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Isolated Tricuspid Valve Libman-sacks Endocarditis in Patient with Systemic Lupus Erythematosus: A Rare Case Report and Literatures Review

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Libman-Sacks endocarditis (LSE), characterized by the formation of verrucous vegetations, is a typical cardiac manifestation of autoimmune diseases such as systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APS). It primarily leads to cardiac valve lesions. The most commonly affected valves in systemic lupus erythematosus are the mitral and aortic valves. Although isolated tricuspid valve involvement is quite rare. Here, we report the case of a 38-year old female with a history of SLE who suffered from acute right heart failure caused by tricuspid vegetation and valve regurgitation. The patient was successfully treated with prednisolone and hydroxychloroquine, and follow-up echocardiography showed the disappearance of the vegetations.

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Keywords: Tricuspid valve; Libman-Sacks endocarditis; systemic lupus erythematosus.

1. INTRODUCTION

Libman and Sacks first published a description of these atypical, sterile, verrucous vegetations in 1924 [1]. Libman-Sacks endocarditis most commonly involves the mitral and aortic valves. However, all four cardiac valves and the endocardial surfaces can be involved [2]. Nowadays, LSE has been recognized as a typical cardiac manifestation of autoimmune diseases such as SLE and antiphospholipid syndrome (APS). The pathogenesis of Libman-Sacks endocarditis would involve the formation of a thrombus on a valve damaged by the deposits of immune complexes, inducing inflammation, which progresses to fibrosis with distortion and dysfunction [3]. LSE often involves the left heart valves, and tricuspid lesions are very rare. Most cases of LSE can be treated with medical therapy, while very few require surgical treatment [4]. Here, we report the case of a 38-year-old female with a history of SLE who suffered from acute right heart failure caused by tricuspid vegetations and valve regurgitation. She underwent treatment with prednisolone and hydroxychloroquine with a favorable outcome.

2. CASE PRESENTATION

38-year-old Moroccan female presented to our department in June 2023, complaining of continuous weakness and shortness of breath for more than 4 months. The patient had a significant past medical history of systemic lupus erythematosus (SLE) for over 13 years. She experienced paroxysmal knee joint pain, paroxysmal nocturnal dyspnea, and orthopnea. Additionally, she reported increased dyspnea on exertion during daily activities and increasing lower extremity edema (Fig. 1). Upon admission, the patient had a mild fever with a peak temperature of 38.5 °C. Heart auscultation revealed a systolic murmur at the 4th intercostal space by the left border of the sternum. Transthoracic and transesophageal echocardiography (TTE and TEE) revealed moderate tricuspid regurgitation and a large single vegetation on the atrial surface of the anterior leaflet (Fig. 2), which was displaced by blood flow. Laboratory tests showed normal complete blood counts, an elevated erythrocyte sedimentation rate, normal C-reactive protein levels, positive antinuclear antibody, decreased complement C3 and C4 levels, and negative anti-double-stranded DNA antibody, anticardiolipin antibody, and lupus anticoagulant. Hepatic and renal functions were normal upon admission. Blood cultures were obtained three times consecutively, but no bacterial growth was observed. The patient was treated with prednisone and hydroxychloroquine, and follow-up echocardiography showed the disappearance of the vegetations.

