Isolated tricuspid valve Libman-Sacks endocarditis in patient with systemic lupus erythematosus: A rare case report and literatures review

Abstract

Libman-Sacks endocarditis (LSE), characterized by verrucous vegetations formation, is a typical cardiac manifestation of autoimmune diseases such as systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APS). It primarily leads to lesions of cardiac valves . The most common valves involved in systemic lupus erythematosus are the mitral and aortic valves. Although isolated tricuspid valve involvement is quite rare. Here we reported a 38-years-old female with past SLE history 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.

Keywords: tricuspid valve, Libman-Sacks endocarditis , systemic lupus erythematosus

Background

Libman and Sacks first published a description of these atypical, sterile, verrucous vegetations in 1924(1). Libman–Sacks endocarditis most commonly involves mitral and aortic valves. However, all 4 cardiac valves and the endocardial surfaces can be involved(2).Nowadays, LSE have been seen 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 involve left heart valves, tricuspid lesions were very rare. And most LSE can be treated with medicine therapy while very few need surgical treatment. Here we reported a 38-years-old female with past SLE history suffered from acute right heart failure caused by tricuspid vegetations and valve regurgitation which underwent treatment with prednisolone and hydroxychloroquine with a good evolution.

Case presentation

A 38-year-old young Moroccan female presented to our department for continuous weakness and short of breath for more than 4 months on June 2023. The girl's past history was significant for systemic lupus erythematosus (SLE) more than 13 years. Symptoms of paroxysmal knee joint ache, paroxysmal nocturnal dyspnea and orthopnea are manifested. She had experienced increased dyspnea on exertion with activities of daily living and increasing lower extremity edema.

After admission the patient had transitional mild fever with the highest temperature of 38.5 °C. Heart auscultation showed systolic murmur at the 4th intercostal space by the left border of sternum. The transthoracic and transesophageal echocardiography (TTE and TEE) showed

moderate tricuspid regurgitation and a large single vegetation on the atrial surface of anterior leaflet, which was swinged by blood flow. Her laboratory test yielded the following: normal regular blood tests, elevated erythrocyte sedimentation rate and normal C-reactive protein level, positive antinuclear antibody , decreased complement C3 and C4 levels, and negative anti-double-stranded DNA antibody, negative anticardiolipin antibody and negative lupus anticoagulant. Hepatic and renal functions were all normal just after admission. Blood culture was taken for 3 times consecutively, but no existence of bacteria was shown. Prednisone and hydroxychloroquine were given after admission and follow-up echocardiography showed the disappearance of the vegetations.

Discussion

LSE have been seen as a typical cardiac manifestation of autoimmune diseases such as SLE and antiphospholipid syndrome(APS). Mitral and aortic valve is the mostly involved valve, while tricuspid valve involvement was rarely reported. Moyssakis reported 38 LSE in 342 SLE patient which were diagnosed by echocardiography, among which there were 24 mitral and 13 aortic involvement. only one tricuspid involvement (4). Doppler echocardiography can be considered as the diagnostic technique of choice. But sometimes it is very difficult to identify LSE and true infectious endocarditis (IE), for the former may also have fever due to the original immunology diseases and the latter may also have vegetation. Echocardiographically, LSE vegetations appear as masses of varying size and shape with irregular borders and heterogeneous echo-density, which are firmly attached to the leaflet surface and exhibit no independent motion (5). While the vegetation of IE may typically exhibit independent motion (6). The role of transesophageal echocardiography (TEE), had been emphasized in assessing vegetation size in a patient with LSE (7). Treatment for LSE consists of drug treatment and surgical intervention. Corticosteroid and anticoagulation drugs are used for LSE drug treatment. Corticosteroids cannot prevent LSE, but they can help healing LSE lesions by lessening inflammation (8, 9, 10). However they can also increase tissue fibrosis and scarring, finally worsening valvular damage and dysfunction. Nonetheless, appropriate and sufficient steroid therapy to control autoimmune disease activity is important. Anticoagulation therapy is required due to the increased risk of thrombo-embolic events in LSE and current therapeutic guidelines for APS did suggest long-term anticoagulation to prevent thrombo-embolic events [6, 8]. So if the patient with LSE is hemodynamically stable, conservative treatments above should be firstly recommended. Moaref had reported a successfully recovered case of LSE with treatment of prednisolone and hydroxychloroquine, but the patient denied worsening symptoms of heart failure and received drug treatments by months (11). If with severe intractable symptomatic valvular dysfunction, surgical intervention for LSE may be required (12,13).

Usually for LSE, it was not very recommended to implant a bioprosthesis since there was reported cases that underwent re-operation in the future, due to rapid calcification, valvulitis and subsequent perforation (14) or massive bioprosthetic thrombosis (15). So a mechanical prosthesis might provide a comprehensive better result for LSE. But there were no expert suggestion on prosthesis selection for tricuspid site for LSE. For normal consideration, mechanical tricuspid valve replacement (TVR) leads to increased early mortality (16) and occurrence of valve-related events, especially the composite of thrombosis, embolism, and bleeding (17). Compared to the

possibility of mid-to-long-term degeneration and failure of bioprosthesis, we could not tell which kind of prosthesis on tricuspid site should have better outcomes.

Conclusion

LSE should be strongly suspected when significant valve vegetation unveiled during the course of SLE and/or APS. Mitral involvement is common but tricuspid LSE is rarely reported. TEE, especially RT3D-TEE, is useful for diagnosis between LSE and IE. Conservative treatment with steroids should be firstly recommended, but patients with untreated and severe intractable symptomatic valvular dysfunction still need surgical intervention.

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