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Lemierre's Syndrome

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ABSTRACT

Lemierre's syndrome is a very rare but potentially fatal condition characterized by suppurative thrombophlebitis of the internal jugular vein (inner vieins of neck region), often following the recent oropharyngeal infestation. It involves bacteraemia and can pave to septic emboli and metastatic infections.

KEYWORDS: *Lemierre's syndrome, potentially, fatal, suppurative thrombophlebitis, internal jugular vein, bacteraemia.*

INTRODUCTION

EPIDEMIOLOGY AND PATHOPHYSIOLOGY

In all the major case series and reviews, Lemierre's syndrome is primarily an affliction of previously healthy children, adolescents and young adults. In a review of 113 patients, researchers found that most cases presented in the 2nd decade of life (52%), followed by the 3rd decade (22%) and then the 1st decade (9%). There is no clear gender predominance with Lemierre's syndrome, and most cases are diagnosed during the "sore throat season" in the late winter and early spring. While not uncommon in the preantibiotic era, Lemierre's syndrome is now a rare condition with an incidence of 3.7 cases per 1 million per year. Several authors have noted an increase in the number of cases of Lemierre'ssyndrome since the late 1980s. Theories as to the cause of this slight resurgence include an aggrandized awareness of the syndrome, population changes and the more judicious use of antibiotics for the treatment of streptococcal pharyngitis (Courmont et al., 1900).

Fuso bacterium species, most commonly Fusobacterium necrophorum, are responsible for the majority of the bacteraemia in cases of Lemierre's syndrome (Goodman et al., 1917). Up to one third of the patients will have a poly microbial bacteraemia, with anaerobic streptococci and the other miscellaneous gram-negative anaerobes frequently present (Schottmuller et al., 1918). F. necrophorum is a gram-negative anaerobic rod that is the key part of the normal flora of the oropharynx. It is not known what causes this specifically non invasive organism to penetrate the mucosal surfaces, though some authors have proposed an alteration in the pharyngeal mucosa by a viral or bacterial pharyngitis might play a role. Several cases of Lemierre's syndrome preceded by the infectious mononucleosis have been reported (Lemierre, 1936). The palatine tonsils and peritonsillar tissue are the primary sources of infection in most cases. Other primary sources of the infection encompass the lungs, middle ear, mastoid, teeth and sinuses. Following the primary infection, there is local invasion of the lateral pharyngeal space and septic thrombophlebitis, specifically of the IJ vein (Mosher, 1920) (Fig-1).

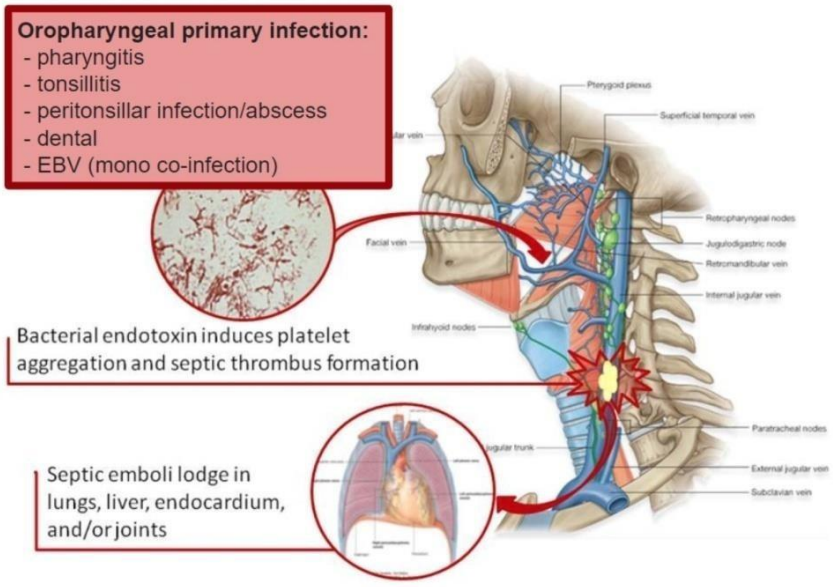

Figure: 1

Metastatic infections following the IJ thrombophlebitis occur in 64%-100% of patients. The lungs are by far the main and common site of metastatic infection in Lemierre's syndrome, followed by the major joints. Metastatic infections of the liver, muscle, pericardium, brain and skin have also been delineated (Kristensen, 2000).

