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Autoimmune disease with libman sacks endocarditis presenting with acute limb ischemia

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Abstract

Background: Libman Sacks Endocarditis is rare, nonbacterial vegetation of the heart valves. Most commonly seen in association with solid cancers and autoimmune diseases.

Case: A 64-year-old male came in with sudden onset left upper arm numbness, pain and pallor which worsened with movement. On examination, he had feeble pulses of the left upper extremity and an aortic diastolic murmur of the heart. He underwent an angiogram which revealed decreased flow distal to the brachial artery and therefore was scheduled for an emergent embolectomy. An echocardiogram showed a mass on the aortic valve. Repetitive blood cultures were negative. Autoimmune workup showed positive lupus anticoagulant and cardiolipin IgM, possibly suggesting antiphospholipid syndrome. Unfortunately, we were unable to follow up on the thrombophilia workup at 12 weeks.

Conclusion: Autoimmune disease has a strong association with Libman Sacks endocarditis, treatment with warfarin is preferred and valve replacement is necessary in some cases.

Keywords: Autoimmune disease; Libman sacks endocarditis; Nonbacterial endocarditis; Limb ischemia.

Abbreviations: LSE: Libman Sacks Endocarditis; APS: Antiphospholipid syndrome.

Introduction

Libman Sacks Endocarditis (LSE) or marantic endocarditis or nonbacterial thrombotic endocarditis is a disease that is characterized by sterile, verrucous vegetations on cardiac valves. It may be associated with malignancy (especially in the solid tumors of the pancreas, stomach, or lung), Systemic Lupus Erythematosus (SLE), or Antiphospholipid Antibody Syndrome (APS) [1]. It most commonly involves healthy mitral valves, aortic valves, and less commonly other valves [2]. LSE is a rare disease, and diagnosis is challenging as most patients are asymptomatic and either found incidentally during cardiac workup or postmortem with a prevalence of 0.9% to 1.6% [1]. The most common clinical manifestation is due to thromboembolic events, like stroke, transient ischemic attack, and limb ischemia. Among all causes of limb ischemia, upper extremity involvement is <5% [3]. We present a case where both these unique conditions – LSE and upper limb ischemia- meet in a patient with possible Antiphospholipid Syndrome (APS).

Case report

A 64-year-old Caucasian male with a history of hyperlipidemia and hypertension arrived at the emergency department

with sudden onset of left upper arm numbness after a shower. It was associated with pallor and left elbow pain which worsened with movement. He observed his extremity gradually becoming cold and clammy and brought himself to the emergency department. He denied any trauma, family or personal history of blood clots, or prolonged immobilization. On physical examination, the left upper extremity was cold to the touch, and pale with feeble distal pulses. Heart auscultation revealed a grade 3, aortic diastolic murmur. Laboratory tests were only remarkable for an elevated white blood cell count of 16.8 K/UL, neutrophilic. The sedimentation rate was normal at 13 mm/hr (normal: 0-20 mm/hr). An upper extremity angiogram was done which showed decreased flow distal to the brachial artery at the antecubital fossa with faint runoff. He was immediately started on a heparin drip and vascular surgery was consulted. He underwent an emergent embolectomy and the thrombus was sent for pathological examination.

To further assess the etiology of the thrombus and due to the patient's aortic murmur, a transthoracic echocardiogram was completed which showed a suspected mass on the noncoronary leaflet of the aortic valve. For better understanding, a transesophageal echocardiogram was performed and it also redemonstrated an aortic valve mass on the noncoronary leaflet of 0.6 x 0.8 mm. Three consecutive blood culture sets were negative. Embolectomy clot pathology resulted in showing a fibrinous thrombus. Nonbacterial thrombotic endocarditis was considered and a thrombophilia workup was ordered which revealed a positive lupus anticoagulant (a PTT 109 sec (normal < 43 sec), a PTT 1:1 mix 73 sec (normal <43 sec), hexagonal phase phospholipid neutralization assay positive) and elevated cardiolipin IgM at 14 MPL (normal </= 12.5 MPL). ANA negative. Unfortunately, we were unable to follow up on the thrombophilia workup 12 weeks after the event to confirm the suspected diagnosis of APS.

