CASE REPORT

Lemierre's syndrome: a rare cause of sepsis presenting with an absence of throat symptoms

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SUMMARY

A 16-year-old boy presented to hospital with a 6-day history of diarrhoea, vomiting and abdominal pain. During his admission he was found to be hypotensive, tachycardic and persistently feverish. Blood cultures taken on admission isolated *Fusobacterium necrophorum*. CT scanning of his neck showed a non-occlusive thrombus of the right internal jugular vein and a small right parapharyngeal abscess. CT scans of the chest and abdomen revealed multiple pulmonary abscesses, bilateral pleural effusions and splenomegaly. Treatment consisted of an unfractionated heparin infusion and intravenous antibiotics. A right-sided intercostal drain was inserted for a complex right-sided empyema. He subsequently developed a left-sided pleural effusion which was treated with a video-assisted thoracoscopic surgery (VATS) pleurodesis procedure. His fever resolved after his VATS pleurodesis procedure 3 weeks after initial presentation. Clinically he made a slow recovery but now is improved after 6 weeks of intravenous antibiotics and was discharged home.

BACKGROUND

Lemierre's syndrome is rare, characterised by internal jugular vein (IJV) thrombosis with septicaemia usually precipitated by oropharyngeal infection.^{1,2} Patients can then develop disseminated infective thromboembolic disease which can present in numerous ways; this presentation was particularly interesting as the patient had an absence of throat symptoms both at presentation and during admission. On retrospective questioning, the patient did recall a mild sore throat a week prior to admission.

Sore throat, neck pain and neck mass are the most common presenting complaints in Lemierre's syndrome.¹ Gastrointestinal symptoms are not uncommon however are rarely recorded as sole presenting complaint.² There are only few recorded cases of patients presenting with solely gastrointestinal symptoms in the absence of throat symptoms.

Lemierre's syndrome has become relatively rare since 1940s and the regular use of antibiotics for sore throat, one report even referring to it as the 'forgotten disease'.³ Despite this it is a diagnosis that the practicing otolaryngologist considers in some cases of throat sepsis.

On referral to secondary care, this patient had no throat symptoms and thus did not present to otolaryngology. This case helps to highlight Lemierre's disease as a differential in patients with evidence of sepsis without an obvious source. The case should

be highlighted to a wide range of specialties within medicine given the possibility for unusual and multisystem presentations.

CASE PRESENTATION

A 16-year-old boy presented to hospital with a 6-day history of diarrhoea, vomiting, fever and upper abdominal pain. He denied any other symptoms on systemic enquiry.

On arrival he was feverish at 39.0°C, tachycardic at 114 beats per minute, with a blood pressure of 105/65 mm Hg. His oxygen saturations were 99% breathing room air and he was tachypnoeic with a respiratory rate of 24 breaths per minute. His ECG showed sinus tachycardia. An admission chest X-ray suggested a developing right-sided pleural effusion.

On examination, he was clinically dehydrated, with a clear sounding chest, normal heart sounds and a soft abdomen with generalised tenderness in left and right upper quadrants and epigastrium.

He had no medical history of note, took no regular medications and had no drug allergies.

The patient reported that other children at his school had a similar clinical picture recently.

Figure 1 Coronal Ct scan showing right internal jugular vein thrombosis.

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Figure 2 Coronal CT showing the right para pharyngeal abscess.

INVESTIGATIONS

Initial blood tests revealed raised inflammatory markers with C reactive protein of 195 mg/L, leucocytosis of $15.0 \times 10^9/L$ and platelet count of $51 \times 10^9/L$. Liver function tests were also mildly deranged with bilirubin $51 \mu\text{mol/L}$, aspartate aminotransferase 130 U/L and alanine aminotransferase 92 U/L. Renal function was impaired with uraemia of 11.6 mmol/L, creatinine $102 \mu\text{mol/L}$, hyponatraemia 124 mmol/L and chloride 82 mmol/L.

His blood film showed neutrophilia with slight left shift, toxic granulation and vacuolation and a genuine thrombocytopenia.

Blood cultures taken on initial presentation grew a *Fusobacterium necrophorum*.

CT scanning of his neck, chest and abdomen revealed evidence of: right IJV thrombosis (figure 1) with a right parapharyngeal abscess measuring 10 mm $\times$ 17 mm $\times$ 20 mm (figure 2). There is evidence of bilateral evolving pulmonary abscesses (figure 3) with bilateral pleural effusions.

He subsequently underwent pleural aspiration of right-sided effusion which showed a hydrogen ion concentration of 5.5 and a lactate of 5.8 with lactate dehydrogenase of 3145 IU/L. This was treated as an empyema and he had a chest drain inserted.

DIFFERENTIAL DIAGNOSIS

Haemolytic uraemia syndrome was the initial differential diagnosis suspected due to the clinical history of infectious diarrhoeal symptoms with other children at school being affected and his biochemical blood picture. He was subsequently started on intravenous fluids replacement. Antibiotics were started for fever of unknown origin on day 3 of admission.