Clinical presentation

Sore throat is the most common symptom in all major series and reviews (Rathore et al., 1990). The onset of the sore throat specifically precedes all other symptoms by 4–5 days, though this interval may be up to 10-12 days. Some patients will have complete resolution of pharyngitis symptoms prior to the onset of the symptoms from the IJ thrombophlebitis or metastatic infection. The neck pain with Lemierre's syndrome is typically unilateral and may be aggravated by turning the head away from the involved side as a consequence of the irritation of the sternocleidomastoid muscle. Since the lungs are the main and common site of metastatic infection, chest pain and pulmonary complaints are the most consistent indicators of the metastatic disease (Venglarcik, 2003).

In 1937, Andre Lemierre published a case series of 20 patients with a syndrome characterized by a history of recent oropharyngeal infection, clinical or radiological evidence of the Internal jugular (IJ) venous thrombosis, and anaerobic septicaemia caused primarily by the Bacillus Fundiform is (now and commonly known as Fuso bacterium necrophorum) (Chow et al.,1978). Lemierre classified this syndrome as the "anaerobic postanginal sepsis" because of the onset of sepsis occurring shortly after the patient shad experienced a sore throat. It was not until the 1990s that anaerobic postanginal sepsis was routinely referred to as Lemierre'ssyndrome. With the introduction of antibiotics in the 1950s and their wide spread use for streptococcal pharyngitis, the incidence of Lemierre's syndrome fell dramatically. In fact, some researchers in the 1980s and 1990s referred to it as a "forgotten disease". For reasons that are not clear, there has been aggrandize in the reporting of Lemierre's syndromes incethelate 1990s. Researchers report here a case of Lemierre's syndrome in an adolescent and review the literature on this uncommon condition (Kaplan et al., 1998).

CASE REPORT

36-year-old woman presented with a three-day history of sore throat, painful swelling of the neck, fever, rigor, hemoptysis, and dyspnea. Examination revealed that the pustular exudates on the right tonsil and thrombophlebitis of the right external jugular vein (Panel A, arrows). Computed tomographic imaging of the chest revealed bilateral pleural effusion and the multiple areas of consolidation with cavitation (Panel B, arrows). Doppler ultra sonography mainly confirmed thrombosis of both the external and the internal jugular veins. Lemierre's syndrome is characterized by disseminated abscesses and thrombophlebitis of the internal

jugular vein after infection of the oropharynx (Monagle et al., 2000).

The predominant pathogen is the gram negative anaerobic bacillus, Fusobacterium Necrophorum*;* in this case, the bacillus was isolated from blood culture after two days. The patient had a sporadic response to therapy with metronidazole, clindamycin, and enoxaparin. She had recovered completely by fourth weeks after discharge (Palmer et al., 1998).

HISTORY

A previously healthy 14 year old boy was brought to our emergency department(ED) by his mother complaining of right sided throat pain present for 6 days (Van Ommen et al., 2001). The pain radiated to his right ear and specifically down the right side of his neck. It was aggravated by the swallowing as well as flexion, extension and rotation of his neck. His mother reported at agile fever also present for 6 days, though noted no change in the child's voice. The patient had been evaluated by his paediatrician 3 days before coming to the ED and was prescribed amoxicillin for suspected strep to coccal pharyngitis. His mother was concerned as the throat pain had mainly worsened despite the antibiotics (Nowak-Gott et al., 2001).

