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Research Article

# Lemierre's Syndrome and Nasopharyngitis in Children: DIAGNOSIS and Treatment

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## Abstract

Introduction: Lemierre's syndrome (LS) is a rare but serious infectious emergency. It can be life threatening. It is a sepsis difficult to recognize and most often due to Fusobacterium necrophorum. The purpose of this case report was to discuss the diagnosis and treatment of Lemierre syndrome in children. Observation: A three-year-old child consulted for rhinopharyngitis complicated by an abscessed cervical adenophlegmon. The symptomatology had been evolving for 7 days with a notion of self-medication. The blood cell count showed hyperleukocytosis at $14.06.10^3$ elements/$mm^3$, anemia at $8.2.10^6$ elements/$mm^3$, and platelets at $102.10^3$ elements/$mm^3$. CRP was 258.5 mg/dL. HIV serology was negative. Bacteriological culture of the biological fluids collected (abscess pus, blood and joint fluid) was sterile. The CT scan revealed a collection of abscesses in the right sub-angulo-mandibular region. It was associated with thrombophlebitis of the internal jugular vein, as well as pulmonary and joint infections. The diagnosis of Lemierre syndrome was done. Cervicotomy allowed drainage of the abscess. A triple parenteral antibiotic therapy (Ceftriaxone, Metronidazole, Gentamycin) with per-os relay by Amoxicillin + Clavulanic Acid allowed to obtain the cure in one month of treatement without complications. Conclusion: LS is difficult to diagnose due to its non-specific clinical presentation and low incidence. Early clinical and imaging diagnosis guarantees a good outcome by allowing adequate treatment.

## Keywords

Lemierre's Syndrome, Thrombophlebitis, Nasopharyngitis, *Fusobacterium necrophorum*

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## 1. Introduction

Lemierre's syndrome (LS) is defined by the presence of septic thrombophlebitis of the internal jugular vein with sepsis and distant septic emboli (most often pulmonary). It complicates an oropharyngeal infection [1-4]. It was described for the first time in 1936 by André Lemierre [2-5]. It is a rare but serious pathology that can be life-threatening [1, 3, 6]. Its low incidence and the radical change in its prognosis with the advent of antibiotics make it a little-known infection [7]. In children, acute nasopharyngitis is an obligatory adaptation disease. It can be complicated by peripharyngeal suppuration if poorly managed [8]. Through this focus, we can witness the appearance of Lemierre's syndrome [2]. This sepsis is hard to recognize [3, 4, 6]. The diagnosis is late because it is not often mentioned and discovered incidentally [4]. Other less typical clinical forms, less complete or associated with other pathologies, are increasingly described like Lemierre syndrome, malignant tumors, autoimmune diseases, digestive disorders, metabolic disorders which can make diagnosis even more difficult and late [3, 9]. However, this syndrome often has devastating after-effects at the cost of prolonged hospitalization, often in intensive care [10]. Early treatment and therefore diagnosis are essential, and the infectious medical-surgical emergency is absolute [6, 10]. It is a condition rarely reported in Sub-Saharan French literature [4]. The objective of this clinical case was to discuss diagnosis and treatment of Lemierre's syndrome in children.

## 2. Observation

A three-year-old patient visited for right sub-angulo-mandibular swelling. The debut was a week ago associated with odynophagia, cough, and right ear pain. This required unspecified traditional and conventional self-medication. The patient had no specific pathological history. The physical examination revealed 39 ℃, a fluctuating right sub-angulo-mandibular swelling (4 cm) in diameter, hot and painful on palpation. The surrounding skin was unremarkable. There was a painful tightness along the ipsilateral sternocleidomastoid muscle causing torticollis. Anterior rhinoscopy found bilateral crusty rhinorrhea. Tongue depressor examination revealed diffuse inflammation of the pharynx and labial perleche. Otoscopy was normal. The pleuropulmonary examination revealed bilateral crackles. A painful swelling of the right knee was found. The Blood Count had found hyperleukocytosis at $14.06.10^3$ Element/$mm^3$, anemia at $8.2.10^6$ Element/$mm^3$, platelets were at $102.10^3$ Element/$mm^3$. CRP was 258.5 mg/dL. HIV serology was negative. The CT scan showed a collection of abscesses (12 HU) in the right sub-angulo-mandibular region with irregular contours. It measured 53x35 mm (coronal reconstruction) in places. It infiltrated the right parapharyngeal fatty tissues and the ipsilateral sternocleidomastoid region. After injection of contrast product, there was peripheral parietal enhancement with central necrotic hypodensity. There was also an enhancement defect along the entire length of the right internal jugular vein with a thickened, enhancing wall (Figure 1).

