Lemierre Syndrome: Rediscovering the "Forgotten Disease"

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Case Presentation

A 77-year-old male with diffuse large B-cell lymphoma, on zanubrutinib, presented with a chief complaint of a one-time fever of 102°F which resolved with over-the-counter Tylenol. He reported four days of right-sided neck edema and odynophagia. He had a right-sided chest port which had not been accessed in six months. He denied any history of VTE, anticoagulant use, or IV drug use. The patient was afebrile on presentation. Physical exam revealed right-sided jugular edema. Labs showed elevated procalcitonin (0.16 ng/mL) with no leukocytosis or neutropenia. A respiratory viral panel was negative. Venous Doppler ultrasound of the right upper extremity revealed an occlusive thrombus of the right internal jugular vein (IJV). Peripheral blood cultures were obtained, and the patient was discharged with Eliquis 10 mg PO BID and outpatient follow-up.

That evening, blood cultures grew positive for Enterococcus faecalis. The patient was contacted and returned the following morning. He was admitted with a diagnosis of Lemierre syndrome and started on empiric IV ampicillin (2 g Q4H) and a heparin drip. Repeat blood cultures from the chest port and a peripheral line confirmed positive for E. faecalis. A throat culture was negative. The chest port, deemed the likely source of infection, was removed on hospital day four, with catheter tip cultures confirming E. faecalis. Transthoracic echocardiogram on day five showed no valvular lesions or vegetations. He was discharged on hospital day seven with ampicillin home infusions for four weeks and Eliquis 10 mg PO BID.

Discussion

Lemierre syndrome is a rare condition characterized by thrombophlebitis of the IJV, bacteremia, and septic emboli. It is typically associated with oropharyngeal infections in young, healthy individuals. Fusobacterium necrophorum is the most implicated pathogen and invades the IJV, causing thrombophlebitis and the formation of septic emboli. Fever, exudative tonsillitis, dysphagia, and unilateral neck pain are clinical indicators of Lemierre syndrome. Diagnosis requires radiographic evidence of IJV thrombosis and positive throat or blood cultures. Contrast-enhanced CT of the neck and chest is the preferred imaging modality. Labs may show elevated inflammatory markers and thrombocytopenia. After diagnosis, prompt initiation of antibiotics is necessary, as delayed treatment is associated with high mortality.

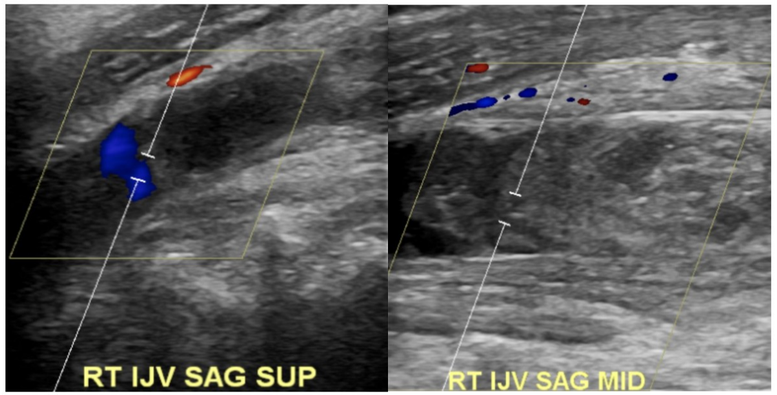

Figure 1: Venous Doppler ultrasound imaging of the right internal jugular vein demonstrating an occlusive thrombus.

Conclusion

This case illustrates a unique presentation of Lemierre syndrome in an immunosuppressed elderly patient with a chest port as the source of infection. Lemierre syndrome remains underrecognized, lacks standardized diagnostic criteria, and is often considered a "forgotten disease" among physicians. No systematic study of Lemierre syndrome has been done to date. Further education about the pathogenesis, diverse clinical presentation, and evolving management strategies for Lemierre syndrome is necessary.

References

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