Sigmoid sinus thrombosis secondary to Lemierre’s syndrome

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Abstract

Lemierre’s syndrome, a rare and almost forgotten cause of internal jugular vein thrombosis, is usually caused by an anaerobic head and neck infection. Left untreated, it can result in the release of septic emboli. We describe the case of a 42-year-old man who presented with fever and a tender, swollen neck mass. Computed tomography revealed an edematous parapharyngeal area and a compressed internal jugular vein. Despite antibiotic treatment, the patient’s condition worsened, and a parapharyngeal fluid collection was drained 4 days later. Six weeks later, the patient returned to the outpatient department complaining of headaches, and he was found to have a tender, firm neck. He was readmitted, and magnetic resonance venography revealed a right internal jugular vein thrombosis that extended intracranially to the sigmoid sinus. He was anticoagulated for 6 months, and he remained well during outpatient follow-up. We examine the controversial roles that anticoagulation and thrombolysis play in Lemierre’s syndrome and sigmoid sinus thrombosis, and we review the diagnostic options.

Introduction

Lemierre’s syndrome (postanginal sepsis, necrobacillosis) is characterized by thrombosis of the internal jugular vein that is usually preceded by an infection in the oropharynx or other head and neck site. It is frequently complicated by sepsis and septic metastasis, which can affect the lungs, the musculoskeletal system, and occasionally the liver. The incidence of Lemierre’s syndrome declined significantly after the introduction of antibiotics, and it has become an increasingly rare phenomenon. Lemierre’s syndrome remains associated with high morbidity, especially when the administration of appropriate therapy is delayed. Therefore, it is still important that clinicians be aware of this classic entity so that they can recognize its presentation and initiate appropriate therapy quickly.

Sigmoid sinus thrombosis can occur as a result of the direct spread of infection from the mastoid bone or as a result of thrombophlebitis in communicating veins or sinuses. A significant association remains between sigmoid sinus thrombosis and intracranial sepsis.

We present a rare case of Lemierre’s syndrome that was caused by sigmoid sinus thrombosis, and we discuss its diagnosis and treatment.

Case report

A 42-year-old man was referred by his general practitioner to our regional ENT department for evaluation of a 2-day history of fever and a tender, swollen mass in the upper anterior triangle on the right side. The patient said he had been feeling generally run-down during the preceding week, but he reported no other symptoms on general inquiry. He was otherwise fit and well, and he was taking no regular medication. He had undergone an elective tonsillectomy at the age of 10 years for recurrent tonsillitis.

Apart from the neck mass, findings on further examination, including endoscopy of the upper aerodigestive system, were normal. The patient had no problem maintaining his airway. Blood testing revealed that inflammatory markers were elevated; his C-reactive protein level was 105 mg/L, and his white blood cell count was 12.6 × 10⁹/L.

In view of the size and position of the mass, urgent computed tomography (CT) of the neck was obtained in order to exclude a parapharyngeal abscess. CT demonstrated enlarged superficial cervical lymph nodes on the right, some edema of the right parapharyngeal space, and compression of the right internal jugular vein; there was no evidence of a collection (figure 1).

The patient was treated conservatively with cefuroxime and metronidazole, but no clinical improvement was seen. Dental examination by the maxillofacial team revealed no dental cause. After 4 days, the patient underwent an incision and drainage of the right parapharyngeal space via a right high-collar incision. A sterile phlegmon was drained, and a surgical drain was left in situ. The fluid that had drained during the operation yielded no microbial organisms on
culture. The patient made an unremarkable recovery and was discharged 4 days postoperatively.

Six weeks later, the patient was examined in the outpatient clinic, and his neck was again found to be tender and firm. He was readmitted for urgent magnetic resonance imaging and magnetic resonance venography. Venography detected a right internal jugular vein thrombosis that extended intracranially to the right sigmoid sinus (figure 2). During this hospitalization, the patient complained of headaches, but he was neurologically normal.

We discussed this case with members of the hematology, neurology, and neurosurgical teams. On the basis of these consultations, we decided to initiate anticoagulation, having determined that the benefit of preventing embolization with anticoagulation outweighed the small potential risk that it would lead to a cerebral hemorrhage. The patient underwent anticoagulation for 6 months. At outpatient follow-up 4 months following the discontinuation of anticoagulation, he was well. Follow-up magnetic resonance venography revealed recanalization of the sigmoid sinus, but the internal jugular vein remained occluded.

Discussion
In 1936, André Lemierre described the presentation, treatment, and outcomes in 20 cases of the syndrome that came to bear his name. During the preantibiotic era, Lemierre's syndrome was not uncommon. It was rapidly progressive and usually fatal by 1 to 2 weeks of symptom onset, as occurred in 18 of the 20 cases reported by Lemierre.

