

LEMIERRE SYNDROME: CLINICAL CHALLENGES AND THE ANTICOAGULATION DILEMMA IN ADOLESCENT CARE

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Abstract

Lemierre syndrome is a rare, life-threatening condition characterized by an acute otorhinolaryngologic infection with septic thrombophlebitis of the internal jugular vein and septic embolism, particularly to the lungs. We describe a case of a previously healthy 15-year-old female patient who initially presents fever and odynophagia but quickly develops neck and pleuritic chest pain. Computed tomography was performed and the radiological findings confirmed the diagnosis of a Lemierre syndrome. She was managed with antibiotics, anticoagulant for three days and symptomatic treatment, with a gradually improving condition. After 17 days of hospitalisation, due to reappearance of pleuritic pain, a new imaging assessment was performed and showed additional septic emboli in the lungs, which prompted the reintroduction of anticoagulant therapy. Awareness of the existence of this syndrome is essential to ensure a radiological evaluation with computed tomography and thus timely diagnosis and treatment.

Keywords: Lemierre Syndrome; Thrombophlebitis; Embolism; Palatine Tonsil

INTRODUCTION

Lemierre syndrome is defined as a septic thrombophlebitis of the internal jugular vein (IJV) as a result of an oropharyngeal bacterial infection. The infection spreads from the oropharynx to the IJV, leading to an infected thrombus in the lumen. Due to bacteraemia, septic embolizations are often observed, mainly in the lungs.^{1,2} Although this syndrome is rare, its incidence has been increasing over the last decade.^{2,3}

This case report illustrates a typical case of Lemierre syndrome, but with interesting and real challenges in clinical practice.

CLINICAL CASE

A 15-year-old female patient, with no significant past medical history, presented with a five-day history of fever and odynophagia, a two-day history of neck pain and pleuritic

chest pain since the previous day.

Her general state of health was reasonable, with no signs of respiratory distress. On chest examination, there was a slight reduction of vesicular murmur with crepitation, bibasilar. She had right-side neck tenderness and trismus.

Investigations revealed leucocytosis ($11.84 \times 10^9/L$) with neutrophilia ($10.66 \times 10^9/L$), thrombocytopenia ($57 \times 10^9/L$), increased C-reactive protein (248mg/L) and activated partial thromboplastin time (aPTT) (44.3 seconds).

Enhancement computed tomography (CT) scan of the neck and lung (Figure 1) revealed a small non-occlusive thrombus in the right IJV (13mm), a peritonsillar abscess and multiple consolidations of the left lower lobe, suggesting septic embolism. Taking into consideration both clinical and radiological findings, a diagnosis of Lemierre Syndrome with septic embolization to the lung was established.

The patient was kept on intravenous ceftriaxone and metronidazole and started anticoagulation with enoxaparin (2 mg/Kg/day).



Figure 1

Contrast-enhanced neck computed tomographic (CT) scan, soft tissue window (axial plane – A and B; coronal plane - C). Image A showing multiseptated collections with peripheral enhancement reflecting a peritonsillar abscess. Images B and C revealing a non-occlusive thrombus in the left internal jugular vein. Contrast-enhanced chest CT scan, lung window (axial plane – D) presenting three consolidated lesions in the left lower lobe.