PHYSICAL EXAM

The patient arrived to the ED with a temperature of 37.9° C, though was found to have a temperature of 38.2° C 1hr later (Wright et al., 2012). He appeared to be in no distress, and examination of the heart, lungs, abdomen and skin was quiet unremarkable. Head examination revealed that normal tympanic membranes and external auditory canals bilaterally and no mastoid tenderness and edema (Fig: 1). Hisoropharynx was noted to be non erythematosus and without the exudates, and his uvula was midline. His neck was supple with enlarged, tender lymph nodes pulp able in the right anterior cervical chain (Moore-Gillon et al., 1984).

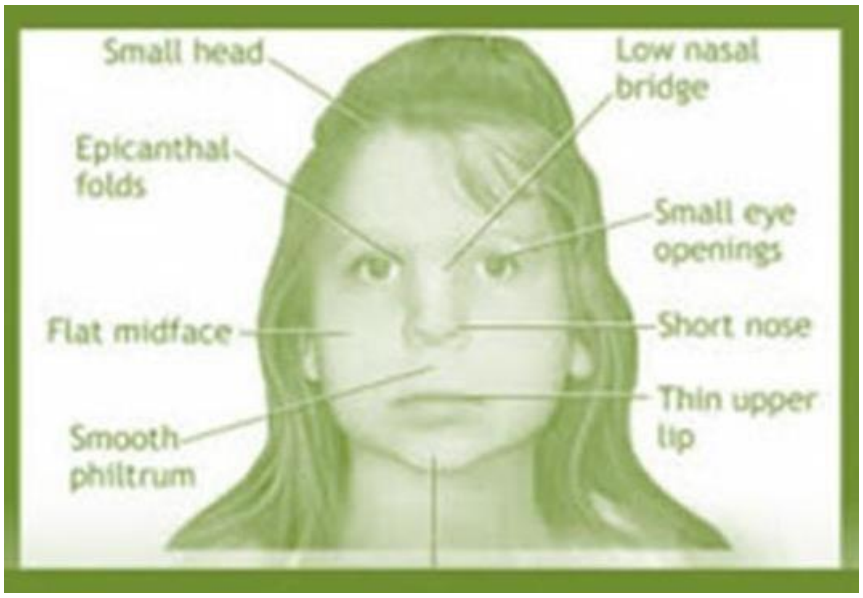

Figure 1: Lemierre's syndrome

DIAGNOSTIC TESTING

A complete blood count mainly included a white blood cell count of 9,400 cells / mcL with 69% neutrophils. Serum electrolytes were all within the normal limits, and urinalysis was negative for signs of infection (Koay et al., 1995). Computed (CT) of the neck with intravenous (IV) contrast was specifically obtained because of concern for a possible deep space abscess. While the CT showed no abscess, an opacification of the specifically right IJ vein was seen extending from the jugular for men to the common jugular confluence, suspicious for high grade partial occlusion (Weesner et al., 1993).

TREATMENT

The patient was given a specific dose of IV ampicillin/ sulbactam for suspected septic thrombophlebitis is of the specific right IJ vein and transferred to a nearby children's hospital for admission and further treatment (Ramirez et al., 2003). During his 6-day hospital course, the patient had no signs of metastatic infection. Two sets of the blood cultures drawn on hospital days 1 and 5 grew no organisms. Workup for the presence of a hyper coagulable state was mainly negative, and the patient was anti coagulated with enoxaparin. He was discharged home to complete a 7 week course of antibiotics and anticoagulation. At a clinic visit 2 month later, the patient was doing well with only a complaint of mild weight loss (Brazier et al., 2002).

