CASE REPORT

Lemierre Syndrome in a Teenager Presenting as Pulmonary Septic Embolism

Rui Domingues[1], João Neves[2], Flora Candeias[1], Paula Kjöllerström[2], Maria João Brito[1]

Port J Pediatr 2021;52:40-2
DOI: https://doi.org/10.25754/pjp.2021.18095

Abstract

Lemierre syndrome is easily missed and may be more common than generally believed. Usually a complication of a deep neck abscess, it can present suddenly with shortness of breath and hypoxemia. Accurate diagnosis and orientation are mandatory for the treatment of an otherwise potentially life-threatening disease. We describe a case of an adolescent with Lemierre syndrome and septic pulmonary embolism.

Keywords: Adolescent; Diagnosis/Differential; Dyspnea/etiology; Hypoxia/etiology; Lemierre Syndrome/diagnosis; Lemierre Syndrome/therapy; Pharyngitis/complication; Pulmonary Embolism/etiology; Treatment Outcome

Introduction

In pediatric patients, the natural course of pharyngitis is complete resolution within one week.[1] If the disease presents worsening symptoms, lasting longer than expected or the emergence of painful unilateral swelling of the neck, other diagnoses should be considered, including pharyngeal abscess and Lemierre syndrome.[1-7] Lemierre syndrome is a rare but life-threatening disease.[4,5,8] It is a condition characterized by infectious thrombophlebitis of the internal jugular vein and bacteremia, following an oropharyngeal infection caused primarily by anaerobes.[1,2] In adolescents and young adults, septic emboli can spread to the lungs or central nervous system.[4,5] First described in 1936, its incidence and mortality rate decreased drastically after the introduction of antibiotics and are currently estimated in 0.6-2.3 cases per 1,000,000 and 4%-18%, respectively.[3] Diagnosis is often only considered when systemic dissemination and life-threatening sepsis occur. This disorder preferentially affects young, previously healthy patients, presenting prolonged pharyngitis later associated with symptoms of septicemia and pneumonia.[2,3] Identification of internal jugular vein thrombophlebitis as well as isolation of anaerobic bacteria in cultures, namely *Fusobacterium necrophorum*, confirms the diagnosis,[4,5,7] However, one third of patients with Lemierre syndrome have polymicrobial infections.[3,8] We present a case of an adolescent with Lemierre syndrome and pulmonary septic embolism.

Case Report

A previously healthy 17-year-old adolescent girl was diagnosed with pharyngitis one week before admission and treated with azithromycin. The patient had a sore throat and persistent fever, despite antibiotic therapy with intravenous amoxicillin and clavulanic acid, 50 mg/kg, three times per day, started two days before referral. On the day of admission at our hospital, the patient presented with cough, chest pain, and dyspnea. Physical examination revealed painless enlarged left cervical lymph nodes (with normal temperature and no flushing), pharyngeal exudate, tachypnea (60 breaths per minute), hypoxemia - 82% oxygen saturation, fraction of inspired oxygen (FiO_2) 21%, intercostal and subcostal retractions, and hepatosplenomegaly. The initial laboratory investigation was remarkable for leukocytosis (24,000 cells/μL), neutrophilia 20,700 cells/µL, normocytic anemia (hemoglobin 10.6 mg/dL), thrombocytopenia (22,000 cells/µL), and elevated C-reactive protein (194.3 mg/L). A chest X-ray revealed multiple nodular lesions and bilateral pleural effusion. The patient rapidly developed severe hypoxemia (80% oxygen saturation despite receiving 15 L/min, FiO_2 nearly 100%) progressing to shock, requiring

