Lemierre's syndrome caused by *Streptococcus pyogenes* in an elderly woman

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ABSTRACT

Lemierre's syndrome is characterized by septic thrombophlebitis of the internal jugular vein. It typically presents in healthy adolescents or young adults, usually preceded by an oropharyngeal infection, with the most common offending pathogen being *Fusobacterium necrophorum*. We present a case of Lemierre's syndrome in an elderly woman without antecedent oropharyngeal infection, caused by *Streptococcus pyogenes*. She was successfully treated with combined surgical and medical management. (J Vasc Surg Cases and Innovative Techniques 2020;6:31-3.)

Keywords: Lemierre's syndrome; Streptococcus pyogenes; Septic thrombophlebitis; Internal jugular vein

Lemierre's syndrome (LS) is a rapidly progressive septic thrombophlebitis of the internal jugular vein (IJV) preceded by an oropharyngeal infection and diagnosed in young healthy adolescents or young adults. The most common bacterium associated with LS is *Fusobacterium necrophorum*. The time interval is short between initial infection and onset of septicemia.[1] Early diagnosis with rapid initiation of antibiotics can be curative, and delayed diagnosis and treatment is potentially fatal. We report a unique presentation of LS in an elderly woman without a recent oropharyngeal infection, caused by *Streptococcus pyogenes*. The patient gave consent to the use of her clinical history and images for this case report.

CASE REPORT

An 80-year-old woman presented with altered mental status and right neck pain for 1 week. She denied chills, dysphagia, odynophagia, shortness of breath, cough, and history of infections or recent dental work. She was febrile to 103°F at presentation. Physical examination revealed erythema, induration, and tenderness over the right neck. Her white blood cell count was 19.6 K/μL (reference range, 4.8-10.8 K/μL), and C-reactive protein level was elevated at 35 mg/dL (reference range, 0.02-1.20 mg/dL). A computed tomography (CT) scan with intravenous contrast and duplex ultrasound examination demonstrated a thrombosed right IJV with adjacent abscess extending into the sternocleidomastoid muscle and extensive surrounding inflammation (Figs 1 and 2).

The patient was immediately started on intravenous piperacillin/tazobactam and vancomycin, then intubated in the emergency department before proceeding to the operating room for planned exploration/washout with possible ligation of the IJV. A longitudinal incision was made along the anterior border of the sternocleidomastoid muscle. The abscess cavity was superficial but the posterior aspect infiltrated the wall of the IJV. Therefore about 5.4 cm of IJV was resected with its thrombus, and the two ends ligated. There was a clear separation between the IJV and the carotid artery. A Jackson Pratt drain was placed and the wound was closed in multiple layers. Her blood cultures grew *S pyogenes* (group A beta hemolytic streptococcus) and tissue culture grew methicillin-resistant *S epidermidis*. Postoperatively, she was anticoagulated with heparin. A CT angiogram was negative for septic emboli to the lungs, and an echocardiogram was negative for endocarditis. She was discharged on hospital day 7 with an additional 10 days of ceftriaxone, metronidazole, and doxycycline as recommended by the infectious disease consultants.

The patient followed up at 2 weeks and 3 months after discharge and was doing well. She had no further signs or symptoms of infection. There was no facial edema or cranial nerve deficits. She was advised to continue the rivaroxaban for a total of 6 months because of the partial dissection of the IJV resulting in a remnant IJV stump with residual thrombus.

DISCUSSION

LS is rare, with a reported incidence of 1 per 1 million.[1] Although an increasing number of cases have been reported since the syndrome was first described and defined in 1936 by Andre Lemierre, there is an absence of evidence to determine change in disease incidence over time.[2] Mortality, however, has been significantly decreased from as high as 90% in the preantibiotic era.[3]

Our patient represents a unique case of an already rare disease for several reasons. First, this patient presented at the age of 80. The typical presentation for LS is

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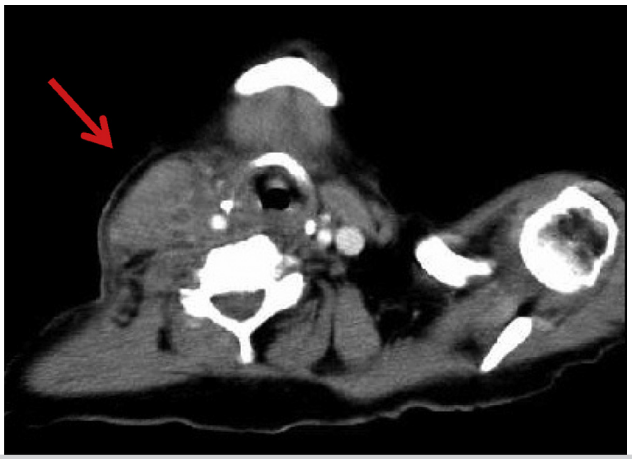

Fig 1. Computed tomography (CT) scan demonstrating right internal jugular vein (IJV) thrombosis and adjacent abscess (*red arrow*).