The syndrome of jugular vein thrombosis (JVT) allied with suppurative anaerobic infection of the upper aero digestive tract, often complicated by the septic pulmonary embolism (PE), was initially described in the early 20th century (Hagelskjaer et al., 1998).Lemierre's syndrome, as it has since become known, has commonly been allied with a pleiomorphic Gram-negative bacillus (currently designated Fusobacterium Necrophorum) identified from anaerobic culture of blood or purulent fluid. However, in children and perhaps adults, the infection frequently seems to be particularly polymicrobial.JVT may arise from infection of the lateral pharyngeal space secondary to tonsillitis, pharyngitis, mastoiditis, and even the odontogenic infection (including tooth abscess). In addition, both uncomplicated JVT and Lemierre's syndrome can rarely be attributed tothe penetrating oropharyngeal trauma ("pencil-point injury") (Hagelskjaer et al., 2008).

Infection and thrombophlebitis in the anterior compartment of the lateral pharyngeal space often presents as the neck pain, swelling, and tenderness along the anterior border of the sternocleidomastoid muscle; however, primary involvement of the posterior compartment may specifically remain asymptomatic until the disease progresses systemically to encompass septicpulmonary embolism and, in some cases, sepsis syndrome with the multisystem organ dysfunction(Moreno et al.,1989).Lemierre's syndrome was particularly recognized in the preantibiotic era as the complication of pharyngit is with parapharyngeal abscess and was rarely reported in the literature during the 1950s and 1970s,when penicillin came into common use. In recent decades, however, an aggrandizing number of reports of Lemierre's syndrome in childhood have been published, principally focusing on the presentation of illness, antimicrobial therapy, and clinical course of the infection, with little attention to thromboembolic outcomes (Jones et al., 2001).

To date, outcomes of thrombosis in children have been mainly determined in at least 3 large paediatric thrombosis registries. Although these studies and research have in general suggested a low rate of clinically apparent and recurrent venous thromboembolism (VTE), they have raised concern that the development of chronic venous insufficiency in the form of the post thrombotic syndrome may be a frequent sequela of thrombosis particularly in children (Sinave et al., 1989).

The recent cohort assessment and the incorporation of uniform radiologic assessment of

thrombus evolution over time in the evaluation and analysis of paediatric VTE outcomes has led to the understanding that, in many cases, thrombosis may persist after standard anticoagulant therapy. The systematic evaluation of VTE outcomes in children, to our knowledge, has never been delineated separately for JVT or for Lemierre's syndrome. Given the clinical severity of this disorder and its emergence as a key clinical entity in children, we sought to evaluate VTE outcomes of JVTallied with aerobic and anaerobic infection of the head and neck (Lemierre's and Lemierre's-like syndromes [LALLS]) from within the context of the larger cohort study of paediatric thrombosis (Leugers et al., 1995).

OVERVIEW

With the approval of the Colorado Multi-Institutional Review Board, clinical and laboratory information and manifestations from children who ranged in age from birth to 22 years and had received a diagnosis of JVT or Lemierre's syndrome at the Children's Hospital, Denver, and The Mountain States Regional Hemophilia and Thrombosis Center (Aurora, CO) between January 2002 and January2005 was reviewed from the database of a larger cohort study of paediatric thrombosis. Case designation of the LALLS required all of the following criteria: (1) radiologic confirmation of the JVT, (2) a clinical diagnosis of pharyngitis or the other febrile illness, and (3) intra operative evidence of loculated infection typically in the head and neck region or radiologic demonstration of bilateral pulmonary infiltrates(Riordan et al.,2004).Isolation of a causative pathogenic organism by microbiologic culture of blood, tissue, or purulent fluid was also necessary diagnostic criterion among patients who had not been treated with antibiotics before culture but was not restricted to anaerobes (Chirinos et al.,2002). A designation of the classic Lemierre's syndrome was reserved for children who met the aforementioned criteria and in whom anaerobic infection was demonstrated microbiologically. Children in whom JVT was associated with the presence of an ipsilateral central venous catheter were excluded from the analysis. Lemierre syndrome is mainly caused by an acute oropharyngeal infection associated with secondary septic thrombophlebitis of the internal jugular vein, frequently complicated by metastatic infections to the lungs or large joints (Eykyn, 1989).

