

# LEMIERRE'S SYNDROME: A FORGOTTEN COMPLICATION OF OROPHARYNGEAL INFECTION

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**Background:** Lemierre's syndrome (postanginal sepsis) usually complicates an oropharyngeal infection as septicemia, septic thrombophlebitis of the internal jugular vein, and metastatic lesions, most frequently in the lungs. *Fusobacterium necrophorum* is the usual etiologic agent. Lemierre's syndrome is not widely known by clinicians. This study aims at creating awareness amongst the clinicians of existence of this potentially fatal but curable clinical entity. **Methods:** All the patients admitted in the ENT department of Ayub Teaching Hospital, during the period of January 2000 to December 2002, for the treatment of acute oropharyngeal infection, were critically assessed by consultants, for features of Lemierre's syndrome, and investigated further, accordingly. **Results:** 156 patients were admitted with acute oropharyngeal infection. Two patients (1.28%) had features suggestive of Lemierre's syndrome. A male and a female, 25 and 28 years old respectively, had a history of a preceding sore throat for a variable duration followed by complications. Attempts were made to make the diagnosis of Lemierre's syndrome and they were treated appropriately, as per recommendations, to a successful outcome. **Conclusions:** Widespread use of antibiotics for pharyngeal infections has significantly reduced the incidence. Rare and a forgotten complication, Lemierre's syndrome is potentially fatal. Early diagnosis and prolonged treatment with appropriate antibiotics are usually curative. A high degree of clinical suspicion is necessary for diagnosis. Modern day clinician should be aware of this potentially life threatening clinical entity that may complicate a trivial oropharyngeal infection.

**Keywords:** Lemierre's syndrome; *Fusobacterium necrophorum*; oropharynx; septicemia;

## INTRODUCTION

Post anginal sepsis was first described by Scottmuller in 1918.<sup>1</sup> However, Andre Lemierre, in 1936, described septicemia secondary to oropharyngeal anaerobic infection, on account of his experience with 20 cases, 18 of whom died,<sup>2</sup> and the condition is therefore more commonly known as Lemierre's syndrome.

Limited number of cases have been reported in the literature and currently the rate of disease is 1 per million per year,<sup>3</sup> which is much infrequent than it was in the pre antibiotic era. Mostly it occurs in adolescents and young adults,<sup>4</sup> evenly distributed between the sexes, and the youngest age group that it has been reported in is 5 years.<sup>5</sup>

In the pre-antibiotic era, it had a 90% mortality rate. Now, with the advent of antibiotics, the prognosis is favorable, but delays in diagnosis may result in increased morbidity and mortality. Its incidence appears to be increasing.<sup>6</sup>

The disease usually begins with an acute oropharyngeal infection followed by unilateral jugular vein septic thrombophlebitis, producing neck pain. Subsequently, bacteremia, mostly by anaerobic bacteria, and multiple metastatic pulmonary abscesses resulting from septic emboli develop.<sup>7</sup> Lemierre noted that the syndrome could occur as a complication of otitis media, mastoiditis, or dental infection.<sup>2</sup> Other sites of septic metastatic lesions include the bone and joints, peritoneum, liver, and kidneys.<sup>2</sup> Typical triad of pharyngitis, a tender/swollen neck, and noncavitating pulmonary infiltrates, is commonly seen nowadays.<sup>8</sup>

*Fusobacterium necrophorum* is the anaerobe most often implicated in Lemierre's syndrome, but other fusobacteria, *Bacteroides* species, and anaerobic *Streptococcus* species have also been isolated.<sup>7</sup>

This study was designed to study the frequency of Lemierre's syndrome in patients presenting with acute oropharyngeal infection at Ayub Teaching Hospital, Abbottabad.

Our aim was to increase the clinician's awareness of this clinical entity, as a high index of suspicion is mandatory to diagnose the syndrome. The disease is easy to recognize and is diagnosed exclusively on clinical grounds. A contrast-enhanced CT scan of the neck and/or ultrasonography with Doppler can be used to detect the jugular venous thrombophlebitis, which confirms the diagnosis.<sup>9</sup>

Ultrasonography is recommended for the initial evaluation because it is sensitive, is inexpensive and does not require the use of intravenous contrast material.<sup>10</sup> It can confirm internal jugular vein thrombosis, showing localized echogenic regions within a dilated vessel,<sup>11,12</sup> however, its utility can be limited by the anatomic location of the clavicle and mandible and by the skill of the technician performing the study.

