INTRODUCTION
Lemierre’s syndrome, also called postanginal sepsis syndrome, is one of the deadliest complications of acute oropharyngeal infection. Most commonly caused by the bacteria of genus Fusobacterium, it is commonly found in the normal flora of gastrointestinal tract and oropharynx and female genitourinary tract. The pathogenesis of the disease is least understood. However, one of the theories state that due to the close proximity of internal jugular vein (IJV) toward the pharyngeal wall, mucous invasion of the bacteria can occur. The pathogenesis of the disease is least understood. However, one of the theories state that due to the close proximity of internal jugular vein (IJV) toward the pharyngeal wall, mucous invasion of the bacteria can occur. The etiological agent could not be identified as blood cultures were negative; she had already received antibiotics prior to admission. She responded to treatment, and there were no metastatic complications. Though the syndrome is rare, it has severe manifestations and mortality can be reduced, if timely diagnosed and managed.

CASE REPORT
A 36-year-old married female presented with the complaints of high-grade fever with chills, sore throat, and odynophagia for 7 days. Initially, she had visited a general practitioner for sore throat for which she was started on oral antibiotics and anti-inflammatory medications for the same. After a period of 3 days, she developed diffuse swelling on the right side of the neck along with difficulty in neck movements and a dry cough. She had no history of headache, altered sensorium, seizure, and blurring of vision. There was no history of any instrumentation in the neck or arm. Her personal history was significant with a history of chronic tobacco chewing with poor oral hygiene and recurrent sore throat. Otherwise, her past history was unremarkable. There was no history of intake of any oral contraceptive pills or any drug abuse.

On examination, she was toxic and febrile with a temperature of 101°F; her pulse was regular with a rate of 110/min, a respiratory rate of 20 breaths/min, and a supine blood pressure of 120/80 mmHg. Oral examination revealed pharyngitis with swelling on the neck on the right side with palpable submental and submandibular lymph nodes bilaterally. Local site ultrasonography revealed thrombosed right IJV with subcentimetric regional lymph nodal enlargement. She was started on prophylactic intravenous antibiotics and anticoagulants, awaiting blood culture report. A detailed workup was done to rule out occult malignancy, and autoimmune as well as thrombotic screening was also negative. The etiological agent could not be identified as blood cultures were negative; she had already received antibiotics prior to admission. She responded to treatment, and there were no metastatic complications. Though the syndrome is rare, it has severe manifestations and mortality can be reduced, if timely diagnosed and managed.

Key words: Internal jugular vein, oropharyngeal infection, thrombophlebitis

ABSTRACT
Lemierre’s syndrome is an infectious thrombophlebitis of internal jugular vein (IJV) which occurs rarely, as a complication of oropharyngeal infection. We report an unusual case of a 36-year-old healthy female who presented with high-grade fever with chills, sore throat, and diffuse swelling of the neck on the right side. Examination revealed pharyngitis with swelling on the neck on the right side with palpable submental and submandibular lymph nodes bilaterally. Local site ultrasonography revealed thrombosed right IJV with subcentimetric regional lymph nodal enlargement. She was started on prophylactic intravenous antibiotics and anticoagulants, awaiting blood culture report. A detailed workup was done to rule out occult malignancy, and autoimmune as well as thrombotic screening was also negative. The etiological agent could not be identified as blood cultures were negative; she had already received antibiotics prior to admission. She responded to treatment, and there were no metastatic complications. Though the syndrome is rare, it has severe manifestations and mortality can be reduced, if timely diagnosed and managed.

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Lemierre’s syndrome is an infectious thrombophlebitis of internal jugular vein (IJV) which occurs rarely, as a complication of oropharyngeal infection. We report an unusual case of a 36-year-old healthy female who presented with high-grade fever with chills, sore throat, and diffuse swelling of the neck on the right side. Examination revealed pharyngitis with swelling on the neck on the right side with palpable submental and submandibular lymph nodes bilaterally. Local site ultrasonography revealed thrombosed right IJV with subcentimetric regional lymph nodal enlargement. She was started on prophylactic intravenous antibiotics and anticoagulants, awaiting blood culture report. A detailed workup was done to rule out occult malignancy, and autoimmune as well as thrombotic screening was also negative. The etiological agent could not be identified as blood cultures were negative; she had already received antibiotics prior to admission. She responded to treatment, and there were no metastatic complications. Though the syndrome is rare, it has severe manifestations and mortality can be reduced, if timely diagnosed and managed.

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Yarappa, et al.: Lemierre’s syndrome complicating oropharyngeal infection

There was a diffuse swelling of the neck on the right side [Figure 1], associated with tenderness, and neck movements were painful. However, there was no generalized petechiae or any rash, and systemic examination was unremarkable. There was no lump in the breast or axilla.