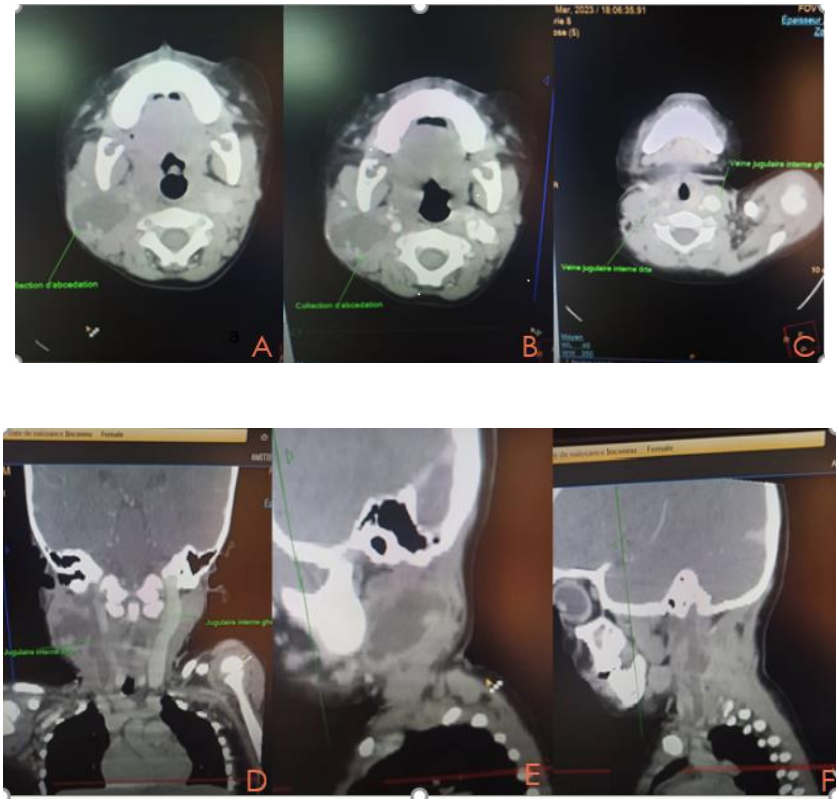


***Figure 1.*** *Craniocervical CT in axial sections (A, B, C), coronal (D) and sagittal (E, F) reconstructions highlighting a collection of abscesses in the right subangulo-mandibular region (A, B, D and E) with thrombophlebitis of the right internal jugular vein (C, D and F).*

Likewise, frank pneumonia of the ventral segment of the left upper lobe associated with foci of alveolar filling of the lower lobes was found (Figure 2). Furthermore, several cervical and axillary lymphadenopathy had been reported.

