clinical picture of Lemierre’s syndrome and it is often difficult to recognize this unusual illness in the Emergency Department (ED). (10) Systemic septic complications may range from deep neck infections over septic arthritis.

Figure 1. Transverse section through the neck (CTA). Arrow is denoting the thrombosed internal jugular vein with a perivascular area of inflammation.

Figure 2. Frontal section through the neck (CTA). Arrow is denoting thrombosed internal jugular vein with massive propagation of the thrombus in a cranial and caudal direction.
REFERENCES