A previously healthy and fully immunized 1 year old girl was admitted with fever, neck pain and moderate dyspnea for four days. On examination, there was bilateral anterior cervical lymphadenopathy and tender swelling specifically in the left laterocervical region extending

from the angle of jaw and parallel to the sternocleidomastoid muscle. Laboratory analysis and further tests showed a non specific inflammatory reaction with leucocytosis and elevated ESR. Throat cultures were negative for Corneybacterium Diphtheriae and beta hemolytic streptococci. The chest *X*-ray showed bilateral perihilar infiltrates indicating metastatic infection from oropharynx. Colour Doppler ultrasonography of the neck revealed thrombophlebitis of left internal jugular vein. Adiagnosis of Lemierre syndrome was made (Karkos et al., 2009). The child was treated with the crystalline penicillin and chloramphenicol, metronidazole and low molecular weight heparin. Blood cultures were sterile. Serial radiologic follow up revealed resolution of the thrombus over time

Fusobacterium Necrophorum is the etiological agent in over 82% of cases of Lemierre's Lemierre Syndrome in the Antibiotic Era syndrome. Researchers did not isolate this organism, as had already started antibiotics before the blood culture was taken (Alvarez et al., 1995). The palatine tonsils and peritonsillar tissue are the prime source of infection in the majority, although pharyngitis, otitismedia and mastoiditis have been described. Lungs are the most common sites of embolic disease. A tender swelling at the angle of the jaw and parallel with the sternocleidomastoid muscle mainly reflects the development of thrombophlebitis of the internal jugular vein. The mainstay of the treatment is prolonged intravenous antibiotics directed at anaerobic microbes and therapeutic anticoagulation.

CASE STUDY

A 36-year-old woman presented with a three-day history of sore throat, painful swelling of the neck, fever, rigor, hemoptysis, and dyspnea. Examination revealed pustular exudates on the right tonsil and especially thrombophlebitis of the right external jugular vein. Computed tomographic imaging of the chest revealed bilateral pleural effusion and multiple areas of the consolidation with cavitation. Doppler ultrasonography confirmed the thrombosis of both the external and the internal jugular veins (Dagan, 1987).

Lemierre's syndrome is featured by disseminated abscesses and thrombophlebitis of the internal jugular vein after infection of the oropharynx. The dominant pathogen is a gram-negative anaerobic bacillus, Fusobacterium Necrophorum*;* in this case, the bacillus was isolated from the blood culture after two days. The patient had a sporadic response to therapy with metronidazole, clindamycin, and enoxaparin. The patient had recovered completely by

3 weeks after discharge (Goldenhagen et al., 1988).

Lemierre's syndrome (synonyms: Jugular vein suppurative thrombophlebitis, postanginal sepsis, necrobacillosis, and "Forgotten Disease") is the potentially lethal condition in a young and healthy patient population. The syndrome comprises of pharyngitis progressing to jugular vein thrombophlebitis. Infection progresses from the oropharynx to the parapharyngeal or the lateral pharyngeal space, typically in less than one week. The disease affects otherwise healthy young adults with the mean age of onset of 22 years. As such, the disease is frequently unsuspected and the imaging findings often precede clinical suspicion. The disease was much common in the pre-antibiotic era, but there has been a reemergence of cases due to antibiotic resistance (Burden, 1991).

The clinical presentation of the young patient with persistent fever despite antibiotic therapy and neck pain should raise suspicion. Occasionally, the patient will present with tonsil abscess or even the purulent drainage of the involved vessel. Septic emboli to the lung is a hallmark of the disease and occurs in 80-98% of cases.

When the diagnosis is suspected in a patient older than 15 years, a CT of the neck with contrast is the imaging study of choice. For the pediatric population under 15 years of age, an ultrasound of the neck can be ordered first. Diagnosis is established with the imaging demonstrating jugular vein thrombosis in the appropriate clinical setting. The classic imaging triad, as present in the case above, comprises of 1) ipsilateral pharyngeal fullness, 2) pulmonary septic emboli, and 3) neck vein thrombosis (Carlson et al., 1994).