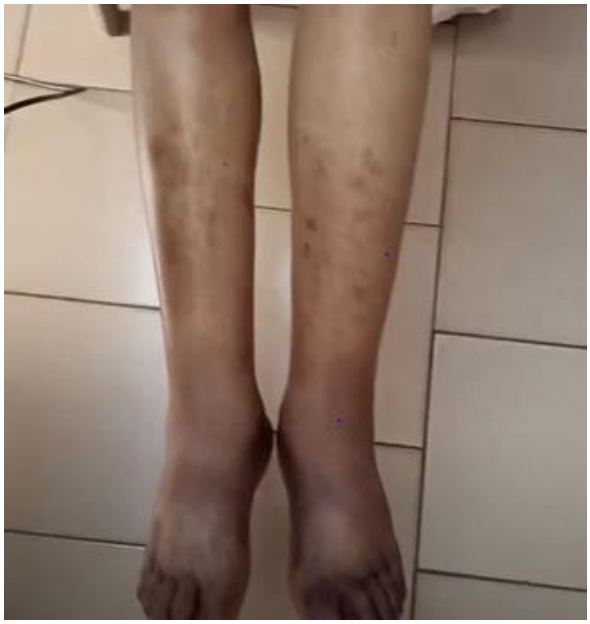

Fig. 1. Lower extremity edema

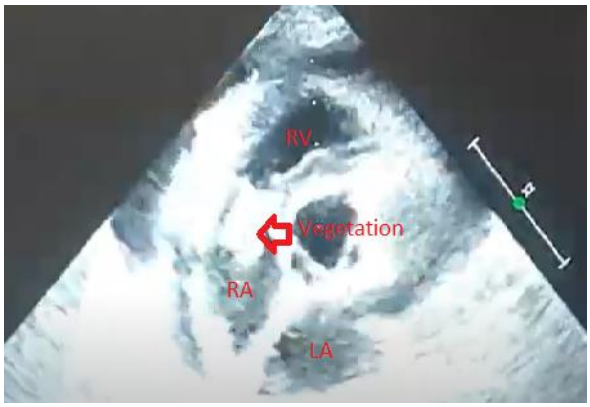

Fig. 2. TTE; 4 chambers view. Vegetation (red arrow) attached to the atrial surface of the anterior tricuspid valve leaflet

3. DISCUSSION

Libman-Sacks endocarditis (LSE) is recognized as a typical cardiac manifestation of autoimmune diseases such as SLE and antiphospholipid

syndrome (APS). While mitral and aortic valves are commonly affected, tricuspid valve involvement is rarely reported. Moyssakis reported 38 cases of LSE in 342 SLE patients diagnosed by echocardiography, including 24 with mitral and 13 with aortic involvement, and only one case with tricuspid involvement [5]. Doppler echocardiography is considered the diagnostic method of choice, although distinguishing between LSE and infectious endocarditis (IE) can sometimes be challenging, as both conditions may present with fever. Echocardiographically, LSE vegetations appear as masses of varying size and shape with irregular borders and heterogeneous echo-density, firmly attached to the leaflet surface and exhibiting no independent motion, while IE vegetations typically exhibit independent motion" [6,7]. The role of transesophageal echocardiography (TEE), especially RT3D-TEE, has been emphasized in assessing vegetation size in LSE patients [8]. Treatment for LSE involves drug therapy and surgical intervention. Corticosteroids and anticoagulation drugs are used for medical treatment. While corticosteroids may not prevent LSE, they can help heal LSE lesions by reducing inflammation [9,10,11]. However, they can also increase tissue fibrosis and scarring, potentially worsening valvular damage and dysfunction. Nevertheless, appropriate and sufficient steroid therapy to control autoimmune disease activity is important. Anticoagulation therapy is required due to the increased risk of thromboembolic events in LSE, and current therapeutic guidelines for APS recommend long-term anticoagulation to prevent thromboembolic events [7,9]. Therefore, if the patient with LSE is hemodynamically stable, conservative treatments as described above should be recommended. Moaref reported a case of successfully recovered LSE treated with prednisolone and hydroxychloroquine; however, the patiens symptoms of heart failure did not worsen and received drug treatments for months [12]. In cases of severe intractable symptomatic valvular dysfunction, surgical intervention for LSE may be necessary [13,14]. In the case of LSE, the implantation of a bioprosthesis is not generally recommended due to reported cases of re-operation in the future, often caused by rapid calcification, valvulitis, and subsequent perforation, or massive bioprosthetic thrombosis [15,16]. Therefore, a mechanical prosthesis might provide better overall results for LSE, but there is no expert consensus on prosthesis selection for tricuspid site involvement. Typically, mechanical tricuspid valve replacement (TVR) is associated with increased early mortality [17] and a higher occurrence of valve-related events, particularly a combination of thrombosis, embolism, and bleeding, [18] when compared to the possibility of mid-to-long-term degeneration and failure of bioprosthetic valves. Thus, determining which type of prosthesis is more favorable for tricuspid site involvement in LSE remains uncertain.

4. CONCLUSION

LSE should be strongly suspected when significant valve vegetation is observed in the course of SLE and/or APS. While mitral involvement is common, tricuspid LSE is rarely reported. TEE, particularly RT3D-TEE, is useful for differentiating between LSE and IE. Initial treatment recommendations should include conservative measures with steroids, but patients with untreated and severe intractable symptomatic valvular dysfunction may require surgical intervention.

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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