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Lemierre's syndrome following perianal abscess: A case report

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ABSTRACT

INTRODUCTION: Lemierre's syndrome (LS) is a rare and life-threatening condition characterized by suppurative thrombophlebitis of the internal jugular vein (IJV), and a history of head and neck (H&N) sepsis. LS is usually caused by *Fusobacterium necrophorum*, which is part of the normal flora in the oro-pharynx, and the digestive and urogenital tracts. We here report the first case of LS following perianal sepsis.

PRESENTATION OF CASE: A 60-year-old man with a painful left neck swelling, dysphagia and worsening sepsis was referred from a peripheral unit where he had an incision and drainage of a perianal abscess a week earlier. Urgent Doppler ultrasound and computed tomographic scans demonstrated suppurative thrombophlebitis of the left IJV, and the patient was subsequently commenced on intravenous Piperacillin/Tazobactam and heparin. The symptoms gradually improved, and the patient was eventually discharged on the 10th day.

DISCUSSION: Vigilant examination of the H&N region searching for a primary source is paramount, but LS following infections in the gastrointestinal or uro-genital tracts has also been described. A high index of suspicion is required for diagnosis, especially in patients with unresolving pharyngitis with a unilateral neck swelling, and septicaemia. Early resuscitation and treatment with broad-spectrum parenteral antimicrobials are important for favourable outcome.

CONCLUSION: LS is well known to specialists in the H&N region, but other disciplines like general surgery, urology, or obstetrics and gynaecology might also rarely encounter the disease. We present a case of LS complicating a perianal abscess that was successfully treated with good outcome.

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1. Introduction

Lemierre's syndrome (LS) is a life-threatening condition characterized by suppurative thrombophlebitis of the internal jugular vein (IJV), and a history of head and neck sepsis [1]. LS is exceptionally rare with a reported incidence of less than one in million, and is often missed earning the reputation of 'The forgotten disease' [2–4]. The condition was named after the French bacteriologist Andre Lemierre, who in 1936 described a series of post-anginal septicaemia cases and confirmed the relationship with *Bacillus funduliformis* (*Fusobacterium necrophorum*) [1]. *F. necrophorum*, is a gram negative, anaerobic bacillus which is part of the normal flora of the oropharynx, gastrointestinal tract, and the female urogenital tract [5]. Infections from the head and neck region are the most commonly reported causes for LS, but other cases have been reported following urogenital and gastrointestinal tract infections

[1,3,5]. Based on the Surgical Case Report Guidelines (SCARE) [6], we here report the case of a 60-year-old male presenting with LS following a perianal abscess surgery. To the best of our knowledge, no similar cases have been previously reported.

2. Case report

A 60-year-old man with a painful unilateral neck swelling, progressive dysphagia and worsening sepsis was referred to our tertiary referral centre from a peripheral unit. The patient had undergone an incision and drainage of an uncomplicated perianal abscess at the referring hospital a week earlier. He did not have any recent sore throat or other co-morbidities, and had no smoking history. On the fourth day after the surgery, the patient started complaining of painful neck swelling, dysphagia and worsening odynophagia. He was being treated with an oral course of amoxicillin/clavulanic acid (500/125 mg/8 h) and pain killers. At admission to our centre, the patient was septic with a temperature of 38.5 ° and a heart rate of 115 beats/minute. The patient did not have any history of invasive procedures to his neck. Local

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Fig. 1. Picture of the patient's neck showing left-sided swelling without notable cellulitis changes in the overlying skin.

