CASE REPORTS

CEREBRAL EMBOLISM: A CHALLENGING COMPLICATION IN LIBMAN-SACKS ENDOCARDITIS

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Abstract

Libman-Sacks endocarditis is an uncommon disorder that can be a risk factor for neurocognitive dysfunction, and brain lesions, either separately or in a combination. We report 57-year-old woman from Colombia who presented with a history of dyspnea, associated with general malaise and arthralgia of small joints. On examination, she had signs of chronic arthropathy and purpuric plantar lesions. Her cardiac exam revealed a pan-systolic murmur of mitral regurgitation, harsh, grade 5/6, best heard at the apex, and a diastolic murmur in the aortic focus. Her echocardiography revealed severe mitral regurgitation and moderate aortic regurgitation with nodular thickening of both valves, consistent with a diagnosis of LSE. Blood cultures and serologic tests were negative. After starting anticoagulant treatment, the patient suffered a stroke, and it was finally decided that she would undergo surgery. She suffered a second stroke with status epilepticus in the immediate postoperative period. She eventually recovered without serious neurological sequelae.

Keywords: *Libman-Sacks endocarditis; marantic endocarditis; stroke; thrombosis.*

INTRODUCTION

Libman-Sacks endocarditis (LSE), also known as marantic or verrucous endocarditis, is a form of nonbacterial thrombotic endocarditis (NBTE) which involves the presence of sterile vegetations on the cardiac valves. LSE was first described in four patients in 1924 by Emanuel Libman and Benjamin Sacks in New York. Vegetations in NBTE commonly involve a single native, usually healthy, mitral and/or aortic valve, and are often small, more friable, and easily prone to dislodge, leading to systemic embolization to the brain, visceral organs, including the lungs, spleen, and kidneys, or extremities. The initial development of Libman-Sacks endocarditis appears to be an endothelial injury in the setting of a hypercoagulable state. Thus, they are mainly observed in patients with malignancies, systemic lupus erythematosus (SLE), which was first described in women in 1985, and antiphospholipid antibody syndrome[1] (APS).

CLINICAL CASE

A 57-year-old woman from Colombia that was previously diagnosed with hypertension, monoclonal gammopathy and erosive arthropathy, arrived in Spain, where she suffered several episodes of arthritis that required infiltrations and immunosuppressive agents. The patient complained of dyspnea, general malaise and arthralgias and was admitted from the rheumatology service due to a new episode of arthritis with cervical involvement and the presence of purpuric plantar lesions on both feet.

Blood tests showed positive rheumatoid factor, ANAS, anti-DNA and anti-Ro; negative anti-La and anti-Sm. Hypocomplementemia C3 and C4, and positive anticardiolipin IgG antibodies. Blood cultures and HACEK serology were negative.

A chest CT angiography was performed due to the appearance of purpuric lesions on the soles of her feet,