Computerized tomography with intravenous contrast may be the best choice because it is more sensitive than ultrasound, can show the extent of abscess formation and allows visualization of the intrathoracic contents.<sup>13</sup> The only drawbacks are that it requires radiation and intravenous contrast material. A positive study shows a dilated internal jugular vein with low attenuation intraluminal contents and enhancement of the vessel wall and surrounding tissue.

Magnetic resonance imaging offers superb detail of soft tissue structures, no radiation and multiplanar views. Reportedly the accuracy of magnetic resonance imaging is excellent with as high as 97% correlation to venography.<sup>14</sup> Gallium scanning lacks sensitivity and specificity,<sup>13</sup> and nuclear scintigraphy lacks the reliability and ease of the other methods.<sup>15</sup>

Retrograde venography carries the obvious risk of perforation or embolic events and should be reserved only for cases in which the other modalities have failed to show the thrombus.

Confirmation of Lemierre's syndrome is provided by demonstration of *Fusobacterium necrophorum* on blood culture,<sup>4</sup> however negative cultures does not deny the diagnosis.<sup>8</sup>

Recommendations for treatment of Lemierre's syndrome include prolonged, high-dose antimicrobial therapy, ranging from 3 to 6 weeks.<sup>7</sup> The use of metronidazole or clindamycin may be necessary to provide good anaerobic coverage in view of reports of penicillin-resistant organisms, including some *Fusobacterium* species and *Bacteroides* species.<sup>16</sup>

Surgical ligation or excision of the thrombosed jugular vein is usually reserved for cases with persistent embolic phenomena and progression of septicemia despite appropriate medical therapy. Aggressive antimicrobial therapy, along with surgical drainage of purulent collections, is successful in the vast majority of patients.<sup>7</sup> Anticoagulation with heparin is recommended,<sup>15,17</sup> especially in cases of extensive thrombosis or progression on antibiotics alone; it shortens course and often avoids surgery, however, routine use of heparin therapy is controversial.

Although Lemierre's syndrome is now rarely seen, physicians should become familiar with this potentially life-threatening condition, which necessitates early clinical recognition and prompt initiation of appropriate therapy.

## **MATERIAL AND METHODS**

This study was conducted in the ENT, Head & Neck Surgery department of the Ayub Teaching Hospital, Abbottabad, during the period of January 2000 to December 2002.

All the patients admitted with acute oropharyngeal infection were included in this study and critically assessed for signs of infection spreading into the neck and metastatic lesions.

Patients with neck space infections in the absence of features of oropharyngeal infection were excluded. Patients included in the study were assessed and the clinical features as described by Lemierre, were looked for. Patients with suspicion of the syndrome had total white cell count and differential white cell count to confirm the presence of infection. Plain chest radiograph was done to check the lungs status. Ultrasonography of the neck was employed as a quick diagnostic tool for the presence of infection in the neck and to check the patency of the internal

jugular vein in suspicious cases, however computerized tomography with intravenous contrast was utilized to confirm or deny the presence of disease in the neck veins and lungs, in suspicious cases.

Data was manually analyzed to calculate frequency of Lemierre's syndrome in our group of patients.

## **RESULTS**

During the period of January'2000 to December'2002, 156 cases were admitted to the ENT department with acute tonsillopharyngitis. 37 cases presented to the out patient department and 119 cases were admitted through casualty department. 88 (56.41%) were male and 68 (43.58%) female patients. Age ranged from 08 years to 70 years, however, majority of the patients were young adults.

Two patients (1.28%), a male and a female, had features suggestive of Lemierre's syndrome. They had a history of a preceding sore throat for a variable duration followed by complications in the neck.

