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A persistent sore throat

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Case study

A previously healthy female, 18 years of age, presented with a 10 day history of multiple symptoms including sore throat, vomiting, diarrhoea, cough, lethargy, neck pain and fever.

She had been seen earlier in the course of this illness and had a negative rapid antigen test for group A *Streptococcus* performed as part of the evaluation. Since that visit, her symptoms had progressively worsened. On first examination her vital signs were:

- temperature 37.3°C
- pulse 120 bpm
- blood pressure 100/58 mmHg
- pulse oximetry of 93% on room air.

Her oral examination showed some mild erythema in the posterior pharynx. She had left neck swelling and tenderness in the left submandibular region.

White blood cell count was $20 \times 10^9/L$. A chest X-ray revealed extensive consolidation in the lower lungs, multiple small nodules and a small right pleural effusion. Computerised tomography (CT) of the chest was performed (Figure 1a, b).

Question 1

What is the most likely diagnosis?

Question 2

What age group is most commonly afflicted with this syndrome?

Question 3

What is the cause?

Question 4

What are the typical clinical findings in this illness?

Question 5

How would you diagnose this condition?

Question 6

How would you treat this patient?

Answer 1

Lemierre syndrome, which is septic thrombophlebitis of the internal jugular vein, is the most likely diagnosis. This entity is relatively rare with a reported incidence of one in 1 million people, although incidence may be increasing.¹ There is a reported mortality rate of 5%.²

Answer 2

Lemierre syndrome is seen most often in previously healthy teenagers and young adults, although any age group can be affected.²

Answer 3

Lemierre syndrome is most commonly caused by *Fusobacterium necrophorum*. It classically arises from oropharyngeal infections, especially tonsillitis and pharyngitis,² although it can occur in conjunction with mastoiditis, dental infections, sinusitis, parotitis and otitis.³

Answer 4

The thrombophlebitis of Lemierre syndrome typically occurs 4–8 days after the onset of a tonsillopharyngeal infection, however, it can occur up to 2 weeks later.³ Patients often present with a history of a sore throat followed by a tender, unilaterally swollen neck. Swelling may be located in the angle of the jaw or anterior to the sternocleidomastoid muscle. Patients usually have fever and rigors. Oral examination can



Figure 1a. Axial contrast enhanced chest CT in this patient. Multiple bilateral pulmonary nodules, many of which are cavitory (arrows) consistent with septic emboli. The largest nodule is located in the left lung base measuring just over 3 cm



Figure 1b. A loculated pleural fluid collection is present with mild pleural enhancement (asterisks) consistent with empyema

show signs of pharyngitis or tonsillitis, but may also be entirely normal.

Septic emboli are common and most frequently occur in the lungs and the joints. Pulmonary septic emboli may manifest as cough, haemoptysis, pleuritic chest pain and shortness of breath. Chest X-ray typically shows multiple bilateral nodular infiltrates. Pleural effusion is frequently present, lesions may quickly cavitate and empyema can occur. Emboli to the joints present as a septic arthritis. Less commonly, septic emboli can spread to the bone, soft tissue, liver, spleen and kidneys.

Answer 5

Diagnosis is based on clinical history, culture results and demonstration of thrombophlebitis of the internal jugular vein. Both ultrasound and CT with contrast can reveal internal jugular thrombophlebitis. Ultrasound has the benefit of avoiding radiation, but has a lower sensitivity than CT as it may miss thrombophlebitis located

below the clavicle or inferior to the mandible.¹ This patient had a magnetic resonance venogram (MRV) to assess the extent of the thrombus (Figure 2).

Blood and any other purulent material, such as that collected from a septic joint, empyema or other septic emboli, should be cultured. The most common causative organism is the anaerobic *F. necrophorum*. It has been suggested that if blood cultures grow this organism, consideration should be given to imaging the internal jugular vein to evaluate for internal jugular vein thrombophlebitis.³

Answer 6

Lemierre syndrome is considered a medical emergency. Patients should be admitted to hospital and started on intravenous antibiotics. *F. necrophorum* is generally sensitive to penicillin, but beta lactamase producing organisms have been reported, thus treatment with penicillin alone is not advised. First line treatment with penicillin and metronidazole has been found to be effective. Clindamycin can be used as an alternative.² Use of ticarcillin clavulanate, ampicillin sulbactam, cefoxitin and amoxicillin clavulanate has also been described.⁴ The role for anticoagulation in the treatment of Lemierre syndrome is uncertain though it is often avoided unless there is extension of thrombus into the cavernous sinus.¹ Unfortunately, due to



Figure 2. A multiple intensity projection gadolinium enhanced MRV of the head demonstrating thrombus extension (arrows) into the left sigmoid sinus, to the junction of the left transverse sinus

the very low incidence of Lemierre syndrome, it is unlikely that controlled trials will be performed in order to address the role of anticoagulation. Surgical therapy, when indicated, includes drainage of abscesses and, rarely, ligation or excision of the internal jugular vein. The latter procedure is reserved for patients with persistent septic emboli despite appropriate antibiotic therapy.⁴

Case follow up

The patient was admitted to hospital and started on intravenous antibiotics and heparin. She required bilateral chest tubes for loculated effusions. She was ultimately changed to oral antibiotics (cefepodoxime and metronidazole) which she continued for 7 weeks. Due to her thrombus extending into the sigmoid sinus, she received anticoagulation for a total of 5 months. On follow up the patient achieved a full clinical recovery without any sequelae.

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