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Radiological Case Report / Caso Clínico

Lemierre Syndrome: An Overlooked Diagnosis in Otherwise Healthy Patients

Síndrome de Lemierre: Um Diagnóstico Esquecido em Doentes Previamente Saudáveis

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Abstract

Lemierre Syndrome is a severe complication of an otorhinolaryngologic infection, resulting in septic thrombophlebitis, bacteriemia and septic emboli, most often in young and previously healthy patients. We present a case of a 24-year-old female patient, that develops an episode of syncope and multiorganic dysfunction, two days after the diagnosis of acute tonsillopharyngitis treated with intramuscular penicillin and anti-inflammatory drugs. Due to worsening of the clinical status, a computed tomography was performed and radiological findings were in keeping with Lemierre Syndrome. The continuous worsening of respiratory and cardiovascular dysfunction led to intensive care unit admission where the patient was submitted to extra-corporeal membrane oxygenation support. Despite being a rare entity, Lemierre Syndrome is potentially life-threatening, therefore it should be recognized as soon as possible. Given its unspecific clinical presentation, a high index of suspicion and radiological evaluation with computed tomography are needed to establish the diagnosis.

Keywords

Embolism; Sepsis; Thrombosis; Palatine Tonsil.

Resumo

A Síndrome de Lemierre é uma complicação grave de uma infeção do foro otorrinolaringológico, que resulta em tromboflebite séptica, com consequente bacteremia e êmbolos sépticos, manifestando-se maioritariamente em adultos jovens previamente saudáveis. Apresentamos o caso clínico de uma doente do sexo feminino com 24 anos que desenvolve quadro de síncope e disfunção multiorgânica dois dias após o diagnóstico de amigdalite aguda, tratada com penicilina intramuscular e anti-inflamatórios. Por agravamento clínico, é realizado estudo tomodensitométrico, cujos achados foram sugestivos de Síndrome de Lemierre. O agravamento da insuficiência respiratória e cardiovascular levaram à admissão da doente nos cuidados intensivos, onde foi submetida a oxigenação por membrana extra-corporal. Embora a Síndrome de Lemierre seja rara na era atual, é uma entidade potencialmente fatal, pelo que deve ser reconhecida o mais precocemente possível. Atendendo à sua apresentação clínica inespecífica, o diagnóstico implica alto índice de suspeição e, muitas vezes, avaliação radiológica por tomografia computorizada.

Palavras-chave

Embolia; Sépsis; Trombose; Amígdala.

Introduction

In developed countries, ENT (ear, nose, and throat) infections are extremely common conditions, typically following a benign course 1, due to the impact of antibiotics on their morbidity and mortality.[2,3]
However, in the last years, there has been an increase in the reported cases of ENT infections that resulted in septic thrombophlebitis of the internal jugular vein followed by systemic embolism in young and previously healthy patients,[4,5] a potentially life-threatening condition known as Lemierre Syndrome.[4,6,7]

Case Report

We present a case of a 24-year-old female patient, with previous adenoidectomy and no other significant past medical history, who presented to the emergency department (ED) after syncope with spontaneous resolution. The patient complained of odynophagia and fever with associated myalgias and headaches over the course of the past three days, which drove her to a primary care facility two days before. There, she was diagnosed with acute tonsillopharyngitis and treated with intramuscular penicillin and anti-inflammatory drugs (ibuprofen), without clinical improvement. At admission to the hospital, she presented leucocytosis (11,86 x 109/L), thrombocytopenia (15 x 109/L), increased C-reactive protein (CRP) (280,4mg/L) and acute kidney injury (serum creatinine of 2,3mg/dL), consistent with sepsis with multiorgan dysfunction.
In the first 24 hours of admission, she underwent clinical deterioration, with severe respiratory insufficiency. Computed Tomography (CT) scan of neck, chest and abdomen (Fig. 1) was performed and showed left thrombophlebitis of the internal jugular vein, associated with an enlarged left tonsil with fluid content and gas bubbles in keeping with an abscess in the left tonsil caused by a gas-producing organism; moreover, there were multiple nodules throughout the lung parenchyma