1. Pediatric Infectious Diseases Unit, Pediatric Department, Hospital Dona Estefânia, Centro Hospitalar Lisboa Central, Lisboa, Portugal
2. Pediatrics Department, Hospital Dona Estefânia, Centro Hospitalar Lisboa Central, Lisboa, Portugal
3. Pediatric Hematology Unit, Pediatrics Department, Hospital Dona Estefânia, Centro Hospitalar Lisboa Central, Lisboa, Portugal
Corresponding Author
Rui Domingues
https://orcid.org/0000-0001-9427-393
rp.domingues87@gmail.com
Hospital Dona Estefânia, Rua Jacinta Marto, 1169-045 Lisboa, Portugal
Received: 20/06/2019 | Accepted: 16/07/2020 | Published: 03/01/2021

intensive care support. Clinical presentation raised the hypothesis of a pulmonary embolism and, due to recent pharyngitis, Lemierre syndrome was considered. The patient was treated with ceftriaxone (100 mg/kg, once a day) and clindamycin (40 mg/kg/day, three times per day). A diagnosis of venous thrombosis of the internal jugular vein was confirmed by Doppler ultrasound, and anticoagulation with subcutaneous low-molecular weight heparin (1 mg/kg, twice per day) was started on day five (D5). Cervical computed tomography (CT) scan on D5 showed multiple left cervical lymphadenopathies without evidence of an abscess.

Computed tomography angiography performed on D15 (Fig. 1) confirmed massive pulmonary thromboembolism. Anaerobic blood culture and throat culture were negative. Immunoglobulins (Ig) G and M for cytomegalovirus, Epstein-Barr virus viral capsid antigen IgM, and early antigen IgG were negative. Epstein-Barr nuclear antigen IgG was positive. The patient progressively improved and, from D3, there was apyrexia and she was weaned from artificial ventilation and transferred from the pediatric intensive care unit. The patient received a total of three weeks of intravenous antimicrobial therapy and was discharged on D23 with normal cervical CT scan. Anticoagulation was stopped after three months. After one year, she does not have apparent sequelae and no recurrent episodes.

Discussion

The diagnosis of Lemierre syndrome is first and foremost clinical and can be based on clinical and radiologic findings in the absence of a positive culture.[4] In our case, all of the blood cultures were negative probably because the patient had received a prior antibiotic course with azithromycin followed by amoxicillin and clavulanic acid and the diagnosis was considered based on the clinical and radiologic findings. Oropharyngeal infection is the single most common initial symptom.[4,8] In young children, this disorder is usually a complication of a deep neck abscess,[2,8] but in adolescents and young adults, it can complicate a milder disease and have a sudden presentation of dyspnea and hypoxemia,[3,5] as in our case. Cytomegalovirus serology was negative and Epstein-Barr serology was consistent with prior resolved infection. These findings rule out the hypothesis of previous infection by these agents. Lemierre syndrome has been associated with several pulmonary complications, including pleuropulmonary emboli, pleural effusion, empyema, and abscess.[5,8]

The management of Lemierre syndrome includes supportive care, surgical drainage of pharyngeal or mediastinal collections, and broad-spectrum antibiotics.[5,7,8] Intravenous antibiotics are the mainstay of treatment,[4,5] and must cover anaerobic bacteria, namely penicillin and clindamycin, or ceftriaxone and clindamycin. Other options like piperacillin/tazobactam should be used in specific cases.[1,4,5] Because of the frequent occurrence of a polymicrobial infection, the association of two antibiotics is recommended, depending on the antibiotics used.[3] Like in our case, most treated patients have a favorable prognosis, but delayed treatment is associated with poorer outcomes.[5,8]

The role and extent of anticoagulation for jugular vein suppurative thrombophlebitis is controversial. Some authors recommend anticoagulation only if there is evidence of thrombus extension.[6,8] In our patient, we decided to start therapeutic anticoagulation that was maintained for three months with complete thrombus resolution. Accurate diagnosis and orientation are mandatory for the treatment of an otherwise potentially life-threatening disease.

