Lemierre’s Syndrome – The Syndrome Quite Forgotten

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Abstract

A 17 year old male presented with seizures, headache, and fever and left chronic suppurative otitis media. A 35 year old male presented with headache, giddiness, vomiting, pain in eyes, diplopia and right chronic suppurative otitis media. Brain imaging in both revealed thrombosis of lateral and sigmoid sinus and also of internal jugular vein on the left and right side respectively. A diagnosis of Lemierre’s syndrome was made in both. They were treated with antibiotics and anticoagulants, and they responded to treatment. We want to report this case as we feel, that with the advent of the antibiotic era, this syndrome has become rare; and so “quite forgotten” or overlooked, by many physicians.

Introduction

Postanginal septicaemia (also called necrobacillosis and Lemierre’s syndrome) is caused by an acute oropharyngeal infection with secondary thrombophlebitis of the internal jugular vein (IJV), frequently complicated by metastatic infection. It was first reported by Courmont and Cade in 1900, although the syndrome was best characterised by Lemierre in 1936 from review of 20 cases.1

Case Report

Case 1

A 17 year old male presented with 3 episodes of generalised tonic clonic convulsions in a period of 24 hours. Each episode lasted for 10 minutes followed by loss of consciousness for 15 minutes. He had continuous bi-frontal headache since 12 days; fever and left earache since 3 days. There was no history of trauma, vomiting, neck pain or weakness of any limb. He had whitish discharge from left ear since 1 year but no tinnitus, cough or cold. Patient never had seizures in the past. There was no history of trauma, vomiting, neck pain or weakness of any limb. He had whitish discharge from left ear since 1 year but no tinnitus, cough or cold. Patient never had seizures in the past. There was no past history of tuberculosis. On examination, he was febrile, temp.101ºF, pulse was 96/min, regular, blood pressure 100/60mmHg, respiratory rate 24/min. Ear examination revealed purulent discharge from left ear and an unsafe chronic suppurative otitis media. There was mild tenderness over left mastoid region. Throat and nose were normal.

On neurological examination, patient was conscious, cooperative, well oriented in time, place and person. There was no neck stiffness. There was no weakness of any limb; the deep tendon reflexes were normal but plantars were bilaterally extensor. Fundus was normal. On auscultation of the chest, coarse crepitations were heard in right infrascapular and mammary region. Rest of the systemic examination was normal.

Investigations revealed Hb – 15.5gm/dl, TLC- 10,400/cmm, PPE, L,E,M,B,J, Platelet count- 2.5 lakh/cmm, haematocrit 44.2%, ESR - 15mm at end of 1 hour. Other biochemical investigations were within normal limits. HIV was negative by ELISA, VDRL – non reactive. Plain radiograph of the chest showed few non-homogeneous shadows in right lower lobe. X-ray of left mastoid showed sclerosis of the mastoid with no evidence of sinus dural plate erosion. Pus culture from ear discharge yielded Pseudomonas aeruginosa sensitive to Gatifloxacin, Cefazidime and Tazobactum. Blood culture did not detect any organism – aerobic or anaerobic. Audiometry showed mixed sensory loss in left ear.

MRI Brain and MR venography was suggestive of left lateral, sigmoid sinus and left IJV thrombosis (Figures 1 and 2) with an acute left occipital infarct. There was no evidence of haemorrhage or hemorrhagic infarct. Left otitis media was noted. Colour doppler of the neck also revealed left IJV thrombus.

Patient had been on oral Cefalexin from a private practitioner since two days prior to the admission. It was omitted and intravenous cefazidime and metronidazole (500 mg 8 hrly) were added. Low molecular weight heparin (Enoxaparin 1mg/kg/wt)

![Fig. 1: T1W axial sequence of the brain (case 1) showing abnormal hyper intense signal within the left transverse sinus suggestive of sinus thrombosis. Normal flow void is seen in the right transverse sinus.](image-url)
s.c. bd and warfarin (5mg)/day with PT and INR monitoring was started. Patient was treated with Inj. Lorazepam and phenytoin and later kept on oral maintenance dose of phenytoin (300mg/day). He had no seizures after admission. Fever and headache subsided by day 3. By day 7, on auscultation, chest was clear and chest radiograph showed partial clearing of lung infiltrates. After 2 weeks repeat MRI/MRV and colour doppler showed resolution of thromboses. Antibiotics were continued for 3 weeks. X ray chest was clear at 4 weeks. Warfarin was given for 6 weeks and omitted and the patient was then subjected to left mastoidectomy.

Case 2

A 35 year old male presented with giddiness, bifrontal headache, severe vomiting since 4 days, with diplopia and blurring of vision. There was no history of trauma, altered sensorium, seizures, fever or weakness of any limb. He was a chronic tobacco chewer but did not smoke or take alcohol. He complained of decreased hearing and minimal discharge in the right ear but no pain or tinnitus. On examination, he was afebrile, pulse was 72/min, blood pressure 120/70; fundus examination was normal. Neurological and rest of the systemic examination was normal. Investigations revealed Hb – 13.5gm/dl, TLC- 11,600/cmm, Platelet count-2.3 lakh/cmm, haematocrit 42%, ESR – 12 mm at end of 1 hour. Other biochemical investigations were within normal limits. HIV was negative by ELISA, VDRL was non reactive. Ear examination revealed purulent discharge from right ear, central perforation and chronic suppurative otitis media. There was no tenderness over the mastoid region. Throat and nose were normal. Chest radiograph was normal. X-ray of right mastoid showed sclerosis of the mastoid with no evidence of sinus dural plate erosion. MRI Brain showed a subacute venous sinus thrombosis of superior sagittal sinus, right transverse, right sigmoid sinuses and the right proximal internal jugular vein (Figure 3). There was no infarct. MR Venography showed absent flow in superior sagittal, right transverse, right sigmoid sinuses and the right proximal internal jugular vein (Figure 3). There was no infarct. MR Venography showed absent flow in superior sagittal, right transverse, right sigmoid and right proximal internal jugular vein. Left sided venogram was normal (Figure 4). Colour doppler of the neck also revealed right IJV thrombus. Pus culture from ear discharge yielded Staphylococcus aureus. Audiometry showed mixed sensory loss in right ear.