Patients may have metastatic infections elsewhere and the clinician should be the aware of empyema, septic arthritis, and/or osteomyelitis as possible complications. Intracranial complications encompass meningitis, abscess, and cavernous sinus thrombosis. Additional diagnostic analysis should be tailored to the signs and symptoms of these manifestations.

Treatment consists of antibiotics for at least 5 weeks. The benefits of anticoagulation and surgery are uncertain, but are frequently employed. Mortality from the disease remains at 5-9% (Lustig et al., 1995).

CONCLUSION

Lemierre's syndrome' s clinical manifestation, the signs and symptoms like signs and symptoms of Lemierre's syndrome vary, but usually start with a sore throat, fever, and general body weakness. These are followed by extreme lethargy, spiked fevers, rigors, swollen cervical lymph nodes, and a swollen, tender or painful neck; these are discussed profoundly in this paper.

REFERNCES

1. Courmont P, Cade A. On a case of sepsis caused by an anaerobic streptobacillus [in French].*Arch Med ExpAnatPathol.* 1900;12:393–418
2. Goodman C. Primary jugular thrombosis due to tonsil infection. *AnnOtolRhinolLaryngol.* 1917;26:527–529
3. Schottmu¨ ller H. On the pathogenesis of anaerobic bacillus infection [in German; abstract].Deutsche Med Wochenschr.1918;44:1440
4. Mosher HP. Deep cervical abscess and thrombosis of the internal jugularvein.Laryngoscope.1920;30:365–375
5. Lemierre A. On certain septicaemias due to anaerobic organisms.Lancet.1936;1:701–703
6. Kristensen LH, Prag J. Human necrobacillosis, with emphasis on Lemierre'ssyndrome. Clin Infect Dis. 2000;31:524–532
7. Venglarcik J. Lemierre's syndrome. Pediatr Infect Dis J. 2003;22:921–923
8. Rathore MH, Barton LL, Dunkle LM. The spectrum of fusobacterial infections in children. Pediatr Infect Dis J. 1990;9:505–508
9. Chow AW, Roser SM, Brady FA. Orofacialodontogenic infections.AnnIntern Med. 1978;88:392–402
10. Kaplan DM, Fliss DM, Pediser Y, Greenberg D, Leiberman A. Internaljugular vein thrombosis in a child due to a "pencil point injury" of the palate. Int J Pediatr Otorhinolaryngol. 1998;44:183–187
11. Palmer AL, Strain JD, Henry DB, Karrer FM, Simoes EA. Postanginal sepsis after oropharyngeal trauma. Pediatr Infect Dis J. 1995;14:249–251
12. Monagle P, Adams M, Mahoney M, et al. Outcome of pediatric thromboembolic disease: a report from the Canadian childhood thrombophilia registry. Pediatr Res. 2000;47:763–766

