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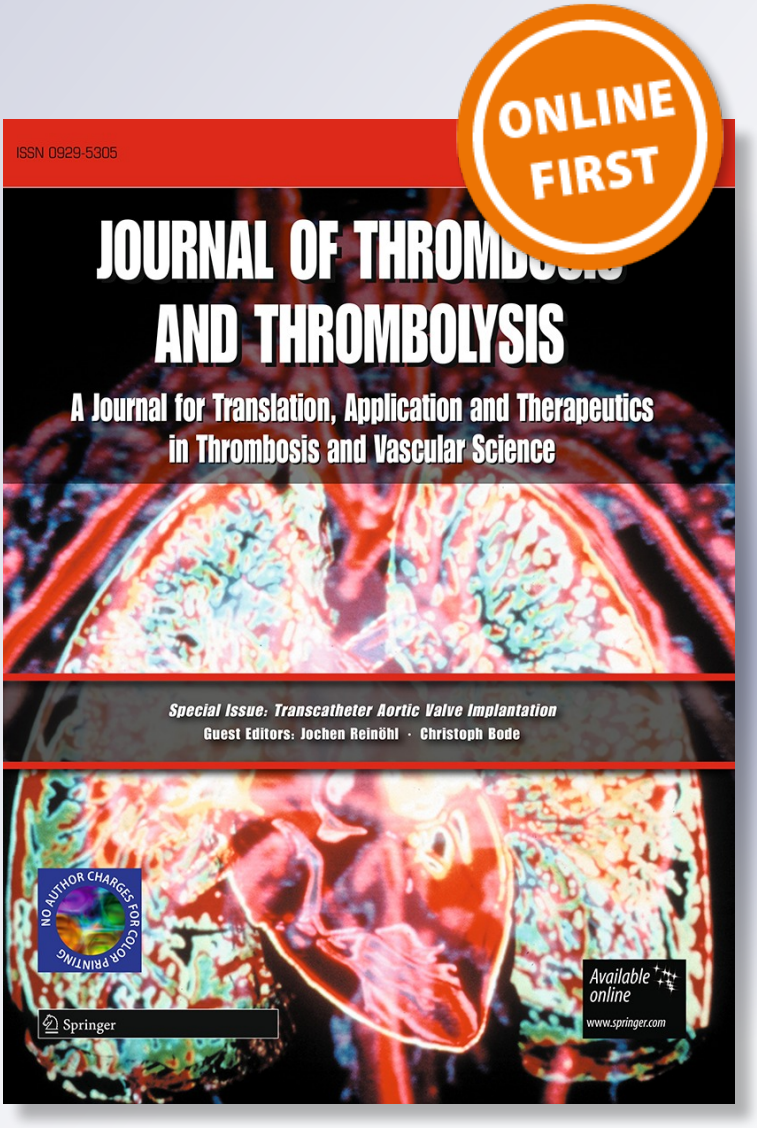

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Lemierre's syndrome: the forgotten disease

Katrine Johannesen · Uffe Bødtger · Ole Heltberg

Abstract Lemierre's syndrome is an often un-diagnosed disease seen in previously healthy young subjects, presenting with symptoms of pharyngitis, fever and elevated markers of inflammation. The syndrome is characterised by infectious thrombosis of the jugular vein due to infection with *Fusobacteria*, causing a variety of infectious complications. Rapid diagnosis and treatment is necessary to avoid severe complications or death. Close collaboration with local microbiologist is pivotal. Treatment consists of longterm treatment with penicillin and metronidazole. This is a case report of Lemierre's syndrome.

Keywords Lemierre's syndrome · Necrobacillosis · Postanginal sepsis · Fusobacterium

Lemierre's syndrome (LS, necrobacillosis, postanginal sepsis) is a potentially fatal infection with septic vena jugularis-thrombophlebitis. LS usually occurs in otherwise healthy young subjects presenting a sudden drastic worsening with sepsis after tonsillitis [1, 4, 5]. Here, we report a young woman with LS with complications of multiple lung abscesses, and affection of liver and spleen.