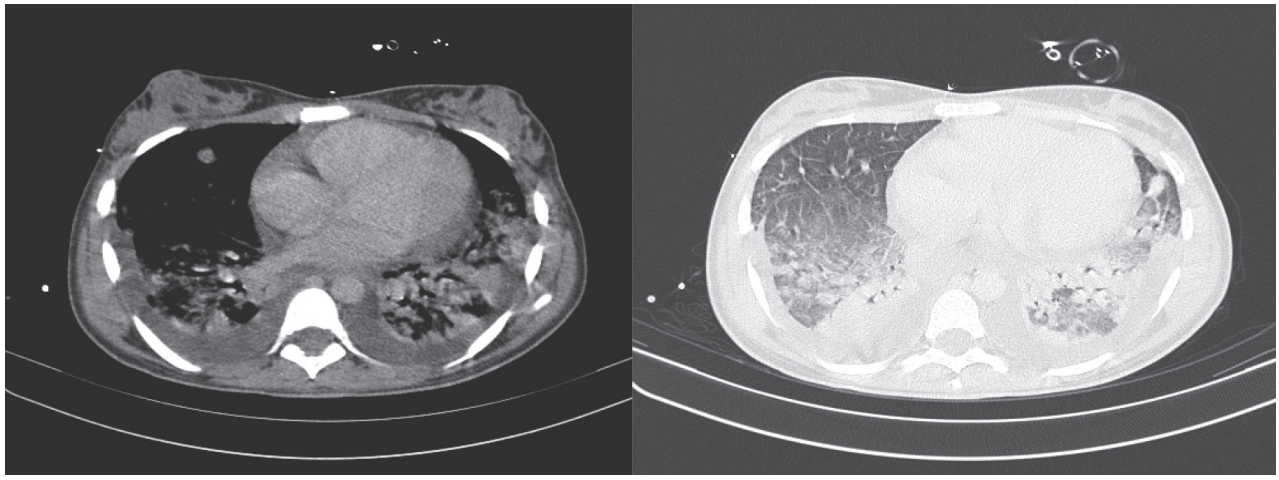

Figure 1. Computed tomography angiography showing massive pulmonary thromboembolism.

WHAT THIS CASE REPORT ADDS

- Lemierre syndrome is a rare but life-threatening disease that should be considered in previously healthy children with prolonged symptoms of pharyngitis later accompanied by symptoms of septicemia and pneumonia.
- Usually is a complication of a deep neck abscess, but in adolescents and young adults, it can complicate a milder disease with empyema and pleural effusion.
- Accurate diagnosis and orientation are mandatory for the treatment of an otherwise potentially life-threatening disease.
- The role of anticoagulation for jugular vein suppurative thrombophlebitis is controversial but can be recommend if there is evidence of thrombus extension.

Conflicts of Interest
The authors declare that there were no conflicts of interest in conducting this work.

Funding Sources
There were no external funding sources for the realization of this paper.

Provenance and peer review
Not commissioned; externally peer reviewed

Consent for publication
Consent for publication was obtained.

Confidentiality of data
The authors declare that they have followed the protocols of their work center on the publication of patient data.

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Embolia Pulmonar Séptica Como Apresentação de Síndrome de Lemierre numa Adolescente

Resumo:
A síndrome de Lemierre passa facilmente despercebida e pode ser mais comum do que habitualmente se julga. É em geral uma complicação de um abscesso profundo no pescoço e pode apresentar-se subitamente como dificuldade respiratória e hipoxemia. Um diagnóstico e orientação precisos são fundamentais para o tratamento desta doença potencialmente fatal. Descrevemos clínico de uma adolescente com síndrome de Lemierre e embolia pulmonar séptica.

Palavras-Chave: Adolescente; Diagnóstico Diferencial; Dispneia/etiologia; Embolia Pulmonar/etiologia; Hipoxia/etiologia; Resultado do Tratamento; Síndrome de Lemierre/diagnóstico; Síndrome de Lemierre/tratamento