Lemierre's syndrome is most commonly associated with Fusobacterium necrophorum—the so-called "spindle of death." This anaerobic, nonmotile, gram-negative rod is found in the normal flora of the oropharynx, gastrointestinal tract, and female genital tract, and it is usually sensitive to clindamycin and metronidazole. F. necrophorum possesses a lipopolysaccharide endotoxin that has been shown to be lethal in animals. In humans, this endotoxin can cause platelet aggregation. Other organisms that have been isolated in patients with Lemierre's syndrome include Fusobacterium nucleatum, Streptococcus ssp, Bacteroides ssp, Eikenella corrodens, and Gemella morbillorum.

Lemierre reported that the clinical manifestation of this entity "constitutes a syndrome so characteristic that mistake is almost impossible." When this rare entity does occur, it is most common in otherwise healthy adolescents and young adults; some cases have been reported in young children. Patients usually have a history of a recent oropharyngeal infection (often pharyngitis), but the syndrome has also followed mastoiditis, otitis, dental infections, and parapharyngeal abscesses. At the onset of symptoms, tonsillitis is often apparent, but the signs of the initial oropharyngeal infection may have already resolved by the time internal jugular vein thrombosis and metastatic infections have become evident.

The development of internal jugular vein thrombophlebitis and associated septicemia usually takes between 1 and 3 weeks. As the parapharyngeal space infection progresses and thrombophlebitis of the internal jugular vein develops, the patient experiences a marked, spiking pyrexia. Many patients will also manifest a palpable tender swelling at the angle of the mandible and along the sternocleidomastoid muscle, and the thrombosed internal jugular vein will be palpable. Infections that primarily involve the posterior compartment of the parapharyngeal space (posterior to the stylohyoid process) may manifest few local signs.

Thrombophlebitis of the internal jugular vein can lead to the formation of septic emboli and therefore to the manifestation of a variety of clinical signs and symptoms. Patients may have havelung involvement (manifesting as breathlessness, chest pain, or hemoptysis), septic arthritis (with joint pain and swelling), or hepatic or splenic abscesses (possibly with jaundice).

With regard to sigmoid sinus thrombosis, mortality rates of 90 to 100% were regularly reported during the preantibiotic era. Since then, rates have ranged from 10 to 40%. As many as 10% of survivors may have residual neurologic deficits.
The clinical features of sigmoid sinus thrombosis vary from subtle and often-vague signs and symptoms to overt toxemia and septic embolization. Symptoms include headache, otalgia, otorrhea, vertigo, vomiting, and rigors. Proptosis, retroorbital pain, papilledema, and ophthalmoplegia suggest further extension of the thrombus.

The clinical features of internal jugular vein thrombosis are subtle. A detailed history, with reference to the possible etiology, may help in making the diagnosis. The most common cause today is trauma, usually as a result of injection (by intravenous drug users) or cannulation of the vein. Infection remains a rare cause. Malignancy and thrombophilia may need to be ruled out. In the presence of high fever and septic embolism, internal jugular vein thrombosis can be mistaken for endocarditis. The predominant features of internal jugular vein thrombosis are pain and swelling below the angle of the jaw, although occasionally a thickened cord of internal jugular vein may be palpable deep to the sternocleidomastoid muscle. If this entity is not observed and treated early, the diagnosis becomes all too apparent upon the release of the septic emboli.

When Lemierre's syndrome is suspected on the basis of extensive thrombosis, retroorbital pain, papilledema, and diminished visual acuity, otalgia, otorrhea, vertigo, vomiting, and rigors. Proptosis, retroorbital pain, papilledema, and ophthalmoplegia suggest further extension of the thrombus.

The treatment of internal jugular vein ligation is controversial, and little but retrospective evidence exists. The efficacy of anticoagulation, hydration, and correction of any underlying cause has been supported and refuted. According to a review of the Cochrane Database, only 2 small trials have suggested that anticoagulation drugs are probably safe and may be beneficial for patients with sigmoid sinus thrombosis; however, the results of these trials were not conclusive.

Aggressive therapeutic measures may be required for patients who do not respond adequately to conservative treatment. These patients may continue to exhibit signs and symptoms of increased intracranial pressure, such as headache, vomiting, papilledema, and diminished vision. Regional infusion of thrombolytic therapy has been suggested as the best way to treat these patients. Most reports involved patients with extensive thrombi that extended into the sigmoid sinus; treatment resulted in few complications. However, a review of the Cochrane
SIGMOID SINUS THROMBOSIS SECONDARY TO LEMIERRE'S SYNDROME

Database failed to identify the indications for or the safety of thrombolytic treatment.  

Although Lemierre's syndrome is rare, clinicians should be aware of it because of its potentially devastating consequences. We hope that this article will increase awareness of this important but often forgotten condition.

References