K. Johannesen (✉) · U. Bødtger
Department of Lung Medicine, Næstved Hospital, Ringstedgade 61, 4700 Næstved, Denmark
e-mail: katrinejohannesen@dadlnet.dk

O. Heltberg
Department of Clinical Microbiology, Slagelse Hospital, Slagelse, Denmark

Case report

A 33-year old otherwise healthy woman was admitted to our emergency department by her general practitioner. She was suspected of having pneumonia after 5 days of sore throat, cephalgia and a fever up to 40 °C. Before admission, she was tested antigen-negative twice for hemolytic Streptococcus group A and for EBV infection.

At admission (day 1), she suffered from dizziness, tinnitus, shortness of breath and a dry cough. She presented with cold sweats, but all vital parameters were stable and normal (tp. 36, 6, O_2-saturation 99 % without oxygen, respiratory frequence 18/min, blood pressure of 105/51 mmHg), except a regular heart rate without murmurs, frequence 107 bpm. Paraclinical values: thrombocytes 37 × 10^9 (normal 150–400) and CRP 366 mg/L (<10; see Fig. 1), as well as alkaline phosphatase 188 U/L (35–105) and bilirubin 57 μmol/L (5–25), along with increased creatinine 205 μmol/L (50–90).

Cholangitis was suspected and intravenous cefuroxime, gentamycin and metronidazole were initiated after blood cultures. Abdominal ultrasonography (day 2) showed splenomegali, but otherwise without abnormal findings.

Anaerobic blood cultures showed growth of *Fusobacterium necrophorum* (day 4). Antibiotics were switched to iv penicillin and metronidazole. Transthoracal echocardiography showed no signs of endocarditis. A CT-scan of the head, neck and thorax showed a 9 cm long thrombosis in the internal jugular vein as well as multiple lung abscesses, largest of volume in the middle lobe (Fig. 2). Warfarin was commenced on day 4, but paused day 6 due to haemoptysis. Clinical and paraclinical improvement on antibiotics was observed (Fig. 1). Day 17, a new CT-scan was performed, showing remission of lung abscesses, but progression of the thrombosis. Fragmin 200 IE/kg/day was

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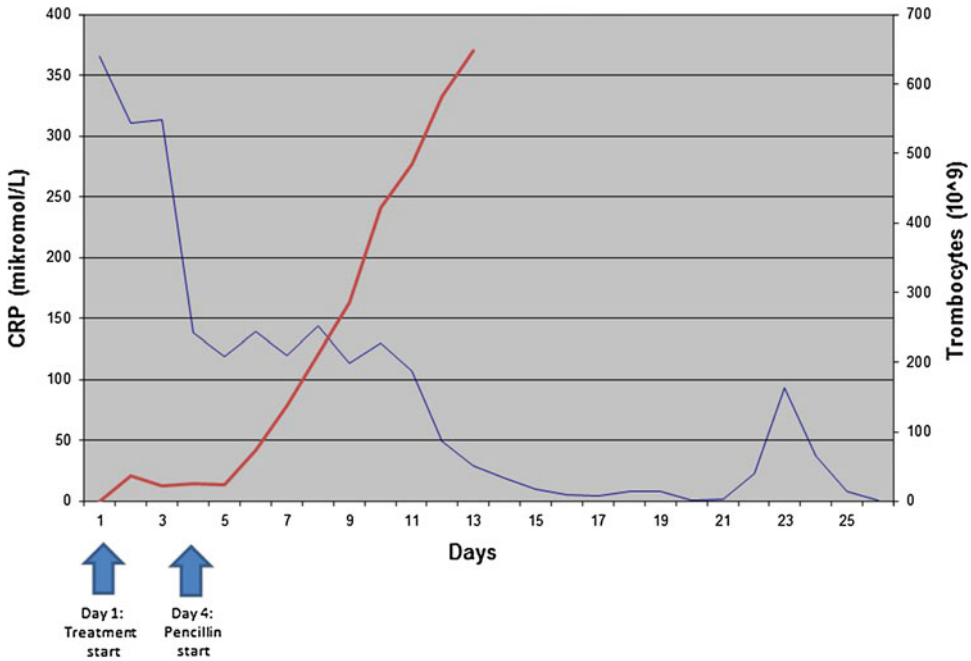