Due to the absence of throat signs it is likely that Lemierre's syndrome would not have been suspected so early if it were not for *F. necrophorum* detected in blood cultures.

Figure 3 Coronal CT scan showing bilateral pulmonary abscesses.

TREATMENT

Treatment consisted of intravenous fluid replacement. Intravenous antibiotics, started on day 3 of admission, were gentamicin, amoxicillin and metronidazole. This was subsequently changed to 2.4 g of benzylpenicillin four times a day and metronidazole 500 mg three times a day as per specialist microbiology advice after *F. necrophorum* was isolated from blood cultures.

An heparin infusion was commenced once the IJV thrombosis and septic emboli had been identified on CT scanning. This anticoagulation was subsequently stepped down to dalteparin 10 000 Units daily.

During admission, the patient required insertion of a right intercostal drain for drainage of right-sided empyema. Unfortunately he continued to develop a large left-sided effusion which was treated with a - assisted thoracoscopic surgery (VATS) pleurodesis procedure by the cardiothoracic team.

An otolaryngology opinion was sought following the CT scanning findings of parapharyngeal abscess. Given the small size of the parapharyngeal collection, surgical drainage was deferred to intravenous antibiotic management in the first instance with a low threshold for surgical management should the abscess increase in size.

OUTCOME AND FOLLOW-UP

The patient was treated with intravenous antibiotics over a period of 6 weeks, his fever resolved on week 3 of his admission after the VATS pleurodesis procedure. He was discharged on oral antibiotics; amoxicillin (1 g three times a day) and metronidazole (400 mg three times a day) to continue for a further 2 weeks. He was also discharged on rivaroxaban 15 mg two times

a day. Clinically he is much improved with no ongoing evidence of systemic infection.

He is to be reviewed as an outpatient following a repeat MRI scan of his neck and his chest with a view to assessing the right IJV thrombus and discontinuing his anticoagulation.

DISCUSSION

Lemierre's syndrome is defined by a pharyngeal infection, complicated by septicaemia and IJV thrombosis followed by septic emboli.³ Given the sequelae of the septic emboli, patients often require intensive care support.⁴ Lemierre's syndrome is known to affect a large age range of the population however the majority of cases occur between 13 and 40 years.⁵ Even when treated appropriately, mortality is reported to be around 2%.⁶

Metastatic infective thromboembolic disease occurs commonly, with the reported incidence being 63%–100%.^{3,6} The lungs are the most common site but others include major joints, muscles and pericardium. As a result, the clinical presentation may vary greatly.³

Fever is the most common clinical finding in the patient with Lemierre's syndrome and is reported in 92%–100% patients,³ this is however non-specific and could potentially represent a number of pathologies. Pharyngitis, peritonsillar abscess and neck mass are the next most common findings and are more specific to Lemierre's syndrome with limited other differentials. An otolaryngologist will be suspicious of Lemierre's syndrome in patients with severe sepsis from tonsillitis or quinsy that is not responding to antibiotic therapy. It is very rare to have complete resolution of pharyngitis symptoms prior to symptoms from metastatic infection.⁶ This can mask the diagnosis. In our case report due to the absence pharyngitis symptoms and primary presentation with diarrhoea and vomiting, the diagnosis of Lemierre's syndrome was not initially clear and suspicions were raised only when the blood culture results were available.

The evidence for the role of anticoagulation in treatment of Lemierre's syndrome is inconclusive.^{6–8} In this disease, the risk of disseminated infection as a result of septic emboli particularly to the lungs is high. This can lead to cavitating lung lesions and empyema despite appropriate antibiotic treatment, as evidenced by this case. Anticoagulation treatment was therefore commenced in an attempt to stop the patient developing respiratory failure from further septic emboli.⁶ Some authors have suggested commencing anticoagulation therapy if little clinical improvement within 2–3 days of starting treatment or evidence of IJV thrombosis propagation.^{8,9} However due to limited research there are no set guidelines on the benefit of anticoagulation, when it should be commenced and the duration or type of treatment if commenced. Despite this it has been reported

that as high as 64% of cases are treated with anticoagulation in a recent systematic analysis.⁶

In conclusion, this case demonstrates the wide variety of presentations of Lemierre's syndrome and the resultant difficulty with initial diagnosis. It is important that a wide range of subspecialty doctors should have knowledge of this condition.

Learning points

- ▶ To have a low index of clinical suspicion for Lemierre's syndrome in an unwell patient presenting with upset gastrointestinal tract.
- ▶ Inclusion of Lemierre's disease in differential diagnosis in a patient with sepsis even in the absence of throat symptoms.
- ▶ To have a low threshold for neck imaging in a patient with sepsis if *Fusobacterium necrophorum* is isolated from blood cultures.

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