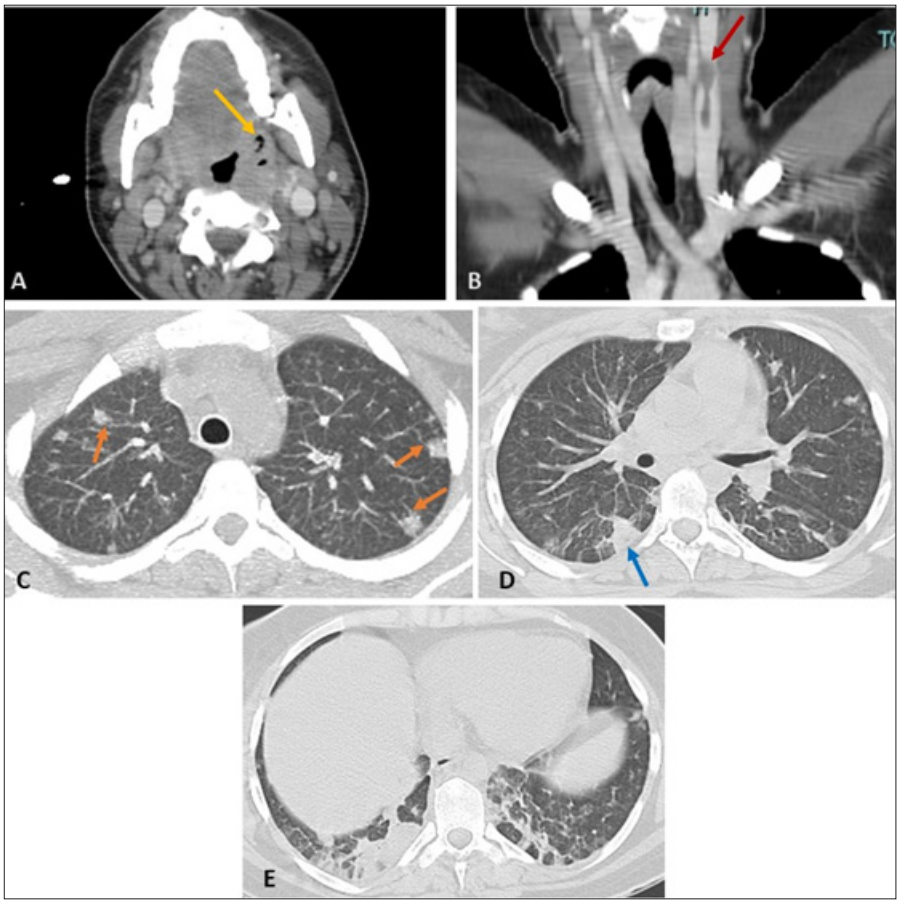

Figure 1 – Contrast-enhanced cervicothoracic CT scan, soft tissue window (axial plane – **A**, coronal reformatted plane - **B**) and lung window (maximum intensity projection in the axial plane at the level of the aortic arch – **C**, axial plane at the level of the pulmonary trunk – **D**, and at the lung bases – **E**) reveals an enlarged left tonsil with fluid content and gas bubbles, in keeping with tonsillitis complicated with an abscess (yellow arrow - A) associated with a thrombus in the left internal jugular vein (red arrow - B). Chest CT shows scattered lung nodules affecting all pulmonary lobes, mostly in a peripheral location, and some of them with a clearly identifiable feeding vessel (orange arrows – C), suggestive of septic emboli. Wedge-shaped peripheral opacities (blue arrow - D), suspicious of septic infarcts, and basal consolidations are also seen (E).

suggestive of septic embolism, that coexisted with wedge-shaped consolidations, suspicious of septic infarcts, and basal consolidations. The first diagnostic hypothesis was Lemierre Syndrome with septic embolization to the lungs.
Blood cultures were positive to group C β-hemolytic streptococci (GCBHS), and susceptibility-guided antibiotherapy was initiated (three days of intravenous ceftriaxone and two days of azithromycin, followed by piperacillin-tazobactam).
The clinical status continued to worsen and the patient was intubated and admitted to the intensive care unit (ICU).
A control CT scan after 5 days of admission was obtained (Fig. 2), showing cavitation of the lung nodules, corroborating the previous diagnosis, and additional

