A syndrome first described in 1936, still relevant today


Clinique universitaire ORL, pôle tête et cou, chirurgie réparatrice, CHU de Grenoble, BP 217, 38043 Grenoble cedex 09, France

Available online 21 December 2010

Clinical case

Mr. Claude R., born August 13, 1945, consulted for fever syndrome associated with severe dysphagia, respiratory distress, and confusion syndrome with spatiotemporal disorientation.

Surgical history comprised a cataract, right rotator cuff tear and right inguinal hernia. There was no history of alcohol abuse or smoking. He was not under medication.

In emergency unit, he had hyperthermia of 39°C associated with respiratory distress and low blood pressure. Pulmonary X-ray found bilateral white lung.

Biological assessment in the emergency room was as follows:

- neutrophil polynucleosis at 25 G/L;
- thrombopenia;
- acute functional renal insufficiency.

Clinical status worsened rapidly, showing septic shock requiring orotracheal intubation and transfer to medical intensive care.

On IC admission, cervical, cerebral and thoracic contrast-enhanced CT was performed; the findings are presented for your interpretation (Figs. 1–4).

Questions

Question n°1: What is your radiological interpretation?
Question n°2: What is your diagnosis?
Question n°3: What treatment?
Question n°4: Which bacterium is usually isolated?

Figure 1  Cervical contrast-enhanced CT.
A syndrome first described in 1936, still relevant today

Figure 2  Cervical contrast-enhanced CT.

Figure 3  Cerebral contrast-enhanced CT.

Figure 4  Thoracic contrast-enhanced CT.

Almost complete condensation of the left lung. Abundant bilateral pleural effusion.

Reply n° 2: What is your diagnosis?
Lemierre syndrome. Lemierre syndrome associates septic thrombophlebitis of the internal jugular vein or one of its collateral branches, causing remote septic embolism, which is usually pulmonary secondary to ENT (most frequently pharyngeal and bacterial) infection.

Reply n° 3: What treatment?
In emergency unit:
- intensive care for septic shock associated to symptomatic treatment of heart failure (vascular filling, catecholamines, respiratory assistance);
- under general anesthesia, drainage of left periamygdalar abscess with sampling for bacteriology;
- consult with chest surgeons: thoracotomy to evacuate pleural effusion and empty pulmonary abscesses by thoracic drainage;
- initiate iv empiric combined antibiotherapy (3rd generation cephalosporin and metronidazole) with secondary adaptation to antibiogram;
- introduce iv anticoagulants: non-fractionated heparin, due to signs suggestive of left cavernous sinus thrombosis.

Reply n° 4: Which bacterium is usually isolated?
Fusobacterium necrophorum is the most frequent.
In the present case, Streptococcus constellatus was isolated in the periamygdalar abscess pus and on blood culture.

Comments
In 1936, André Lemierre [1] reported on a series of 20 cases of septicemia implicating anaerobic bacteria, mostly associated with oropharyngeal infection extending to the neck, with disseminated (mainly pulmonary) abscesses.

Replies

Reply n° 1: What is your radiological interpretation?
Neck: Left peritonsillar abscess associated with left internal jugular vein thrombosis. Hypodense left periamygdalar image, 4 cm in diameter, related to a left periamygdalar abscess. Left internal jugular vein enhancement defect, with enhanced and thickened walls, indicative of thrombophlebitis. Infiltration of the cervical fatty tissue.

Brain: Thrombosis of left cavernous sinus. Enlarged left cavernous sinus, slightly convex externally, associated with moderate dilation of the ophthalmic veins to the left: overall aspect of partial left cavernous sinus thrombosis, despite the sinus opacity.

Chest: Multiple pulmonary abscesses. Multiple pulmonary abscesses, several excavated.
This association was later reported on as "Lemierre syndrome".

It comprises septic thrombophlebitis of the internal jugular vein or one of its collateral branches, inducing remote septic emboli.

The ENT infection is mainly oropharyngeal.

The most frequently isolated bacterium is *Fusobacterium necrophorum*.

Other bacteria reported in the literature are *Fusobacterium* species and *nucleatum*, *Bacteroides species*, *Peptostreptococcus*, *Streptococcus B and C*, *Streptococcus oralis*, *Streptococcus constellatus*, and *Eikenella corrodens* [2,3].

The advent of antibiotherapy drastically reduced incidence and associated mortality. It is now so rare as to be often unrecognized.

Clinically, Lemierre syndrome evolves through three stages:

- the initial stage of ENT (generally oropharyngeal) mucosal infection usually consists of tonsillitis with or without signs of periamygdalar phlegmon, fever and sensitive but non-inflammatory cervical adenopathy;
- in the second stage, infection spreads to the parapharyngeal spaces, with thrombophlebitis of the internal jugular vein or one of its collateral branches and septicemia. Cavernous sinus thrombosis is rare [3]. At this stage, local clinical examination finds painful inflammatory cervical induration, usually anterior to the sternocleidomastoid muscle, ipsilateral to the pharyngeal involvement. Cervical cellulitis with or without abscess formation should be systematically considered in which case suprasternal and/or supraclavicular extension of the cutaneous inflammation, with a risk of extension to the anterior mediastinum, should be explored for: there is in the neck no anatomic barrier between, on the one hand, the vascular sheath (around the internal jugular vein and internal carotid artery) and the fascias around the thoracic large vessels and, on the other, the cervical visceral space and anterior mediastinum;
- finally, signs of septicemia appear, with very high fever (>40°C) and chill, usually without associated signs of hemodynamic instability; the clinical picture is completed by septic emboli, which are usually pulmonary and articular, although other organs such as the liver, pericardium and kidneys may also be affected. The septic emboli induce remote infarcts containing microorganisms, leading to abscess formation.

Treatment is primarily medical, sometimes surgical.

Most authors [4,5] recommend combined penicillin (or 2nd or 3rd generation cephalosporins) and metronidazole (or clindamycin monotherapy), as *Fusobacterium necrophorum* is usually susceptible to these. Aminoglycosides, in contrast, provide no benefit. Two to four weeks’ parenteral antibiotherapy should be followed by oral administration. Optimal duration and dosages have not been determined.

In case of parapharyngeal, cervical or mediastinal abscess, surgical drainage should be performed, with approach adapted to location.

The role of anticoagulants is not well defined [3]. Theoretically, they entail a risk of septic embolism by fragmentation of the venous thrombus, but remain recommended in case of retrograde extension of the thrombus toward the intracranial venous drainage, especially the cavernous sinus as in the present case.

**Conflict of interest statement**

None.

**References**