Fig. 1 Progression of CRP and thrombocytes during treatment

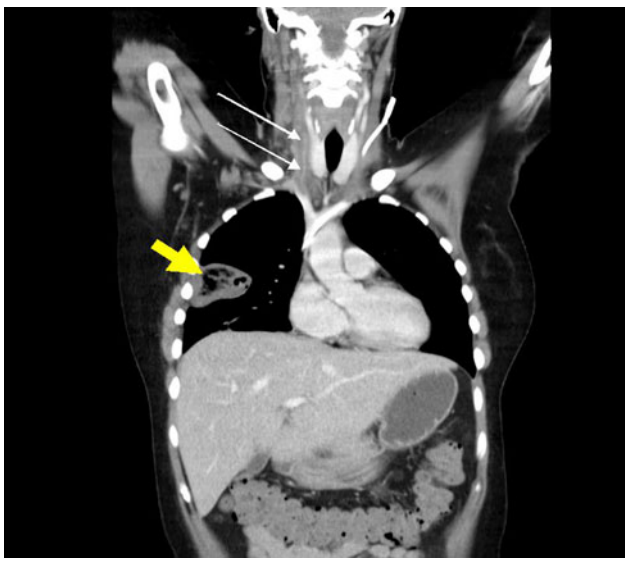

Fig. 2 Sagittal CT with contrast: *white thin arrows* thrombosis in vena jugularis interna dexter. *Fat yellow arrow* abscess in the right posterior lobe

re-inserted with transition to warfarin on day 22 without haemoptysis. On day 26, the patient was discharged with a follow-up after 3 weeks of oral phenoxymethylpenicillin 1 MIE × 4 and metronidazole 500 mg × 3, as well as 3 months of Warfarin treatment (INR-target 2–3). Ultrasound of the veins of the neck on day 46 showed complete remission of the thrombus in the internal jugular vein. She was followed up on day 90 (chest X-ray: satisfactory regression of the middle lobe infiltrate), and without LS-recurrence since then (17 months as of now).

Discussion

Lemierre's syndrome is seen after tonsillitis, pharyngitis or dental infection, facilitating a local invasion of Fusobacterium, most often *Fusobacterium necrophorum*, in the pharyngeal space or the internal jugular vein, where the infection causes septic thrombophlebitis. This occurs within 1–3 weeks. LS is often complicated by a hematogeneous spread, typically to lungs and larger joints, skin, liver, spleen, heart and CNS [5].

Karkos et al. [5] found that *F. necrophorum* was the cause of 10% of all throat infections (21 % of recurrent infections), but the frequency of LS remains largely unknown [4]. An increase has been reported since the 1990s [2, 4]. Untreated, the mortality reaches 17 %, and even with treatment mortality is still significant, especially with meningitis [2, 3]. In the Nordic countries, a combination of (initially intravenous) penicillin and metronidazole for a minimum of 3 weeks is recommended. Development of resistance is rare [2, 4, 5].

Anticoagulation remains controversial, and without any consensus [4, 5]. In this case report, progression of the thromb was seen despite 17 days of relevant antibiotic treatment.

In children, Fusobacterium infection is often seen with otitis media. In the elderly, Fusobacterium infection with a focus caudal of the head is more common, and often in

relationship with cancer [2]. Infection in these last mentioned has a higher mortality, up to 25 %.

Conclusion

Lemierre's syndrome should be considered in otherwise healthy young subjects with development of sepsis after a recent tonsillitis or pharyngitis. Anaerobe blood culture is crucial for achieving diagnosis. Antibiotics effectively reduce mortality. Addition of anticoagulation is subject to debate, and currently the need is assessed by the clinical picture and imaging.

Conflict of interest The authors have no conflict of interest, and no funds have been received.

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