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Lemierre Syndrome Revealed by Orbital Cellulitis

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Abstract

Lemierre's syndrome is a rare and severe sepsis, capable of rapidly compromising the vital prognosis in the absence of early intervention, characterized by an oropharyngeal infection complicated by septic thrombosis of the internal jugular vein and the formation of septic emboli primarily localized at the pulmonary level. We report the case of a 14-year-old boy, with no notable pathological history, who had presented for 15 days with unilateral orbital swelling which then became bilateral, all evolving in a context of fever.

Keywords

Lemierre's Syndrome, Oropharyngeal Infection, Jugular Septic Thrombosis, Septic Embolism

1. Introduction

Lemierre syndrome is a rare and serious form of sepsis that can rapidly become life-threatening if not treated early, combining oropharyngeal infection complicated by septic thrombosis of the internal jugular vein and predominantly pulmonary septic emboli [1] [2].

This pathology generally affects healthy children and adults. The main bacterium involved is a Gram-negative anaerobe belonging to the Bacteroidaceae family [3].

Its diagnosis is often delayed, as it is not frequently evoked and is usually discovered incidentally. Treatment is both medical and surgical, while the use of anticoagulation is controversial [4].

We report the case of a 14-year-old boy, with no previous pathological history of note, who presented with a 15-day history of unilateral orbital swelling which

then became bilateral, all evolving in a context of fever.

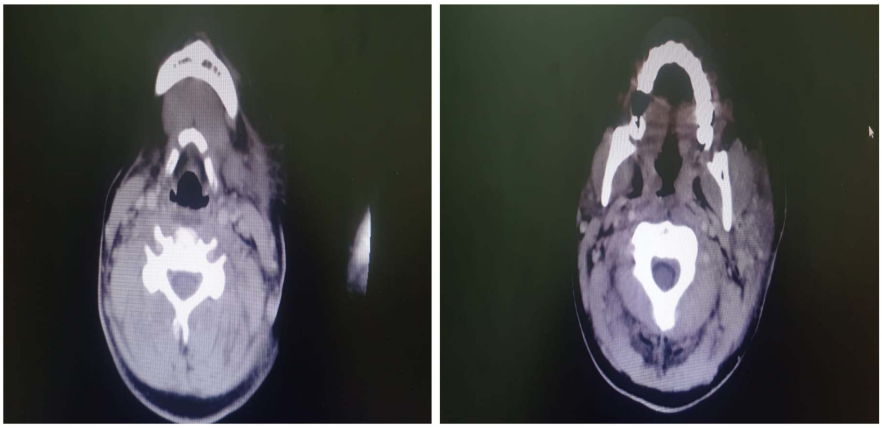
2. Clinical Case

In the pediatric emergency, we received a 13-year-old child with no notable medical history, particularly with regard to hemostasis disorders in the family. The child was conscious, with a low-grade fever of 38˚C. Blood pressure was 12/8 mmHg, heart rate 90 beats per minute, respiratory rate 26 cycles per minute, and saturation 95%. The child was conscious with a Glasgow Coma Score (GCS) of 15, weighed 65 kg and measured 165 cm. Facial examination revealed bilateral palpebral edema with extensive collection of the right upper eyelid, showing inflammatory signs, with extension of the cellulitis to the bilateral frontal and jugal region. The eyeballs did not open spontaneously, and there was bilateral chemosis accompanied by purulent secretions. The rest of the somatic examination was unremarkable.

Paraclinically, the patient underwent a blood test that revealed hyperleukocytosis with a white blood cell count of 16,000 cells/mL, predominantly neutrophils (PNN) with a count of 14,000 cells/mL, and thrombocytopenia with a platelet count of 56,000 cells/mL. Additionally, the patient had normocytic normochromic anemia with a hemoglobin level of 14 g/dL, a mean corpuscular volume (VGM) of 80 fL, and a mean corpuscular hemoglobin concentration (TCMH) of 31 pg. The infectious workup yielded clear positive results, with a C-reactive protein (CRP) level of 320 mg/L. HIV serology returned a negative result, and immunoglobulin assays and lymphocyte subpopulation testing for immune deficiency all fell within normal limits. Liver and kidney function tests showed no abnormalities.

