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Ischemic Stroke Revealing Libman-Sacks Endocarditis: A Case Report

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Abstract

Libman-Sacks endocarditis (LSE) is a characteristic but rare feature of systemic lupus erythematosus (SLE), which can be found in association with antiphospholipid syndrome (APS), exposing the patient to an increased embolism risk, particularly the occurrence of ischemic stroke.

We present a case involving a 64-year-old man who was admitted for ischemic stroke accompanied by a fever. Cardiac investigation revealed mitral vegetations along with severe mitral regurgitation. The diagnosis of Libman-Sacks endocarditis associated with SLE and APS was made after a laboratory work-up showing negative blood cultures and positive antibodies. This case underlines the importance of early diagnosis for better management of this pathology.

Keywords: Libman-Sacks endocarditis, Systemic lupus erythematosus, Endocarditis, Mitral regurgitation

1. Introduction

Libman-Sacks endocarditis (LSE) is defined by the presence of sterile vegetations on the heart valves. It is a rare cardiac manifestation of systemic lupus erythematosus, and the association of antiphospholipid syndrome is even rarer [1]. Manifestations may go unrecognized and progress to severe cardiac and systemic complications (cerebral and peripheral embolisms, valve dysfunction, superinfection), accentuating the risk of mortality [2].

2. Case report

We present a case involving a 64-year-old man with a history of previous ischemic stroke, primary biliary cirrhosis, chronic renal failure of unknown etiology, who was admitted with an ischemic stroke extended to parietal territory. Clinically, the patient had a fever of 38.6° Celsius, dysphasia, and a mitral systolic murmur at 4/6 on cardiac auscultation.

Transthoracic echocardiography (TTE) revealed severe mitral regurgitation (Fig. 1) and moderate dysfunction of the left ventricular ejection fraction, followed by transoesophageal echocardiography which showed mitral vegetations measuring 7 mm × 9 millimeter in diameter adherent to the anterior leaflet and 8 × 14 mm on the posterior leaflet (Fig. 2). On admission, laboratory results revealed normocytic normochromic anemia at 9 g/dl, an inflammatory syndrome with positive C-reactive protein at 66 mg/l (reference range: 0–8 mg/l), negative procalcitonin and negative blood cultures. The diagnosis of infective endocarditis was made, and antibiotic therapy was initiated with penicillin and gentamycin, as well as treatment for heart failure: angiotensin inhibitors, diuretics and beta-blockers. However, due to the increase in the size of vegetations and persistent inflammatory syndrome after 15 days, vancomycin was added.

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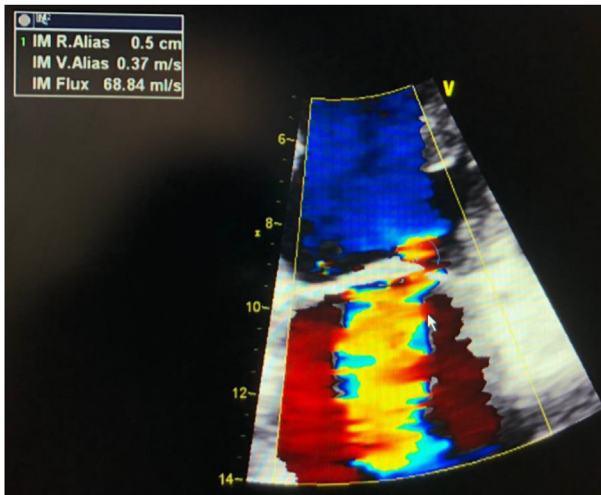


Fig. 1. Transthoracic echocardiography showing severe mitral regurgitation.

The patient's condition deteriorated with worsening renal function, anemia, thrombocytopenia and multiple ischemic micro-strokes. An immunological origin was thus suspected, supported by a biological work-up revealing positive antinuclear antibody and anti-double stranded DNA with complement consumption, as well as antiphospholipid antibody. This confirmed the diagnosis of Libman-Sacks endocarditis associated with SLE and APS.

The patient received a 3-day bolus of methylprednisolone, followed by oral corticosteroids. Immunosuppressive therapy with Cyclophosphamide was administered, leading to a significant improvement of pancytopenia. Anticoagulation with

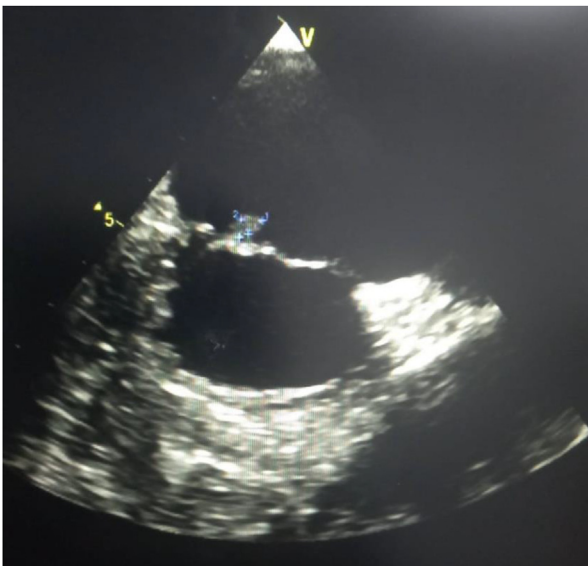


Fig. 2. Transoesophageal echocardiography showing mitral vegetation.