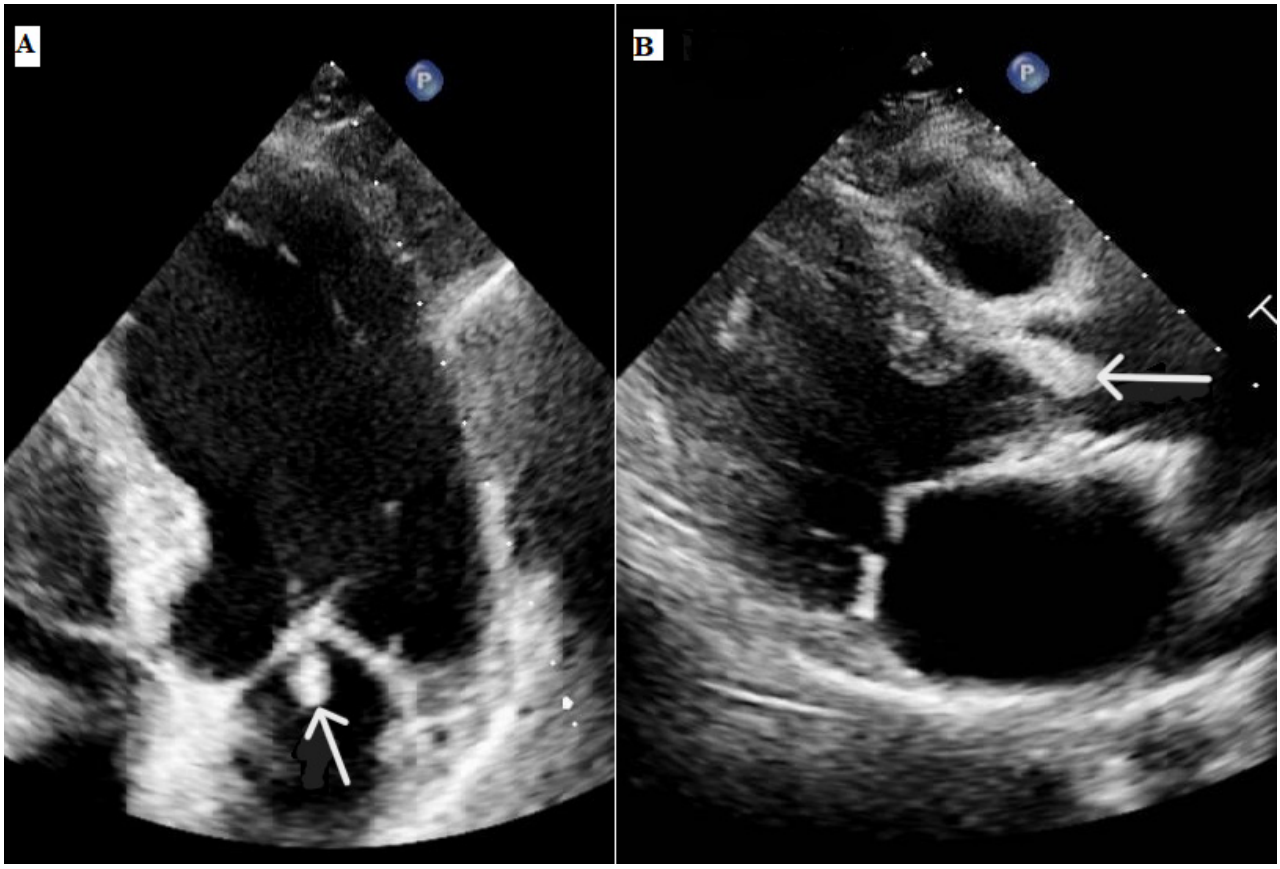

Figure 1 *Two-dimensional TEE revealed a vegetation attached to the atrial surface of the anterior mitral leaflet (A) and a vegetation attached to the aortic side of the aortic non-coronary leaflet (B). Both valves appeared thickened and inflammatory.*

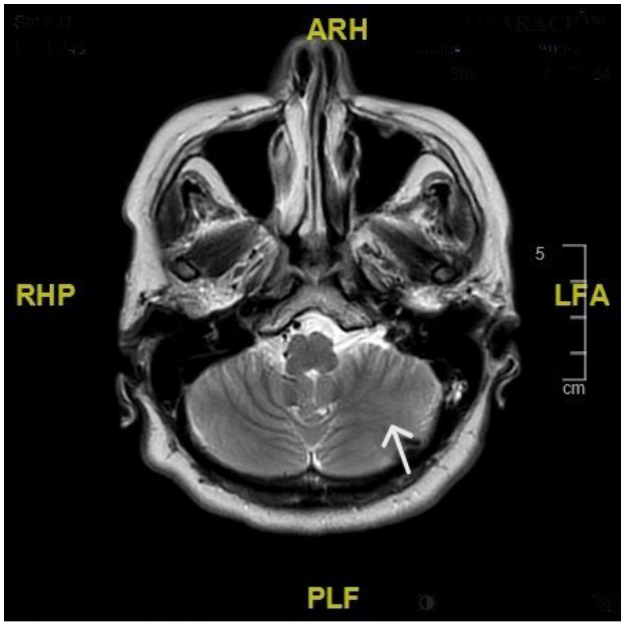

Figure 2 *Brain MRI revealed an acute left cerebellar lacunar ischemic stroke.*

with no evidence of thrombi in the cardiac cavities or in the pulmonary vascular tree. Skin biopsies were performed and showed evidence of vasculitis.

The transesophageal echocardiogram (TEE) showed a vegetation on the atrial side of the anterior mitral leaflet measuring 10x7 mm and an extensive vegetation on the aortic side of the non-coronary aortic leaflet measuring 20x10 mm, with diffuse thickening of the leaflets and severe mitral and moderate aortic insufficiency (Fig.1).

These results confirmed the diagnosis of SLE, and the possibility of LSE was considered. Anticoagulation treatment was instituted. The patient had a sudden headache, so a brain magnetic resonance imaging (MRI) scan was performed, which revealed an acute left cerebellar lacunar ischemic stroke (Fig.2) which is why surgical treatment was decided upon.

Intraoperative analysis of the valves showed a destructured mitral and aortic valve with diffuse thickening and appearance inflammatory of the leaflets (Fig.3). Conventional sternotomy and extracorporeal circulation were performed, and the patient underwent mitral and aortic valve replacement with 27 mm and 21 mm mechanical

prosthesis St. Jude Medical® Mechanical Heart Valves (SJM; Minneapolis, Minn), respectively. Microscopic examination and cultures of operative specimens revealed sterile vegetations. Histopathology and immunofluorescence studies did not reveal relevant data.

On the first postoperative day, she experienced a seizure with status epilepticus, aphasia, and left-sided gaze deviation. Brain CT angiography with perfusion revealed an acute infarction in the left frontal territory, which was successfully treated with mechanical thrombectomy (Fig.4).