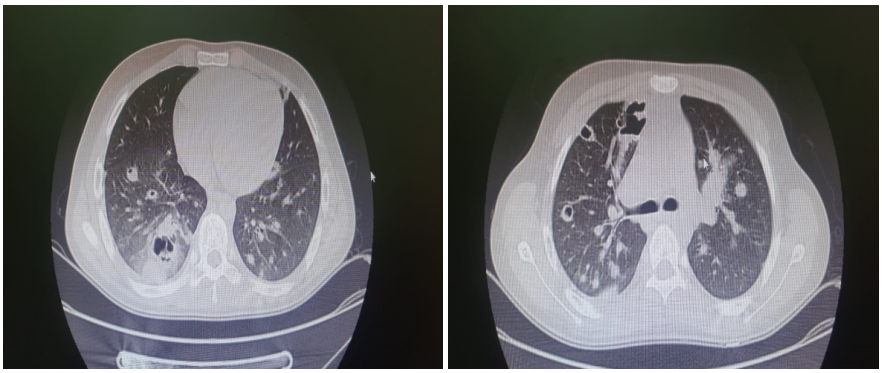
For treatment, the patient received a combination therapy including a 3rd-generation cephalosporin at a dose of 80 mg/kg/day, an aminoglycoside at a dose of 5 mg/kg/day, and metronidazole at a dose of 30 mg/kg/day.

However, within 48 hours of hospitalization, the patient's condition improved clinically. The child became apyretic, with a slight regression of inflammatory signs, and the infectious workup improved, with CRP reduced to 100 mg/L. The blood count rose to 13.8 g/dL, PNN to 9000 cells/µL, and platelets to 100,000/µL, with regression of the palpebral collection on follow-up brain CT. On the third day of antibiotic treatment, the child presented with seizures, respiratory distress and hemoptoic sputum. Clinical examination revealed a child in respiratory distress with altered consciousness, his Glasgow Coma Score (GCS) being 11. Imaging was therefore indicated Cerebral and thoracic CT scans were performed, revealing the following findings: Cerebral: septic thrombosis of both internal jugular veins, complicating orbital cellulitis (**Figure 1**). Thoracic: pulmonary parenchymal septic emboli (**Figure 2**). Taken together, these anomalies suggest Lemierre syndrome. In addition to antibiotic therapy, the child received curative anticoagulant treatment due to the extent of thrombosis and pulmonary embolisms. Unfortunately, the child's outcome was fatal due to profuse hemoptysis secondary to pulmonary embolism.



(a) (b)

Figure 1. Axial section through the cervical level after venous contrast agent injection: Lack of opacification of the left jugular vein (image a), bilateral (image b).



(a) (b)

Figure 2. Axial slices passing through the thoracic level in parenchymal window demonstrating septic emboli in the form of excavated lesions (images a, b).

3. Discussion

Lemierre syndrome (LS) was identified in 1936 by French microbiologist André Lemierre in Paris, in an article published in the Lancet. He based his work on a series of 20 cases, inspired by Courmont and Cade's first observations dating back to 1900 [5] [6].

According to a recent retrospective study in Denmark, Lemierre syndrome (LS) showed an incidence of 2.8 cases per million per year in the general population and an incidence of 9.4 cases per million per year in young people aged 15 to 24 between 2010 and 2014 [7]. These figures are close to the data reported in the 1990s by another Danish study, where incidence was 1 case per million per year in the general population and 14.4 cases per million per year in 15 - 24 year-olds.

This study suggests that the incidence of SL may be underestimated, and highlights an upward trend in this disease over the last two decades [8]. Several factors could explain this increase, including a reduction in antibiotic prescribing, the rise of resistant strains due to inappropriate antibiotic use, excessive use of

non-steroidal anti-inflammatory drugs during ENT infections, and a decline in the number of tonsillectomies performed since the 1970s [9] [10] [11] [12].

Lemierre syndrome (LS) usually occurs within a few days to three weeks of an upper respiratory tract infection. This infection is the main diagnostic finding on medical questioning [1] [13] [14] [15]. The resulting symptoms may be noisy and associated with various infectious localizations, manifesting as high fever (≥38.5˚C), chills, sweating, accelerated heart rate, low blood pressure, rapid breathing, signs of meningitis, difficulty swallowing, jaw muscle contraction, neck tilt, pain in the neck, chest, abdomen or joints, gastrointestinal problems such as diarrhea and vomiting, enlarged eyes, and even paralysis of certain cranial nerves [1] [11] [16] [17].