Investigations showed a high total leukocyte count of 17,000/mm³ (96% polymorphs) and a platelet counts of 3.4 lakhs/mm³; kidney and liver function tests were within normal range. Erythrocyte sedimentation rate and C-reactive protein were raised. Her viral screen was negative including HIV, HBsAg, and hepatitis C virus. Monospot was negative.

Chest radiograph was normal. Local site ultrasonography revealed evidence of a thrombosed right IJV with significant soft-tissue edema [Figure 2]. There was subcentimetric bilateral submandibular and submental lymph node enlargement.

Two sets of blood samples were withdrawn 12 h apart and sent for both aerobic and anaerobic blood cultures; they were followed up for growth after 12, 24, and 48 h, but were negative. In the meantime, she was started on intravenous (IV) piperacillin tazobactum 4.5 g 6 hourly, clindamycin 600 mg IV 8 hourly, and anti-inflammatory drugs along with low-molecular-weight heparin subcutaneously 12 hourly. Further, a detailed workup was done to rule out occult malignancy. Her ultrasonography of the abdomen and pelvis was normal. Breast and axillary screening was negative.

Contrast-enhanced computed tomography (CT) of the neck [Figure 2] and chest revealed the same thrombosis of right IJV with reactive lymph nodes and the chest was normal. Thrombotic and autoimmune profile was negative which included prothrombin time, activated partial thromboplastin time, protein C and protein S deficiency, Factor V Leiden mutation, homocysteine, antinuclear antibody, peri-nuclear antineutrophil cytoplasmic antibody (ANCA) and cytoplasmic ANCA, and antiphospholipid antibodies.

After 1 week of antibiotic and supportive care, her swelling regressed significantly, and she was symptomatically better [Figure 3]. Repeat ultrasonography of the neck was done which revealed a thrombosed right IJV with significantly reduced soft-tissue swelling and edema.

She was later discharged on antibiotics for 14 days and with oral anticoagulants for 6 months.

DISCUSSION

Lemierre’s syndrome, also referred as human necrobacillosis, is the infectious thrombophlebitis of IJV.[1] First reported by Andre Lemierre in 1936, it was highly feared for its fatal complications.[5] It is most commonly caused by the bacteria of genus Fusobacterium like Fusobacterium necrophorum but also occasionally by Fusobacterium nucleatum, Fusobacterium...
mortiferum, and Fusobacterium varium. These are anaerobic, Gram-negative, nonmotile, nonsporulating, pleomorphic bacilli. They usually harbor human gastrointestinal tract, oropharynx, and urogenital tract.\(^1,6\)

Literature reports many theories to the pathogenesis of the disease.\(^2^1\) One such theory suggests that the presence of viral or bacterial pharyngitis weakens the integrity of oral mucosa. After the invasion of the mucosa of pharynx has occurred, the local structures provide no barrier to the further spread of Fusobacterium from the peritonsilar space into the vessel, especially IJV.\(^3,4\) Further, platelet aggregation and subsequent thrombus formation in the IJV are proposed to be a direct consequence of bacteremia and also it is a potential source of metastatic septic emboli.\(^1,6\) In a few cases, the thrombophlebitis has also been reported in the branches of external jugular vein in addition to IJV.\(^7\)

The classical presentation of the illness begins with a sore throat and high-grade fever with chills. In some cases, it has been observed that sore throat resolves following a brief asymptomatic phase in between and then a full-blown septicemic phase occurs with neck swelling and severe dysphagia. However, in some cases, signs of pleuropulmonary embolization may be the only finding where oral infection resolves. Metastatic complications in the form of septic arthritis, osteomyelitis, and meningitis have been well documented in literature.\(^8\)

The proposed antibiotic regimens include beta-lactam agents and metronidazole or clindamycin for anaerobic coverage for 2–6 weeks. Some authors have strongly supported that anticoagulants should be added to the antibiotic regimen.\(^4\) Moore \textit{et al} in their study of 41 cases of Lemierre’s syndrome stated that 11 patients were found to have been benefitted by the addition of anticoagulation treatment, thus highlighting the role of anticoagulation.\(^9\)

**CONCLUSION**

Lemierre’s syndrome is a rare but highly lethal condition with its catastrophic complications. It primarily affects young people, but should be suspected in any person who develops signs or symptoms of internal jugular vein thrombophlebitis secondary to oropharyngeal infection irrespective of association of sepsis. Blood cultures, chest radiographs, and contrast-enhanced CT scanning should be definitive enough to provide a diagnosis. In our case, the diagnosis was consistent with the syndrome, but confirmation of the etiological agent was unsuccessful as the patient was already on antibiotics. We aim to create awareness about this syndrome, best known as “the forgotten disease” as it can complicate any simple case of sore throat.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

**Disclosure**

This material has never been published and is not currently under evaluation in any other peer reviewed publication.

**Ethical approval**

The permission was taken from Institutional Ethics Committee prior to starting the project. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

**Informed consent**

Informed consent was obtained from all individual participants included in the study.

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