He was treated with injection Ampicillin-Cloxacillin and Metronidazole. Later Vancomycin was added as per sensitivity;
anaerobic bacterium of some kind. 6

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throat associated cases. 6 Our patients, otogenic cases, had both,
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of internal jugular vein thrombophlebitis, but a much higher
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those arising from jaw infections, ear infections, from the gut,
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however, regarding the definition of Lemierre’s Syndrome,
different authors have, over the years, used different criteria
there has been a lack of consensus. In fact, Lemierre himself,
in 1936, included cases of post angina septicaemia, as well as
those arising from jaw infections, ear infections, from the gut,
genitourinary tract. 4 Hughes in 1996, and Shetty et al in 1998
described cases following mastoiditis. 2 Riordan, in his prior
article in 2004, have also stated that authors have included in
Lemierre’s syndrome cases in which the source of infection arises
not from the throat but from the ears, mastoid, or tooth infection. 4
Our definition of the syndrome is also with the latter authors, i.e.
history of angina illness or infection from ears/mastoid, jaw, etc.

It has been found that the otogenic cases have a low incidence of
internal jugular vein thrombophlebitis, but a much higher
incidence of intracranial complications than those secondary to
throat associated cases. 6 Our patients, otogenic cases, had both, IJV thrombosis and intracranial complications. Occasionally
external jugular vein instead of IJV thrombophlebitis has been
reported. 3

There are a wide variety of organisms that have been
cultured from patient’s blood with Lemierre’s syndrome. Most
common are Fusobacterium necrophorum (hence the condition is
also called as necrobacillosis). It is a long non-spore forming,
non-motile, gram negative, anaerobic rod and is part of normal
flora of the oropharynx, gastrointestinal tract and genitourinary
tract in females. 5-3 Both our cases have isolated organisms
different from F. necrophorum. Although, it is the anaerobe
most commonly implicated in Lemierre’s syndrome, other
fusobacteria, Bacteroides species, anaerobic streptococci species
have been described. 2 Streptococci, Peptostreptococci, Eikenella,
Staphylococcus aureus, Ps. aeruginosa, Candida etc have also been
cultured. 3 In a series of cases reviewed by Sinave et al, only 23
out of 37 cases had Fusobacterium necrophorum detected and the remaining 13 patients had a range of other organisms, both
aerobic and anaerobic. 4 In the extensive review of 222 cases by
Riordan, however, 68% grew F. necrophorum, and 90% grew an
anaerobic bacterium of some kind. 6

Mainstay of treatment is a prolonged course of high-dose
antibiotics with antibiotic coverage. Antibiotics such as
clindamycin, metronidazole, penicillin and chloramphenicol
are recommended. They should be given intravenously for
1-2 weeks and then prescribed orally for an additional 2-4
weeks. Prolonged antibiotic use is necessary for eradication of
endovascular infection. 7 Because of the frequent occurrence of
mixed infection, monotherapy is inadvisable and metronidazole
with another antibiotic is recommended. 4 The second element
of management is the identification of the collection of pus
amenable to drainage. A more drastic form of therapy that has
been employed is ligation or resection of the internal jugular
vein, indicated in patients with persistent septic embolization
despite antibiotics. 4

The role of anticoagulant therapy in Lemierre’s syndrome is
controversial as the outcome in most patients is good without it.
One study suggested that addition of heparin was associated with
a quicker resolution of septic pelvic thrombophlebitis and this observation might support the use of anticoagulant in
Lemierre’s syndrome. However, there are no controlled studies
that assess its value in septic thrombophlebitis of IJV. Some
authors recommend anticoagulation when septic emboli persist
despite antibiotic therapy and others reserve it for patients with
clinical evidence of IJV thrombosis propagating retrogradely to the cavernous sinus. 4

There are some special features in our cases. Our patients had
some acute symptoms like headache, vomiting, seizures; and
chronic suppurrative otitis media, but not acute and fulminant
infection as in the case reports described earlier; the seizures and
headache were sudden though. Local findings were minimal. Also, as mentioned above, unlike other otogenic cases
of Lemierre’s Syndrome, our patients had both IJV thrombosis
and intracranial complications. The first case had infiltrates in
the right lung, probably septic emboli. But blood culture did not
grow any organism probably because of prior use of antibiotics.
Pus culture grew Pseudomonas and Staph. aureus respectively in
the 2 cases. They responded to antibiotics and other appropriate
symptomatic therapy.

In the pre antibiotic era, Lemierre’s syndrome was not
uncommon and it had a fulminant, usually fatal evolution in 7
- 15 days. Since introduction of antibiotics and their widespread
use for treatment of throat infection, there has been substantially
decrease in incidence of postanginal septicaemia. Because of
this, the syndrome is frequently overlooked and actually “quite
forgotten” when it appears today. With appropriate therapy, a
cure is to be expected in overwhelming majority of patients, so it
is essential for clinician to be aware of this not so rare syndrome,
have a high index of suspicion for early diagnosis, timely and
appropriate therapy.

References