Physical examination should systematically look for signs of oropharyngeal infection, possibly associated with lymph node complications, swelling or indurated areas in the cervical region along the sternocleidomastoid muscle. In addition, careful examination of the skull and face is essential [1] [9] [12]. On cervical examination, abnormalities are detected in almost half of cases (47.7%), although induration of the internal jugular vein is often not palpable due to its position in the posterior lateral pharyngeal space [18]. Finally, it is vital to exclude any associated circulatory, respiratory or neurological insufficiency [11].

Paraclinical examinations include biological analyses, bacteriological tests, as well as imaging examinations such as ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI).

Optimal antibiotic treatment, although the lack of randomized trials is a limitation, should encompass broad coverage of anaerobic germs, including Fusobacterium nucleatum (FN), as well as streptococci and staphylococci [12] [18]. The first-line regimen for dual antibiotic therapy combines a beta-lactam antibiotic with a beta-lactamase inhibitor (such as piperacillin-tazobactam) or a third-generation cephalosporin (such as ceftriaxone, 10 - 15 mg/kg three times daily in children) with metronidazole [9] [13]. If necessary, second-line monotherapy with imipenem or moxifloxacin can be considered [12] [19].

Anticoagulant therapy in the context of Lemierre syndrome (LS) remains controversial, with uncertainties regarding indications, choice of agent, dosage and duration [12] [20]. The theoretical dilemma lies in the potential risk of promoting the dissemination of septic thrombosis, which generally tends to progress well with antibiotics alone, versus the risk of possible retrograde septic extension in the absence of anticoagulation. Furthermore, the use of anticoagulants can be problematic in the event of emergency surgery and/or the development of a multivisceral failure syndrome with coagulopathy [21]. Consequently, the available studies present conflicting results regarding anticoagulation [15] [22].

For example, a recent systematic review by Johannesen *et al.* based on the analysis of 137 cases reported that anticoagulation was used in 87.5% of cases, mainly in the form of low-molecular-weight heparin (LMWH), whereas an earlier study by Riordan in 2007 reported only a 23% rate of anticoagulation use [13] [14]. Further work showed that in a paediatric population of 11 patients

with cephalic infection (mainly mastoiditis) associated with internal jugular or cerebral venous thrombosis and treated with LMWH, 10 patients (90.9%) experienced complete resolution of their thrombosis without bleeding complications, with a median treatment time of 3.4 months, while only one patient experienced thrombosis progression, attributed to lack of compliance [21]. An additional retrospective study in 2017 of 18 cases of SL occurring between 1998 and 2014 in children and adults, 6 of whom were treated with LMWH for ≥ 4 weeks (median: 23 weeks), revealed no significant difference in terms of resolution, progression or recurrence of thrombosis at three months, when monitored by echo-Doppler, compared with antibiotic treatment alone [23].

Ultimately, surgical intervention including drainage of infected cervicofacial tissue and abscess evacuation may be necessary in around 50% of cases [9] [13] [15]. However, ligation-exeresis of the internal jugular vein remains an exceptional procedure, reserved for situations where the evolution is unfavorable, notably in cases of persistent septic emboli despite optimal antibiotic treatment, extensive septic thrombosis, or uncontrollable severe sepsis [1] [12].

Complications of SL lead to significant morbidity and mortality in the absence of prompt administration of anti-infective therapy [1]. Indeed, the risk of septic dissemination is closely linked to the delay in treatment [18]. Mortality, which André Lemierre estimated at around 90% in the absence of antibiotic treatment, can be reduced to around 2% - 10% with optimal management [5] [13] [18]. The majority of patients make a full recovery with treatment, but in some cases the disease can unfortunately lead to death [12] [20].

4. Conclusion

The diagnosis of Lemierre syndrome must be confirmed rapidly, usually by a cervicothoracic CT scan. The standard treatment is urgent antibiotic therapy. Anticoagulation should be considered in situations presenting a high risk of thrombosis. In some cases, surgical treatment may be necessary.

Conflicts of Interest

The authors declare that there are no conflicts of interest with respect to the publication of this article.

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