The first case was a 25 years old man admitted with history of acute sore throat for a week followed by development of a swelling in the left neck that was progressive and painful. He had odynophagia of moderate degree and restricted neck movements. He had clinical signs of acute tonsillopharyngitis and neck examination revealed a firm and tender induration of the left neck without any fluctuation (Fig.1). He had pyrexia of 102°F. White cell count was elevated with polymorph leukocytosis and ESR was elevated to 35mm. Blood culture was negative however, throat swab cultured bacteroides and pseudomonas. Plain X-ray neck demonstrated soft tissue edema on the left side. Chest radiology was unremarkable, whereas computed tomography with contrast showed soft tissue swelling and thrombosis of the internal jugular vein in the neck (Fig.2). He was treated with appropriate antibiotics, intravenously, along with supportive therapy. His condition improved over the next five days to the extent that the neck induration and pain had resolved and so was his pharyngeal infection. He was able to take food and was discharged on day 7, on oral antibiotics for further two weeks. On follow-up he was asymptomatic and there were no clinical signs of infection in the throat and neck..

**Fig-1: A firm and tender induration on left neck**

### **Fig-2: Jugular Vein thrombosis**

The second case was a 28 years old lady referred from a rural health center with suppurating left neck swelling. She had a history of severe sore throat of two weeks duration and left neck swelling of one-week duration. The neck swelling was high initially but within days it significantly increased in size and became more obvious in the lower part of the neck (Fig.3). She had severe pain in the neck with restricted movements. The neck swelling became soft over the days and started discharging two days before she was referred into our unit. Clinically, she had acute tonsillitis (Fig.4) and a fluctuant left neck swelling that was discharging pus through a small opening and had concentrated in the lower left neck. She had polymorph leukocytosis and elevated ESR, however, blood culture was negative. Surgical drainage of the abscess was achieved and intravenous antibiotic therapy to cover anaerobes instituted. Plain X-ray neck showed soft tissue swelling with trapped air, whereas, chest radiology was unremarkable. Contrast enhanced computed tomography revealed soft tissue edema in the neck and thrombosis of the internal jugular vein. She had a variable response over the next few days with spiking pyrexia and eventually settled down by the end of first week. She was sent home on oral antibiotics for two weeks. On review she had completely healed and was asymptomatic.

**Fig-3: Suppurating swelling left side of neck**

**Fig-4: Acute Tonsillitis**

**Table-1: Laboratory and Radiological findings in the two cases**

**Laboratory Findings**

Case	TLC	DLC	ESR mm/1hr	Urea/ electrolytes	Blood culture	Throat swab
1	9000	N: 66% L: 28% M: 05% E: 01%	43	Normal	-ve	Bacteroides Pseudomonas
2	7800	N: 72% L: 18% M: 08% E: 02%	36	Normal	-ve	Negative

## Radiological Findings

Case	Plain x-ray neck	Plain chest x-ray	Contrast enhanced CT scan
1	Soft tissue edema	Normal	IJV thrombosis
2	Soft tissue edema with air fluid level	Normal	IJV thrombosis

## DISCUSSION

Lemierre's syndrome is not widely known by clinicians. It appears to be a forgotten complication that is potentially fatal.

This study emphasizes the importance of awareness about this condition, as it is relatively easy to make a diagnosis on the basis of clinical findings alone.

We supported our diagnosis by the presence of severe oropharyngeal infection not responding to empirical treatment and subsequently presenting with complications in the neck. Culture studies were negative in our patients and so has been reported in the literature.<sup>9</sup>

Furthermore, neck ultrasonography and contrast enhanced computerized tomography of the neck and chest were employed to help us clinch the diagnosis in our cases, however, magnetic resonance imaging would be a better choice, if available.

Following the treatment recommendations in the literature, our cases had a successful outcome. Our finding of two cases amongst 156 cases in no way clashes with a reported incidence of 1 in a million per year as our study population was totally different and consisted of all diseased persons in contrast with the population suggested by Hagelskjaer et al.<sup>3</sup>

## RECOMMENDATIONS

Clinicians should become aware of this potentially life threatening condition that necessitates early recognition and management. If we keep in mind the nature of this rare complication, it becomes relatively easy to make a diagnosis on the basis of clinical findings

- alone.
- Mortality is uncommon, however, significant morbidity persists.
- Modern techniques should be employed early to diagnose the condition.
- Prolonged appropriate antibiotic therapy coupled with surgery, usually, provides a successful outcome.

It must be remembered that this forgotten disease is not extinct

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