13. Van Ommen CH, Heijboer H, Buller HR, Hirasing RW, Heijmans SA, Peters M. Venous thromboembolism in childhood: a prospective two year registry in the Netherlands. J Pediatr.2001;139:676–681
14. Nowak-Gott; U, Junker R, Kruez W, et al. Risk of recurrent venous thrombosis in children with combined prothrombotic risk factors. Blood.2001;97:858–862
15. Wright WF, Shiner CN, Ribes JA: Lemierre syndrome. South Med J 2012, 105(5):283–288.
16. Moore-Gillon J, Lee TH, Eykyn SJ, and Phillips I: Necrobacillosis: a forgotten disease. BMJ 1984, 288(6429):1526–1527.
17. Weesner CL, Cisek JEL: Lemierre syndrome: the forgotten disease. Ann Emerg Med 1993, 22(2):256–288.
18. Koay CB, Heyworth T, and Burden P: Lemierre syndrome: a forgotten complication of acute tonsillitis. J Laryngol Otol 1995, 109(6):657–661.
19. Brazier JS, Hall V, Yusef E, Duerden B: Fusobacterium Necrophorum infections in England and Wales 1990–2000. J Med Microbiol 2002, 51(3):269–272.
20. Ramirez S, Hild TG, Rudolph CN, Sty JR, Kehl SC, Havens P, Henrickson K, Chusid MJ: Increased diagnosis of Lemierre's syndrome and other Fusobacterium Necrophorum infections at a children's hospital. Pediatrics 2003, 112(5):e380–e385.
21. Hagelskjaer, Kristensen L, Prag J: Lemierre's syndrome and other disseminated Fusobacterium necrophorium infections in Denmark: a prospective epidemological and clinical survey. Eur J ClinMicrobiol InfectDis 2008, 27(9):779–789.
22. Hagelskjaer LH, Prag J, Malczyski J, Kristensen JH: Incidence and clinical epidemiology of necrobacillosis, including Lemierre's syndrome in Denmark 1990–1995. Eur J ClinMicrobiol Infect Dis 1998, 17(8):561–565.
23. Moreno S, Garcia Altozano J, Pinilla B, Lopez JC, de Quiros B, Ortega A, Bouza E: Lemierre's disease: postanginal bacteraemia and pulmonary involvement caused by Fusobacterium Necrophorum. Rev Infect Dis 1989,11(2):319–324.
24. Jones JW, Riordan T, Morgan MS: Investigation of postanginal sepsis and Lemierre's syndrome in the south west peninsula. Commun Dis PublicHealth 2001, 4(4):278–282
25. Leugers CM, Clover R. Lemierre syndrome: postanginal sepsis. J Am Board FamPract. 1995; 6(5):384–391.
26. Sinave CP, Hardy GJ, Fardy PW. The Lemierre syndrome: thrombophlebitis of the internal jugular vein secondary to oropharyngeal infection. Medicine (Baltimore)

1989; 6(2):85–94.

27. Riordan T, Wilson M. Lemierre's syndrome: more than a historical curiosa. Postgrad Med J. 2004; 6(944):328–334.
28. Chirinos JA, Lichstein DM, Garcia J, Tamariz LJ. The evolution of Lemierre syndrome. Medicine (Baltimore) 2002; 6(6):458–465.
29. Eykyn SJ. Necrobacillosis. Scand J Infect Dis. 1989; 6:41–46.
30. Karkos PD, Asrani S, Karkos CD, Leong SC, Theochari EG, Alexopoulou TD, Assimakopoulos AD. Lemierre's syndrome: a systematic review. Laryngoscope. 2009; 6(8):1552–1559.
31. Alvarez A, Schreiber JR. Lemierre's syndrome in adolescent children - anaerobic sepsis with internal jugular vein thrombophlebitis following pharyngitis. Pediatrics. 1995; 6:354–359.
32. Goldenhagen J, Alford BA, Prewitt LH, Thompson L, Hostetter MK. Suppurative thrombophlebitis of the internal jugular vein: report of three cases and review of the pediatric literature. Pediatric Infect Dis J. 1988;6(6):410–414
33. Dagan R, Powell KR. Postanginal sepsis following infectious mononucleosis. Arch Intern Med. 1987; 6(9):1581–1583.
34. Gruber B, Mhoon EE. Bilateral deep space neck abscesses complicating infectious mononucleosis. Otolaryngol Head Neck Surg. 1987; 6(1):66–68.
35. Burden P. Fusobacterium Necrophorum and Lemierre's syndrome. J Infect. 1991; 6(3):227–231. doi: 10.1016/0163-4453(91)92684-W.
36. Carlson ER, Bergamo DF, Coccia CT. Lemierre's syndrome: two cases of a forgotten disease. J Oral Maxillofac Surg. 1994;6(1):74–78
37. Lustig LR, Cusick BC, Cheung SW, Lee KC. Lemierre's syndrome: two cases of postanginal sepsis. Otolaryngal Head Neck Surg. 1995; 6(6):767–772.