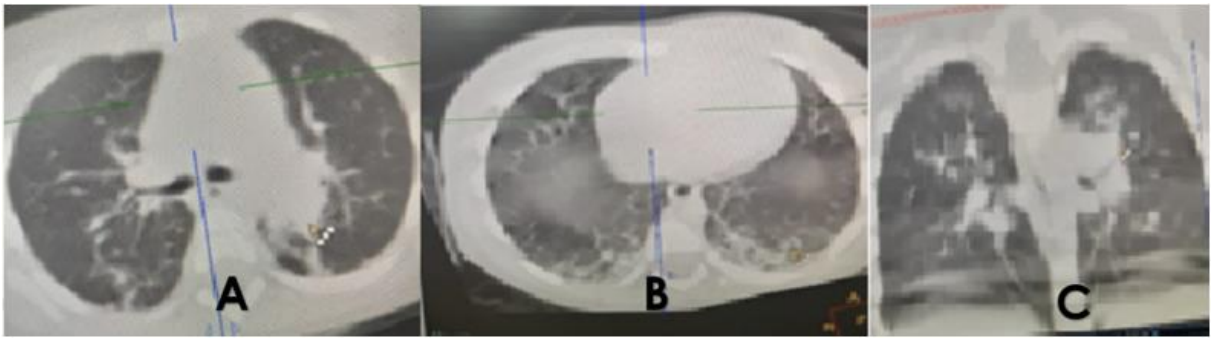


***Figure 2.*** *Thoracic CT in axial sections (A, B) and coronal reconstruction (C) revealing frank pneumonia of the ventral segment of the left upper lobe (A, C) associated with foci of alveolar filling of the postero-basal segments of the lower lobes (B).*

The comparative x-ray of the knees did not find any bone lesions. The cytobacteriological examination carried out after puncture of the puncture fluids from the right knee and the cervical swelling was sterile. It was seo-hematic fluid for the first and thick frank pus for the second. The cervicotomy allowed the drainage of 20 cc of pus. A triple parenteral antibiotic therapy based on Gentamycin, Ceftriaxone, Metronidazole allowed a rapid improvement of the signs after 72 hours. It was continued for three weeks with Gentamycin stopped after five days. The oral relay was provided by Amoxicillin + Clavulanic Acid for seven days. Paracetamol and a splint immobilizing the right lower limb allowed sedation of pain and fever. Nasopharyngeal disinfection was carried out with physiological saline (three times a day for one week). The healing was complete with disappearance of all signs. There were no complications.

## 3. Discussion

Lemierre's syndrome or necrobacillosis or "forgotten disease" [2, 3, 6]. Its incidence is 2.8 cases/million/year in the general population, and 9.4 cases/million/year in the population of young people aged 15 to 24 in Europe [6]. The African incidence is poorly known [5]. It is a pathology of young, immunocompetent male adults in 75% of cases [3, 5, 11]. However, the incidence of infants and children is not exceptional [3]. They represent 20% of cases. LS progresses in three stages [11]. It most often begins with pharyngitis or tonsillitis [2]. Isolated cases in which sinusitis, otitis media or mastoiditis and dental abscess were the primary focus of LS have been reported [2, 5-7]. After a few days, flu-like and anginal-like symptoms appear such as fever, chills, nausea, vomiting, neck pain and cervical lymphadenopathy [2, 10]. In a second phase, the infection continues to progress to the neurovascular system via the parapharyngeal space, which gives rise to thrombophlebitis of the internal jugular vein. In front of the sternocleidomastoid muscle, in the angle of the jaw, a unilateral swelling develops, painful on pressure, and more rarely trismus [2, 5]. Thrombosis of the internal jugular vein can only very rarely be palpated, in the form of a palpable cord in front of the sternocleidomastoid muscle [2, 5, 12]. Damage to cerebral nerves X–XII and Horner syndrome are very rare. During the third phase, from jugular vein thrombophlebitis, dissemination and septic embolism occur, preferably in the lungs (80%), with bilateral nodular infiltrates, lung abscesses and pleural effusions. In 16.5% of cases, joint damage occurs, with arthralgia or septic arthritis. The patient then finds himself increasingly in a state of septic shock, with beginning multi-organ dysfunction and respiratory failure [2, 5]. Clinical manifestations and symptoms vary depending on the location of the septic emboli. It could be an isolated infectious syndrome, pulmonary, neurological, digestive or even joint symptoms... [2]. It occurs on average a few days to 3 weeks after an ENT infection [2, 3]. Cases of infectious endocarditis have also been reported [12]. LS must systematically be part of the differential diagnosis in a child, adolescent or young adult presenting with an oropharyngeal infection which progresses unfavorably, with the appearance of local complications (painful swelling of the cervical region) or a serious septic state not present. controlled [7, 13]. The pathogen most commonly responsible is a Gram-negative anaerobic bacterium, *Fusobacterium necrophorum* (FN) [2, 3, 5, 6]. It is a commensal bacterium of the oropharynx, digestive tract and female genital tract [6, 7]. It can be detected by blood culture which remains the reference examination, or from biological fluids, or by sampling abscessed collections [3, 6]. The negativity of the cultures can be explained by the significant fragility of the germ, particularly during its transport to the laboratory [3, 4, 6]. Infections decapitated by prior administration of antibiotics are also a source of sterile cultures [6]. In blood cultures, FN bacteria are detected after 48 hours in approximately 70% of patients. Direct detection by PCR of FN is favored because of the rapid obtaining of results and the greater specificity and