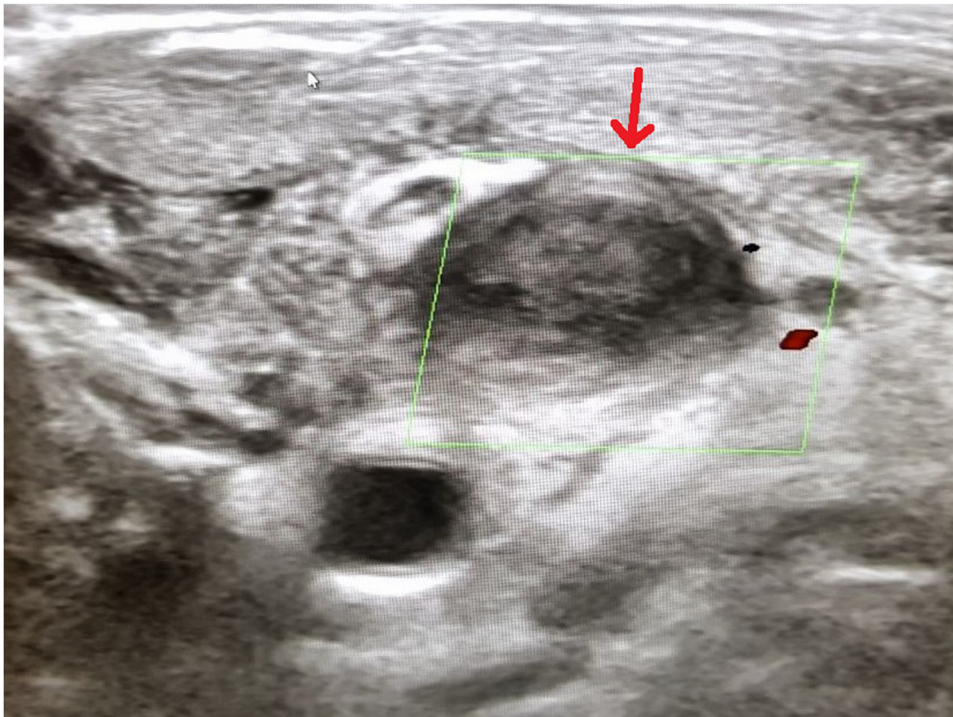


Fig. 2. Colour flow Doppler ultrasound of the neck showing a large echogenic thrombus in the left internal jugular vein (red arrow).

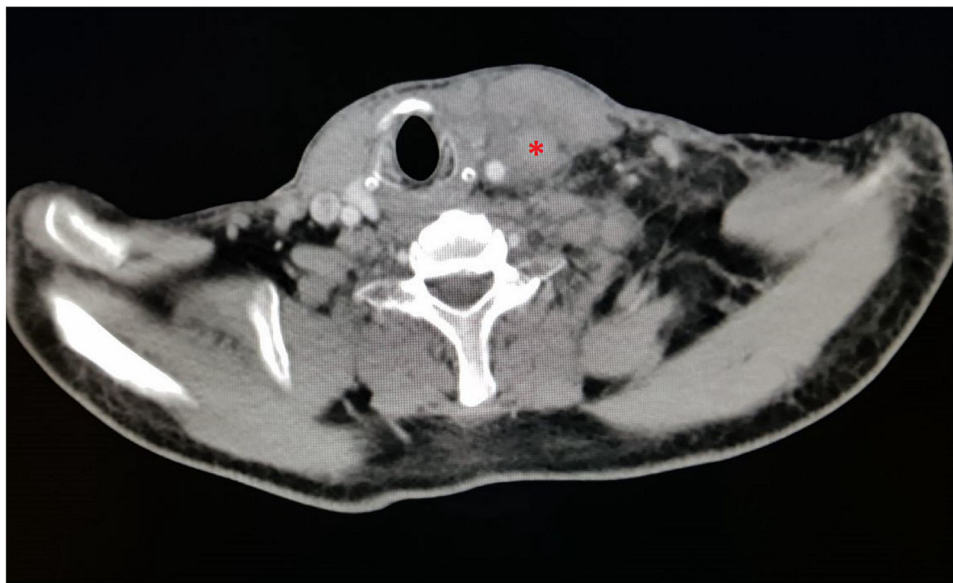


Fig. 3. Computed tomographic scan (axial view) of the neck showing a large filling defect completely obstructing the left internal jugular vein (Asterisk).

examination of the neck revealed normal skin appearance with a soft, tender, non-pulsatile swelling in the left anterior triangle (Fig. 1), and a palpable cord-like structure at the anterior margin of left sternocleidomastoid muscle. Following detailed examination by the oral and maxillofacial (OMFS), and ear, nose and throat (ENT) surgery teams, no primary source of infection was found in the head and neck region. Subsequent examination of the perianal region demonstrated a clean and healing wound, with no signs of local inflammation or ongoing infection. Laboratory investigations demonstrated leucocytosis of $16.4 \times 10^9/L$, raised C-reactive protein (CRP) of 389 mg/L, and elevated D-Dimer. The liver, renal and coagulation biochemical screen was normal. Pus cultures from the perianal abscess did not show any bacterial growth. An urgent colour flow Doppler (CFD) ultrasound of the neck revealed a non-compressible and completely thrombosed left IJV as shown in Fig. 2, but no underlying neck collection was found. A computed tomographic (CT) scan with intravenous (IV) contrast confirmed the findings of suppurative thrombophlebitis of the left IJV, Fig. 3. The diagnosis of LS was provisionally made, and intensive treatment with broad-spectrum Piperacillin/Tazobactam 4.5 g intravenously every 8 h was started. Blood cultures isolated the classical *F. necrophorum*, and hence LS diagnosis was confirmed.

The patient improved markedly with supportive therapy and parental antibiotics. However, on the 4th post-admission day the patient developed progressive heaviness, dull ache and swelling in his left arm. Another CT scan with contrast demonstrated extension of the thrombus to the mediastinum with involvement of the left subclavian vein, Fig. 4. The patient was started on therapeutic dose intravenous heparin with strict arm elevation. The symptoms gradually improved, and subsequently discharged on day 10 on oral amoxicillin/clavulanic acid (500/125 mg/8 h), and metronidazole (400 mg/8 h) for a week, and treatment dose rivaroxaban anti-coagulation for 45 days. Clinic review three months later combined with CFD neck ultrasound showed complete resolution of the disease.

3. Discussion

Various epidemiological studies demonstrated that Lemierre's syndrome affects young patients primarily, typically in those aged 18–20 years, and does not appear to have a gender predominance

[7]. Following the introduction of antibiotics in the 1940s, the disease incidence reduced markedly, but has become more eminent in published literature over the last decade [2,3,5,8]. This resurgence of LS might be attributed to a more conservative attitude in antibiotics prescription, the emergence of drug-resistant microorganisms, and the recent advances in diagnostic modalities facilitating early detection of IJV thrombosis.