Discussion

LSE was first described by Emanuel Libman and Benjamin Sacks in 1924 [4]. It is a rare diagnosis since most patients are asymptomatic and mainly present as a thromboembolic phenomenon, like in our case. The pathophysiology of LSE is not clearly understood. It appears to be a reaction to endothelial injury from circulating cytokines or interleukins, which results in vegetations comprised of immune complexes, fibrin, and platelet [1]. The vegetation size may vary from 2 mm to 10 mm, with an irregular border, and can be valvular or subvalvular with independent motion and are sterile [1,5]. When left untreated vegetation may become a fibrous plaque with focal calcifications [6]. Patients suspected to have LSE should be evaluated with at least three sets of blood cultures, transthoracic echocardiography (sensitivity of 60-75%) and transesophageal echocardiography (sensitivity >95%) [1,7].

About 32%-38% of patients with primary APS have valvular defects on echocardiography [6]. It is also interesting to note that patients with positive APS in SLE (secondary APS) have a higher prevalence of valvular defects [6]. At a molecular level, APS is an autoimmune disease characterized by antibodies against phospholipid-binding proteins [8]. It presents as recurrent miscarriages and arterial/venous thrombosis. It can be primary or secondary (SLE, drug-induced). Diagnosis is based on the presence of lupus anticoagulant, IgG or IgM anticardiolipin antibodies, or anti-beta-2 glycoprotein antibodies. Repeat testing is done 12 weeks after the initial diagnosis to rule out any transient antibodies. APS and LSE can contribute to a greater likelihood of embolic events due to their risk of thromboembolism [6].

Treatment of LSE in APS mainly consists of antithrombotic agents. The preferred agent in patients with APS and proven thrombus is warfarin with a target INR of 2-3 [1]. A study by Khamashta et al in patients with APS-associated valvular disease resulting in thromboembolism showed oral anticoagulant with INR >/= 3 was more effective than INR <3, although associated with higher bleeding risk. Most patients require life-long anticoagulation. Treatment of the underlying etiology of SLE and APS also may prevent recurrent thromboembolism. There are concerns that steroids may worsen valvular dysfunction by facilitating the healing of vegetation. Surgical valve replacement is considered, if necessary, in patients with recurrent embolisms or severe valvular dysfunction. The prognosis of LSE is considered to be poor and depends on the etiology.

Conclusion

LSE is rare and usually diagnosed after patients present with thromboembolic events. It may be secondary to autoimmune diseases like APS, SLE, and rarely rheumatoid arthritis. The mainstay of treatment is via antithrombotic therapy, but some patients with severe valvular dysfunction or recurrent thromboembolism might require surgical repair of the valve.

Declarations

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References

1. Ibrahim AM, Siddique MS. Libman Sacks Endocarditis. [Updated 2022 May 15]. In: Stat Pearls [Internet]. Treasure Island (FL): Stat Pearls Publishing; 2022. Available from: https://www.ncbi.nlm.nih.gov/books/NBK532864/

2. Behbahani S, Shahram F. Electrocardiogram and heart rate variability assessment in patients with common autoimmune diseases: A methodological review. Turk Kardiyol Dern Ars. 2020; 48: 312-327.

3. Mc Carthy WJ, Flinn WR, Yao JS, Williams LR, Bergan JJ, et al. Result of bypass grafting for upper limb ischemia. J Vasc Surg. 1986; 3: 741–746.

4. I Moyssakis, MG Tektonidou, VA Vasilliou, M Samarkos, V Votteas, et al. H. M. Moutsopoulos, Libman-Sacks endocarditis in systemic lupus erythematosus: Prevalence, associations, and evolution, The American Journal of Medicine. 2007; 120: 636–642.

5. Sotiris C Plastiras, Constantinos A Pamboucas, Maria Tektonidou, Savvas T, et al. Toumanidis, Real-time three-dimensional echocardiography in evaluating Libman– Sacks vegetations, European Journal of Echocardiography. 2010; 11: 184–185. https://doi.org/10.1093/ejechocard/jep172

6. Hojnik M, George J, Ziporen L, Shoenfeld Y. Heart valve involvement (Libman Sacks endocarditis) in the antiphospholipid syndrome. Circulation. 1996; 93: 1579-1587.

7. Yuan XC, Liu M, Hu J, Zeng X, Zhou AY, Chen L, et al. Diagnosis of infective endocarditis using echocardiography. Medicine (Baltimore). 2019; 98: e17141.