sensitivity [2]. In 5% of cases, other pathogens such as *Klebsiella pneumoniae* are isolated [2, 4, 5, 9]. Mixed cultures are found in 10 to 30% of cases. A synergistic effect between different pathogens is discussed. Furthermore, scientists also discuss the possibility that LS caused by FN may be favored by a pre-existing infection with the Epstein-Barr virus (EBV), cytomegalovirus (CMV), influenza A virus, or even *Mycoplasma pneumoniae* [2, 6]. Different endotoxins, hemolysins and hemagglutinins cause a weakening of the local immune response, an intense inflammatory reaction and the formation of thrombi [2]. Biologically, the condition is characterized by an inflammatory syndrome. CRP often exceeds 150 mg/dl [2, 9]. There may also be hyperbilirubinemia, elevated liver parameters, increased renal values, electrolyte abnormalities, thrombocytopenia (consumption coagulopathy, disseminated intravascular coagulation) and neutrophilia [2]. The imaging modality of choice is contrast-enhanced computed tomography (CT) of the neck and thorax [2, 3]. It makes it possible to visualize and characterize an endoluminal defect in favor of thrombosis of the jugular vein or one of its collaterals. Locoregional complications as well as distant septic localization, particularly pulmonary, are also objectified [3]. Depending on the clinical picture, it is also possible to perform an abdominal CT if there is suspicion of intra-abdominal organ involvement or a cranioencephalic CT if there is suspicion of intracranial involvement (meningitis, sigmoid sinus thrombosis or of the cavernous sinus) [2]. Cervical venous Doppler ultrasound is a non-irradiating, operator-dependent examination [3]. As an alternative or complement to CT, it has excellent sensitivity [6]. It can contribute to the diagnosis by objectifying a vein increased in diameter, incompressibility of the vein, and/or the absence of flow [3, 6, 12]. As a result, it constitutes an excellent element for monitoring patients [3, 6]. However, it does not allow the identification of subclavicular, retro-mandibular and early thrombi [6, 12, 13]. Cervical MRI allows a precise study of the extension of the thrombus and a mapping of possible loco-regional complications [3, 11]. The 18-fluorodeoxyglucose PET scan allows early diagnosis, especially peripheral septic emboli. It contributes to early treatment as a complement or alternative to conventional imaging. The high costs and availability of these last two examinations constitute limitations in our context [3]. LS is a pathology that can be fatal with mortality estimated at 90% in the absence of early and adequate treatment. Optimal management makes it possible to lower this rate to a percentage between 2 and 10% [2, 3, 6]. The majority progresses towards complete recovery under treatment but can also lead to the death of the patient [6]. Treatment is based on first-line double antibiotic therapy combining a beta-lactam with a beta-lactamase inhibitor or a third-generation cephalosporin, with metronidazole intravenously then monotherapy with imipenem or moxifloxacin orally for a total duration of 3 to 6 weeks [3]. Some authors recommend intravenous monotherapy with clindamycin. For *Fusobacterium necrophorum*, there is resistance to macrolides, quinolones, trimethoprim/sulfamethoxazole, amino-glycosides and partly also to penicillin [2]. Anticoagulation remains highly debated between the risk of thrombus dissemination and retrograde septic extension, so its use must be the subject of multidisciplinary discussion on a case-by-case basis [2, 3, 6]. Indications for anticoagulation are reserved for high-risk situations linked to thrombosis (retrograde septic extension from the internal jugular vein, sigmoid sinus thrombosis, thrombophilia, lack of response to antibiotic treatment, or even cerebral infarction) [6]. Surgical drainage of abscessed collections with abundant washing is of great benefit [2, 3, 6, 9]. It represents an essential part of the care. It makes it possible to eliminate all local sources favoring anaerobic conditions [13]. This treatment allows for a favorable evolution, which was the case for our patient. Ligation-excision of the internal jugular vein remains exceptional and limited to certain cases of progression (persistence of septic emboli under optimal antibiotic treatment, extensive septic thrombosis, or even severe uncontrolled sepsis) [2-4, 6].

