ABSTRACT

Objectives:
1. To review the presenting symptoms and findings of classic Lemierre's syndrome, including radiological and bacteriological findings, and report an atypical presentation of Lemierre's syndrome in a patient with facial vein thrombosis.
2. To reinforce the need for accurate and timely diagnosis, as well as prompt initiation of treatment.

Methods:
The clinical presentation, physical examination, imaging features, surgical findings, bacteriological profile and pathology slides are reviewed in a case of an adolescent patient with a variant presentation of Lemierre's syndrome. The literature on Lemierre's syndrome was reviewed as was that of its typical findings.

Case Report:
A previously healthy 16 year old female presented with a 5 day history of worsening sore throat and fever along with unilateral jaw swelling. Examination revealed a subcutaneous tender cord like structure raising a suspicion for facial vein thrombophlebitis. Computed tomography (CT) scan of the neck with contrast confirmed thrombosis of the left facial vein with patent internal jugular veins. Similar studies of the chest identified multiple septic pulmonary emboli. Neck examination revealed septic thrombus formation within the lumen of the facial vein. Cultures were consistent with Fusobacterium species. Review of the literature has identified only a few such reported cases.

Conclusion:
Although classic Lemierre's syndrome is well described in the literature, it has been termed the “forgotten disease” because it is frequently overlooked and underreported. Prompt diagnosis of Lemierre's syndrome and its variants is essential in limiting morbidity and mortality related to this infectious disease.

CONTACT
Walid I. Dagher, M.D.
Department of Otolaryngology – Head and Neck Surgery
Tufts Medical Center
Boston, MA
wdagher@tuftsmedicalcenter.org

INTRODUCTION

Lemierre's syndrome, also known as postanginal sepsis or human necrobacillosis, is a severe complication of an acute oropharyngeal infection that results in septic thrombophlebitis of the ipsilateral internal jugular vein (IJV) followed by sepsis. (Ref) The condition is frequently complicated by metastatic infection originating from microbes within the mouth or pharynx. The most common causative organism is Fusobacterium necrophorum, a commensal organism of the oral cavity.

After the widespread introduction of antibiotic therapy, largely with penicillin in the 1960s and 1970s for oropharyngeal infections, Lemierre's syndrome was referred to as the “forgotten disease.” Today, with the decreased use of antibiotics for oropharyngeal infections and the increased numbers of antibiotic-resistant organisms, the disease has re-emerged. Andre Lemierre, a French microbiologist, was the first to fully describe the syndrome in 1936 reporting 20 cases. The mortality rate of Lemierre's syndrome was 90% in the pre-antibiotic era. (Ref) We describe a rare case of atypical Lemierre's syndrome, which required surgical intervention, and presented as thrombophlebitis of the facial vein.

CASE REPORT

A previously healthy 16-year-old girl was evaluated for a 5-day history of odynophagia and malaise. She had also developed progressive, tender left-sided facial edema. On presentation temperature was 105°F with rigors. Oropharyngeal exam was notable for left sided tonsillar erythema without trismus or palatal fullness. Neck examination revealed a left tender cord-like structure extending from the level of the submandibular gland to the angle of the mandible superiorly.

A computed tomography (CT) scan of the neck with contrast revealed findings consistent with facial vein thrombophlebitis, including a filling defect in the left facial vein (FIGURE 1). The patient was started on Vancomycin and Zosyn empirically. Despite antibiotic therapy, the patient developed hypotension and an increased oxygen requirement. Her condition worsened and she was intubated and mechanical ventilation was initiated.

A CT scan of the chest with contrast showed multiple pulmonary lesions consistent with bilateral septic emboli. The patient was persistently hypotensive requiring vasopressors and the decision was made to surgically ligate the facial vein. Intraoperatively, the submandibular gland appeared slightly inflamed, but the focus of inflammation was confined to the left facial vein, which was white in appearance. A transition point was identified inferiorly and the vessel was ligated just below this point to prevent further clot propagation. Intravascular purulent fluid was sent for microbiology and the thrombotic facial vein was resected (FIGURE 2) and sent for pathology (FIGURE 3). Blood cultures grew Fusobacterium necrophorum and tissue cultures grew Streptococcus viridans.

The patient was maintained on intravenous antibiotics, exubated on postoperative day 1 and discharged home on intravenous Zosyn/Metronidaizole for 4 weeks on postoperative day 4.

FIGURES

FIGURE 1: Computed tomography scan of the neck with contrast (axial and coronal cuts) showing a filling defect in the left facial vein (arrow) with thickening and enhancement of the vessel wall. There is surrounding soft tissue inflammation. Note patency of the internal jugular veins.

FIGURE 2: Intraoperative findings of purulence filling the left facial vein, which has been ligated inferiorly. Thickening of the vessel wall seen in cross section (arrow)

FIGURE 3: H&E stain 2X showing the vein lumen with a thrombus consisting of necrotic debris and chronic inflammation.

DISCUSSION

Lemierre's syndrome is a very rare disease and usually affects young healthy adults. The incidence has been reported between 0.6 and 2.3 per million, with mortality rates between 4% and 18%. Clinically, the progression of symptoms occurs in 3 stages:

The primary infection includes exudative tonsillitis, oropharyngeal ulcers, cervical lymphadenopathy, and pharyngeal hyperemia. Fever is present in 80% of patients.

The second stage involves invasion of the lateral pharyngeal space.

The third stage consists of development of metastatic emboli. Embolic disease in the lungs, the most common site of metastatic spread, leads to symptoms resembling an aseptic pulmonary embolism. Frank respiratory failure can occur and has been reported as high as 15%.

A high degree of clinical suspicion is required for accurate diagnosis. Criteria that are accepted as strong evidence for the presence of Lemierre's syndrome include an anaerobic primary infection in the oropharynx (Fusobacterium necrophorum), subsequent sepsis (with at least one positive blood culture), metastatic infection at one or more distant sites, and thrombophlebitis of the IJV.

An interdisciplinary approach is necessary to treat patients with Lemierre's syndrome. Treatment involves use of appropriate antibiotics (ceftriaxone and metronidazole) and, in many cases, surgical drainage.

Ligation or resection of the involved vein is only indicated in cases of uncontrolled sepsis or ongoing septic emboli despite antibiotic therapy. The routine use of anticoagulation in Lemierre's syndrome has not been recommended because of limited data.

CONCLUSION

Lemierre's syndrome is a serious complication of head and neck infections, initially involving the oropharyngeal space and ultimately leading to severe systemic compromise. Although a rare clinical entity today, Lemierre's syndrome remains a disease of considerable morbidity and potential mortality. Timely recognition of disease progression is crucial in preventing severe systemic manifestations. Empirical broad-spectrum antibiotic treatment should not be delayed and should include a third generation cephalosporin. Surgery is reserved for cases of uncontrolled sepsis or ongoing septic emboli despite antibiotic therapy.

REFERENCES