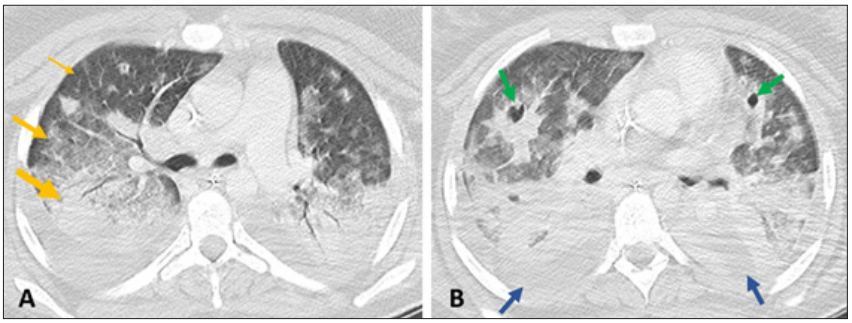

Figure 2 – Contrast-enhanced chest CT scan, lung window (axial plane at two different levels – A and B) shows increase in size and cavitation of the previously identified lung nodules (green arrows - B), confirming the hypothesis of pulmonary septic emboli. In contrast to the previous CT scan, bilateral pleural effusion is now seen (blue arrows) and a radiological pattern of acute respiratory distress syndrome (ARDS), with an anteroposterior density gradient, resulting from dense consolidation in the most dependant regions (thick yellow arrow), followed by ground-glass attenuation (medium yellow arrow) and normal lung in the non-dependant regions (thin yellow arrow), in line with the progressive respiratory insufficiency.

bilateral pleural effusion and central alveolar consolidations presenting with an anteroposterior density gradient, in line with acute respiratory distress syndrome (ARDS).

During the ICU admission, the patient maintained severe systemic inflammatory response syndrome (SIRS) with fever, high CRP (370 mg/L) and leucocytosis (23 x 109/L), and developed progressive haematologic dysfunction (5 x 109/l platelets) and "de novo" liver dysfunction, presenting with mild cytocholestasis and hepatomegaly. There was also renal and cardiovascular dysfunction requiring vasopressor support.

Due to the need of prolonged and aggressive mechanical ventilation, the patient was placed on extracorporeal membrane oxygenation (ECMO) support, where she remained for 42 days. During this time, the most relevant complication was haemoptysis, requiring right bronchial artery embolization.

After ECMO suspension, the patient was intubated for another 15 days, and a tracheostomy was performed. Following another 64 days of mechanical ventilation, decannulation was tried and succeeded. Doppler evaluation showed no signs of jugular vein thrombosis.

At the 80th day of admission, the patient presented with recurrent tonsillitis and was treated with another course of antibiotics (10 days of intravenous ceftriaxone and clindamycin).

After 86 days of hospitalization, the patient was discharged and follow up was made by Otorhinolaryngology. In the months that followed, the patient had four more episodes of tonsillitis and a tonsillectomy was performed without complications.

Discussion

Lemierre syndrome was extensively described for the first time in 1936[3,8] and is characterized by septic thrombophlebitis of the internal jugular vein and bacteremia that can result in septic emboli, most often to the lungs.[3,9] If pulmonary involvement is present, patients can develop respiratory failure, usually due to ARDS.[3,6] Given this, up to 37% of the overall cases of Lemierre syndrome require intubation.[6] This clinical picture is seen in association with an ENT infection, arising more frequently in the oropharynx, and affects primarily younger patients that were previously healthy, mostly in the 2nd and 3rd decades of life.[3,6,7,10] Its incidence significantly decreased in the postantibiotic era and it is a rare disease nowadays; however, it can be fatal with mortality rates reaching up to 6-15%,[4,11] reason why it should be promptly identified and treated.[4,10]

This infection is most commonly caused by anaerobic bacteria, usually Fusobacterium necrophorum, although polymicrobial bacteremia is reported to happen in up to one third of the cases, with anaerobic streptococci being frequently present,[10,12] as in our case.

The first symptoms are usually related to an oropharynx infection and include sore throat, fever, weakness, and malaise.[3,11] The presence of neck pain, pleuritic chest pain and/or respiratory distress in a previously healthy young adult with recent history of pharyngitis or tonsillitis should raise the suspicion of Lemierre syndrome, although its initial indolent course, unspecific clinical presentation and rarity often lead to a delayed diagnosis.[11]

Radiologists have an important role in the diagnostic process, as CT is usually included in the initial workup of a patient whose clinical condition deteriorates, enabling the identification of most of the findings that define this rare condition.[3]

Given the life-threatening potential of Lemierre syndrome and its typical imagiological findings, this condition should be known and promptly recognized by clinicians and radiologists.[3,4,10]

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

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

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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