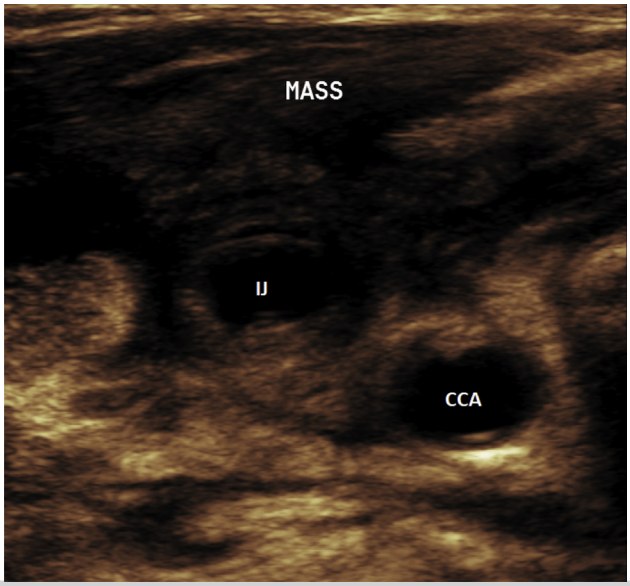

Fig 2. Duplex ultrasound scan of the right neck showing overlying abscess (*MASS*) in relation to the internal jugular (*IJ*) vein and common carotid artery (*CCA*).

adolescents and young adults between the ages of 10 and 35.[4] In our literature search, we came across only one other geriatric case, an 80-year-old man who did not survive his infection owing to a delay in his diagnoses.[5]

Second, this patient had no evidence of a sore throat or recent dental work. The oropharyngeal infection commonly associated with LS is pharyngitis with infectious spread to peritonsillar tissue or the palentine tonsils. Otogenic infections, including otitis media and mastoiditis, odontogenic infections, and sinusitis, are common presenting symptoms.[6] The only presenting symptom seen in our patient conducive to the final diagnosis was unilateral neck pain.

Last, the offending bacteria for our patient with LS was *S pyogenes*. Isolated *S pyogenes* causing LS is rare and has been described in a limited number of cases.[7] Since 2005, there have been five reports of LS caused by *S pyogenes*: two adults, one adolescent, a 4-year-old, and a 22-month-old (Table).[5,7-10] The common etiology noted among all of these patients was a sore throat with odynophagia. The most common causative pathogen is invasive *Fusobacterium necrophorum,* a gram-negative obligate anaerobe, found in up to 81.7% of LS cases.[8] Other organisms associated with LS include group A streptococci, *Bacteroides melaninogenicus*, *Eikenella corrodens*, *Leptotrichia buccalis*, and *Klebsiella pneumoniae*.[10,11]

The initial management is medical, with resuscitation and early initiation of antibiotics. The workup of these patients should include laboratory tests, blood cultures, and a CT scan of the neck. Although surgical intervention has a limited role, it is important to quickly assess the overall clinical condition of the patient and determine whether surgical intervention is required. Indications for surgical intervention include failure of medical therapy, sepsis, or drainage of an abscess. Pencle et al[12] performed and extensive review of surgical management of LS. They concluded that a multidisciplinary team approach is ideal, and surgical management is highly recommended if there is no resolution with antibiotics alone. Surgical options may include ligation and resection of the IJV, drainage of a neck abscess, or drainage of abscesses from septic emboli. Our patient required operative management secondary to a neck abscess. During the exploration, the wall of the IJV seemed to be compromised; therefore, we elected to ligate and resect this portion of the jugular vein as well.

One area of controversy in the treatment of LS is use of anticoagulation. There are no absolute guidelines, but its use has been increasingly reported in LS.[13,14] A possible indication for anticoagulation in LS is to prevent the propagation of thrombus.[15] Another indication proposed is to decrease the risk of metastatic embolisms to other organs, specifically the lungs. A high percentage of LS cases reported have shown to have septic pulmonary lesions that can lead to respiratory failure.[16] Anticoagulation has been suggested to start within 48 to 72 hours of diagnosis if there is clinical deterioration despite adequate antibiotic therapy.[17] The duration of anticoagulation treatment for maximal benefit has also not been determined.

The antibiotic choice should be tailored to the individual and the causative organism. One common regimen is ceftriaxone with metronidazole, but some monotherapies may be adequate also. There is no consensus for the duration of antibiotic therapy.[18] It has been suggested a 6-week course is necessary to adequately penetrate

Table. Lemierre's syndrome (LS) cases caused by *Streptococcus pyogenes*

Age, years/sex	Year	Presentation	Complications	Blood cultures	Imaging study showing IJV thrombosis	Management	Outcome
4/Female	2009	Pharyngitis, sepsis	None	GAS	Neck CT	Antibiotics and anticoagulation	Alive
50/Female	1995	Pharyngitis, odynophagia, sepsis	ARF, pneumonia, pleural effusion	GAS	Neck CT, Doppler U/S	Antibiotics alone	Alive
80/Male	2007	Pharyngitis, odynophagia, sepsis	Shock	GAS	Neck CT	Antibiotics and anticoagulation	Died
13/Male	2007	Pharyngitis, sepsis	Thrombocytopenia, ARF	GAS	Doppler U/S	Antibiotics and anticoagulation	Alive
64/Female	2016	Pharyngitis, odynophagia, dyspnea, sepsis	ARF, PE	GAS	Neck CT	Antibiotics and anticoagulation	Alive
22 months /Female	2011	Fever, lethargy, worsening hemodynamic and mental status	SIRS, pneumonia, osteomyelitis	GAS	Neck CT, Doppler U/S	Antibiotics and anticoagulation	Alive

ARF, Acute respiratory failure; *CT*, computed tomography; *GAS*, group A streptococci; *IJV*, internal jugular vein; *PE*, pulmonary embolism; *SIRS*, systemic inflammatory response syndrome; *U/S*, ultrasound examination.

into the fibrin clot.[4] A shorter 2.5-week course was used for our patient given her remarkable recovery after surgical source control.

CONCLUSIONS

We encountered a patient with LS who presented at an advanced age and absent a typical antecedent oropharyngeal infection. LS should not be dismissed in elderly patients presenting with neck pain. Primary therapy is with antibiotics, but surgical intervention still has an important role in advanced disease.

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