Vigilant head and neck examination looking for the primary source of infection is paramount, but in around one quarter of the patients a head and neck source cannot be found [3,9]. Many reported cases highlighted other sources of primary infection, such as the gastrointestinal and urinary tract infections and endometritis [3–5]. Perianal sepsis is a common general surgical presentation [10] but there are no existing reports of its association with LS. The pathophysiology of a classical LS starts with a suppurative infection in the head and neck area, with subsequent involvement of the IJV tributaries and main trunk [11,12]. The symptoms of the infection in the primary site usually start to improve before the neck symptoms of rapidly progressive odynophagia, dysphagia and unilateral neck swelling worsen. Septicaemia usually worsens with the dissemination of septic micro-emboli to the lungs, joints and other organs [5,9]. Retrograde progression of the IJV thrombus into the sigmoid or transverse sinuses may result in meningitis or cerebral abscesses [3].

It is still unclear how an infection in a primary site outside the head and neck area, such as in our case or other cases reported, can lead to a suppurative localised IJV thrombosis. Our patient had atypical course of events compared to those with classical LS, and he did not have any preceding symptomatic head and neck infection. A thorough head and neck examination done by ENT and OMFS specialists did not reveal any signs of infection. Negative pus swabs taken from the abscess site made it more challenging to prove the link between perianal suppuration and LS. Intra-operative bacteraemia caused by the drainage and curettage of the abscess wall might have led to dissemination of *F. necrophorum* micro-emboli. However, the possibility of a subclinical or missed head and neck infection causing LS in our patient cannot be entirely ruled out.

A high index of suspicion is required for the diagnosis. The disease should be suspected in patients with unresolving pharyngitis, a rapidly waning course with a unilateral neck swelling, and systemic illness. *F. necrophorum* has been cultured in 81.7% of cases¹³, whereas a variety of gram negative, gram positive and anaerobic



Fig. 4. Computed tomographic scan (coronal view) of the thoracic outlet showing extension of the thrombus to the left subclavian (red arrow) and brachiocephalic veins, surrounded by fat stranding indicating an inflammatory process. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

microorganisms have been implicated in other cases [3]. LS is a serious disease with high mortality approaching 20% if not treated promptly [3]. Septic micro-embolisation to the lungs is the commonest complication, occurring in around 70% of cases, but other organs like the brain can be affected with devastating consequences [8]. Radiological investigations are essential for confirmation of diagnosis, defining the extent of the disease and identifying the development of complications. The use of CT scanning with IV contrast is considered the best diagnostic modality [11], although CFD ultrasound remains a valuable test for diagnosis and follow-up [14].

Treatment with broad-spectrum parenteral antimicrobials is the most important step in the management of the disease. Most authors recommend a combination of penicillin and metronidazole for a duration of at least 3 weeks [2,13]. Blood cultures play an important role in guiding the antimicrobial treatment, but negative results are encountered in up to 10–20% of cases [13]. Intravenous hydration is also essential since affected patients suffer from dysphagia and hence they are commonly dehydrated at presentation. Most cases respond to antimicrobial therapy and the role of surgery is nowadays restricted to drainage of purulent fluids and debridement of necrotic tissues. Invasive surgical treatment with ligation and resection of the IJV can be life-saving, but only required in 8% of cases [13,15]. The role of anticoagulation for thrombus control in LS is still controversial. Some authors believe that the disease course is not altered by anticoagulation, and hence routine use of anticoagulants in LS is not necessary [7,16]. However, anticoagulation may be indicated in cases of ante- or retrograde thrombus extension, and in thrombophilic patients [17].

4. Conclusion

Lemierre's Syndrome is a rare but potentially fatal disease if not diagnosed and treated promptly. LS can rarely occur following

infections outside the head and neck region, and specialists in other areas like general surgery, urology, or obstetrics and gynaecology should have high index of suspicion especially with classical progressive neck symptoms following infections in the gastrointestinal or uro-genital tracts. Close clinical follow-up is required and anticoagulation therapy is indicated in cases of thrombus extension, but the duration of anticoagulation remains uncertain. In the case presented, the patient was diagnosed with LS complicating a perianal abscess with no preceding head and neck symptoms. Following the appropriate intensive medical treatment, our patient was eventually discharged with a favourable outcome.

Conflict of interest

None to declare.

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Ethical approval

As the publication is for case report, it was exempt from ethical approval in our institution.

Consent

Written informed consent has been obtained from the patient for publishing this case and the accompanying images.

Author contribution

1-Qusai Aljarrah: Article conception, data collection, writing the paper, reviewing and critiquing the manuscript, corresponding author.

2-Yara Khazaleh: Writing the paper.

3-Mooath Al-Jarrah: Data collection.

4- Jordan Oldbury: Writing the paper.

5- Ahmad K. Abou-Foul: Writing the paper, reviewing and critiquing the manuscript.

Guarantor

Qusai Aljarrah.

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None to declare.

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