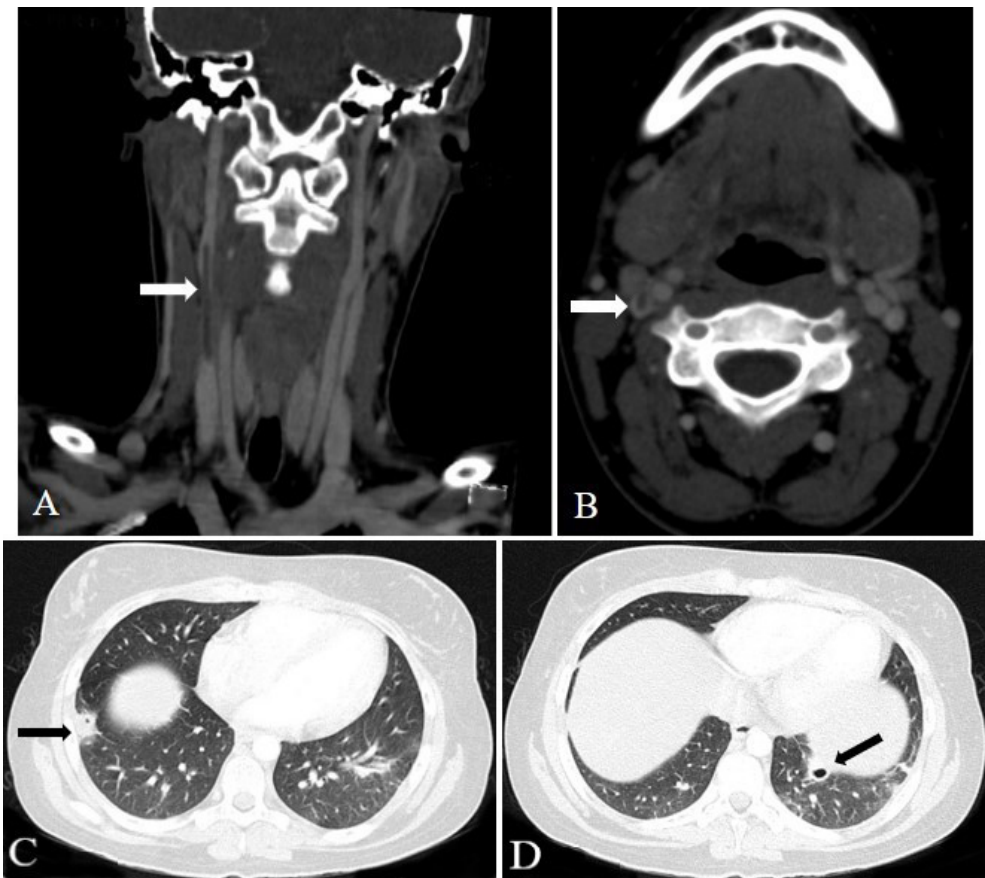


Figure 2

Contrast-enhanced neck computed tomographic (CT) scan, soft tissue window (coronal plane – A; axial plane - B) revealing an increased size of the non-occlusive thrombus in the left internal jugular vein. Contrast-enhanced chest CT scan, lung window (axial plane – C and D) showing two cavitary lesions in the left and right lower lobes.

During the first three days of anticoagulation, the patient had several episodes of epistaxis and coughing with bloody secretions, causing an emotional disruption for the teenager and her family, leading to the decision to stop anticoagulation. On the fourth day, the patient was evaluated by otorhinolaryngology for neck pain and initiated dexamethasone that maintained for nine days. Blood cultures were positive to *Fusobacterium necrophorum*.

A control CT scan of the neck (Figure 2), performed ten days after hospitalisation, revealed resolution of peritonsillar abscess and a greater extension of the non-occlusive thrombus in the IJV (49mm). On day seventeen post admission, the patient complained of pleuritic pain in the lower right side of the chest. An additional CT-angiogram (Figure 2) was obtained, showing areas of septic infarcts of the left lower lobe and new septic emboli in the right lower lobe of the lung. Laboratory tests revealed an increase of D-dimers (1758ng/mL). Anticoagulation was reintroduced with enoxaparin bridging acenocoumarol.

The patient completed thirty days of antibiotics, being discharged from hospital with oral anticoagulant.

DISCUSSION

Lemierre syndrome is a well-recognised and described entity, characterised by septic thrombophlebitis of the IJV due to an infection that usually starts in the oropharynx, invades the pharyngeal mucosa and reaches the parapharyngeal space. Once the infection reaches the IJV, haematogenous dissemination to other sites can occur, leading to several complications and eventually death, due to septic shock^{1,3}. About 80-90% of septic embolism cases occur in the lungs.²⁻⁵

This entity mainly affects younger and healthy patients, especially adolescents or young adults,³⁻⁵ as in our case. According to the literature, *Fusobacterium necrophorum* is the most common pathogen (33,2%), although other agents have been reported (e.g., Methicillin-resistant *S. aureus* - 4.1%; *Fusobacterium nucleatum* – 2.3%; *Klebsiella pneumoniae* – 2.3%).^{1,3,4} The initial presentation with fever,odynophagia and neck pain is consistent with Lemierre syndrome and the development of pleuritic pain should raise suspicions of pulmonary embolisation. Thrombocytopenia is frequently present due to the production of hemagglutinin by *Fusobacterium necrophorum* while aPTT can increase in the context of thrombotic phenomena with consequent activation of the coagulation cascade.

The priority of treatment involves immediate antimicrobial therapy with a B-lactamase inhibitor, since beta-lactamase production by *Fusobacterium necrophorum* has been reported.^{6,7} Additionally, oral streptococci should also be targeted.⁶ The response to the treatment is usually slow, because of limited antibiotic penetration into fibrin clot and/or necrotic abscesses.^{7,8} Treatment duration is not defined, but it may vary from 3-6 weeks.⁶ In our case, a combination of ceftriaxone and metronidazole was initiated and maintained for about four weeks, with good clinical results.

The role of anticoagulation is uncertain in Lemierre Syndrome and should not be used routinely, unless extension of thrombus is evident. Most evidence is limited to case reports.^{2,8-10} Opponents of anticoagulation argue that the clots of IJV usually resolve on their own with good outcomes.⁷ Those in favour of anticoagulation defend its use for faster resolution of clots, thus reducing morbidity.^{7,11} A recent meta-analysis, collecting data from 712 patients with Lemierre syndrome, found that 14.3% of patients were diagnosed with new venous thromboembolism or peripheral septic lesions during hospitalization and the rates of both events were lower if anticoagulation was used.⁹ On the other hand, Nygren et al,⁵ failed to demonstrate the benefit of anticoagulants in preventing progression or recurrence of thrombosis and peripheral septic complication.

In our patient, anticoagulation was suspended due to minor haemorrhages and few days later, an increase in the extension of the thrombus and new septic emboli were detected. It was decided to start acenocoumarol instead of direct oral anticoagulants (DOACs), since physicians had more experience with this drug, the therapy duration would be limited to 3 months and because there are no approved antidotes for DOACs for paediatric patients, which is relevant, taking into account the potential for worsening and the uncertainty of this complex and rare clinical situation.

Some authors recommend using anticoagulation only if there is a lack of response despite 48-72 hours of adequate antimicrobial therapy, underlying thrombophilia and progression of thrombosis or retrograde cavernous sinus thrombosis.^{6,10} When the decision about starting anticoagulation is taken, the risk of haemorrhage should always be assessed, yet the number of reported cases of severe major haemorrhage is very low.^{5,9} This is especially true in young and healthy individuals, as our patient.

Although rare, considering its potentially fatal condition and typical imagiological findings, Lemierre syndrome should be early recognized in the presence of a suggestive presentation.

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