## 4. Conclusion

The natural history of Lemierre's syndrome should teach us lessons. The diagnosis is difficult to establish due to a non-specific clinical presentation and a low incidence. Prevention is better than cure. Acute nasopharyngitis in children must be very well managed. Chills are prodromes of serious infections. Treatment begins with raising awareness among the population about the need to quickly seek primary health care. Knowledge of the different phases of the disease must be essential among health professionals. The availability of additional examinations is essential. Reasonable effective antibiotic therapy prevents fatal outcome and after-effects in the majority of cases.

## Author Contributions

**Samaké Djibril:** Conceptualization, Resources, Data curation, Formal Analysis, Supervision, Validation, Investigation, Visualization, Methodology, Writing – original draft, Project administration, Writing – review & editing

**Sidibé Youssouf:** Supervision, Visualization, Writing – original draft

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**Kéïta Mohamed Amadou:** Writing – original draft

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## Conflicts of Interest

The authors declare no conflicts of interest.

## References

[1] Rissoud M., Mortuaire G., Chevalier D., Rysman B. Atypical Lemierre syndrome. European Annals of Otorhinolaryngology, Head and Neck diseases 2016; 133(2): 123–124.

[2] Schahab C. Lemierre syndrome, A partly "forgotten" condition, but which has not disappeared. Swiss Medical Forum 2012; 12(19): 386–388.

[3] Imane A., Sayad Z., Laassikri O., Nitassi S., Bencheikh R., Benbouzid M A., Oujilal A., Boulaadas M., Essakalli Hossyni L. An unexpected cause of forgotten illness: about 'a case. PAMJ Clinical Medicine. 2020; 4(21): 1-9.

[4] Salami A., N'guessan N D., Kadre A., Assouan C., Anzouan-Kacou E., Konan E. Lemierre syndrome: know it to better recognize it. Rev Col Odonto-Stomatol Afr Chir Maxillo-fac 2018; Special: 41-44.

[5] Kewe I., Djomou F., Ze Minkande J., Kenna E., Mbéé P., Meva'a R., Fokouo V., Njdolo A., Bengono G. Lemierre Syndrome Revealed by Otomastoiditis in a Girl 12 years old in Yaoundé Health Science and Disease 2016; 17 (2): 84-87.

[6] Mesrar H., Mesrar J., Maillier B., Kraoua S., Chapoutot L., Delclaux B. Lemierre syndrome: diagnosis, exploration, treatment. The Journal of Internal Medicine 2018; 39: 339–345.

[7] Perreault M. A case of Lemierre syndrome. Pharmacactual 2006; 39(4): 213-217.

[8] Christopher Maroun A., Habib Zalzal G., Ayman Mustafa A., Carr M. Transoral versus Transcervical Drainage of Pharyngeal Abscesses in Children: Post-Operative Complications. Annals of Otology, Rhinology & Laryngology 2021; 130(9): 1052–1056.

[9] Kharoubi S. Lemierre syndrome due to Klebsiella Pneumoniae: Clinical and therapeutic considerations. J TUN ENT 2020; 43: 73-76.

[10] Panchavati P K., Kar B, Hassoun A., Centor R M. Fusobacterium necrophorum tonsillitis with mild cases of Lemierre's syndrome. Anaerobe 2017; 43: 102-104.

[11] Ben Abdallah Chabchoub R., Riquet A., Ramdane A., Vallée L., Raccoussot S. Lemierre syndrome revealed by torticollis. Archives of Pediatrics 2015; 22: 540-543.

[12] Benhammou A., Mimouni H., Nazih N., Boulaadas M., Benbouzid M A., Boulaich M., Essakali L., Kzadri M. Lemierre syndrome secondary to chronic cholesteatomatous otitis media. The ENT and head and neck surgery letter 2009; 318: 29-31.

[13] Assouan C., Salami A., Anzouan-Kacou E., Nguessan N., Konan E. Cervical necrotizing cellulitis revealing Lemierre syndrome. Rev Stomatol Chir Maxillofac Oral Chir 2016; 117: 158-160.