Abbreviation list

LSE	Libman-Sacks endocarditis
SLE	Systemic Lupus Erythematosus
APS	Antiphospholipid antibody syndrome
TTE	Transthoracic echocardiography

low-molecular-weight heparin, followed by anti-vitamin K, was gradually introduced to avoid hemorrhagic infarction. The patient's condition improved with the resolution of the inflammatory profile and the disappearance of fever. However, due to the persistence of vegetations, severe mitral regurgitation and embolic recurrence, mitral valve replacement was successfully performed using a mechanical valve, with favorable clinical outcome. Bacteriology of the valve was negative, and histopathological examination showed degenerative changes and chronic inflammatory cell infiltrate with fibroid necrosis, further supporting this diagnosis (Fig. 3). Six months after this surgery, the patient's condition is actually stable and there is an improvement in his dysphasia.

3. Discussion

Libman-Sacks endocarditis is a cardiac manifestation of Lupus, first described in 1924 in 4 patients with sterile aortic vegetations [3]. Its prevalence ranges from 0.9 % to 16 %, without gender preference, in a population aged between 40 and 80 years [4]. However, this may be underestimated: post-mortem studies have found a prevalence of 30 % in a population of SLE sufferers [5]. Endocarditis may also be associated with rarer autoimmune diseases



Fig. 3. Resected mitral valve of our patient.

such as antiphospholipid syndrome, an arterial or venous thrombotic disease accompanied by high levels of antiphospholipid antibodies.

The pathophysiology is poorly understood: the classic lesion is a verrucous vegetation that develops from an immunologically-induced endothelial lesion leading to the formation of fibrin, platelets and immune complexes on the valve, causing fibrosis and valve dysfunction [4]. Diagnosis is made on TTE, which shows vegetations with specific characteristics: they are mobile, elongated, narrow-based, with irregular borders and well attached to the valve surface [2].

The location is often mitral, aortic and more rarely tricuspid, and the severity of lesions is associated with the duration of lupus, its activity and the presence of antiphospholipid syndrome, which often carries a poor prognosis [5]. Transoesophageal echocardiography is more sensitive (over 74 %), allowing better characterization of localization, extension and, most importantly, the elimination of differential diagnoses (thrombus, tumors) [2]. Biological tests can also provide useful clues, such as low C-reactive protein levels, low white blood cell counts due to SLE activity, and negative blood cultures.

The complications of this pathology are serious, including cerebral embolism, as seen in our patient. In these patients, cerebral perfusion is reduced, neurocognitive disorders increase and the risk of ischemic stroke is elevated. Eleven percent of Libman-Sacks patients suffer from ischemic stroke [6]. Young ischemic stroke victims should be investigated for APS, and SLE patients presenting with fever or heart murmurs should not hesitate to undergo echocardiography to look for Libman-Sacks endocarditis [7,8].

While the treatment of lupus is well defined, that of Libman-Sacks endocarditis is unclear, and relies mainly on anti-inflammatory therapy [9]. Corticosteroids have anti-inflammatory and antithrombotic aims, but may accelerate fibrosis and hence valve dysfunction, making them controversial. Immunosuppressants and hydroxychloroquine aim to reduce SLE activity. However, there is no consensus on the efficacy of these therapies on valve damage [8,9]. Treatment of heart failure, if present, should also be initiated. When LSE is associated with APS, the addition of an anticoagulant is necessary. Surgery should be considered in cases of uncontrolled infection, severe valve dysfunction or embolic recurrence [7]. Our patient presented with a recurrence of ischemic stroke and persistent severe mitral insufficiency, which led to the indication for valve replacement. According to some studies, valve plasty may also be considered when SLE activity is low with isolated lesions [10].

4. Conclusion

Libman-Sacks endocarditis is a disease with serious cardiac and extra-cardiac manifestations, emphasizing the importance of early recognition to adopt appropriate therapeutic measures [3]. Confirmation of the diagnosis is challenging, relying on a range of clinical, biological, echocardiographic arguments, and on the patient's immunological profile. Nowadays, therapeutic management remains a challenge.

Author contribution

Conception: MAY, FZM. **Literature review:** MAY, FZM, RH. **Methodology:** MAY, FZM, HR. **Software:** MAY. **Analysis and/or interpretation:** MAY. **Investigation:** MAY, HB, IB. **Resources:** MAY, HB, IB. **Data collection and/or processing:** HB. **Writer-original draft:** MAY, HR, HB. **Writing- review & editing:** MAY, HR. **Visualization:** MAY, FZM, RH. **Supervision:** IB, RH. **Project administration:** RH. **Fundings:** RH.

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Conflict of interest

None declared.

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