The patient recovered without serious neurological sequelae and was discharged from the hospital two weeks after surgery. Follow-up at 1, 6 and 12 months showed no evidence of systemic or neurological complications, and the mitral and aortic valve prostheses were functioning normally.

DISCUSION

Libman-Sacks endocarditis is a rare disease that is mostly found postmortem with a prevalence of about 0.9% to 1.6%. LSE most commonly affects patients between 40 and 80 years of age, although it can occur in any age group. Studies do not show any sex predilection. However, SLE and antiphospholipid syndrome show a predominance in women of childbearing age, being 5 to 9 times more common than in men[2,3].

Patients are typically asymptomatic, and the condition is often an incidental finding during evaluation for other cardiac illnesses or at postmortem. The vegetations can affect both the ventricular and atrial sides of the valves. The most affected valves are those of the left side

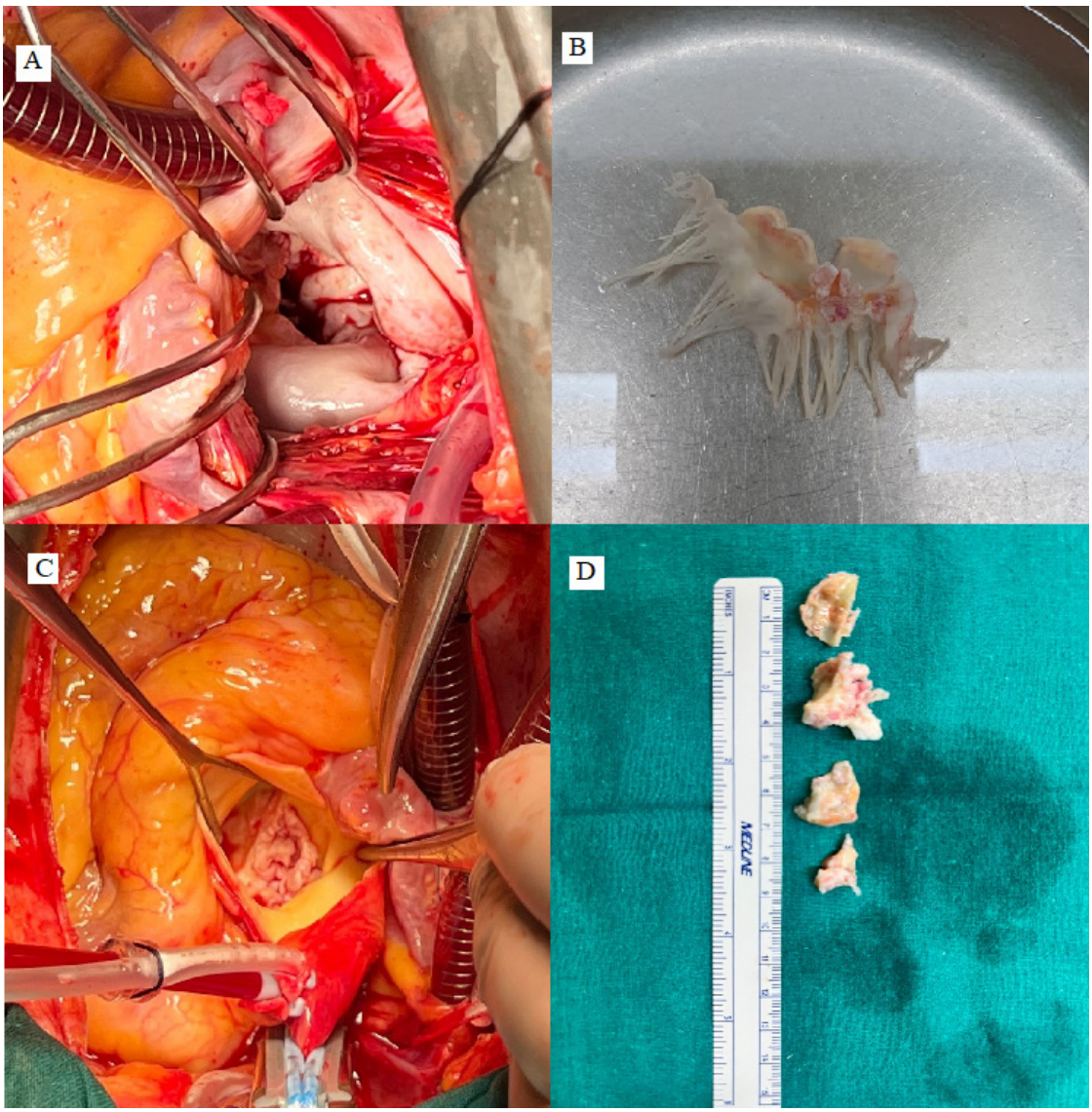

Figure 3 *Intraoperative appearance of the mitral valve (A) and anterior mitral leaflet (B). Intraoperative appearance of the aortic valve (non-coronary leaflet) (C) and aortic leaflets (D) shows a thickened and inflammatory appearance of both valves.*

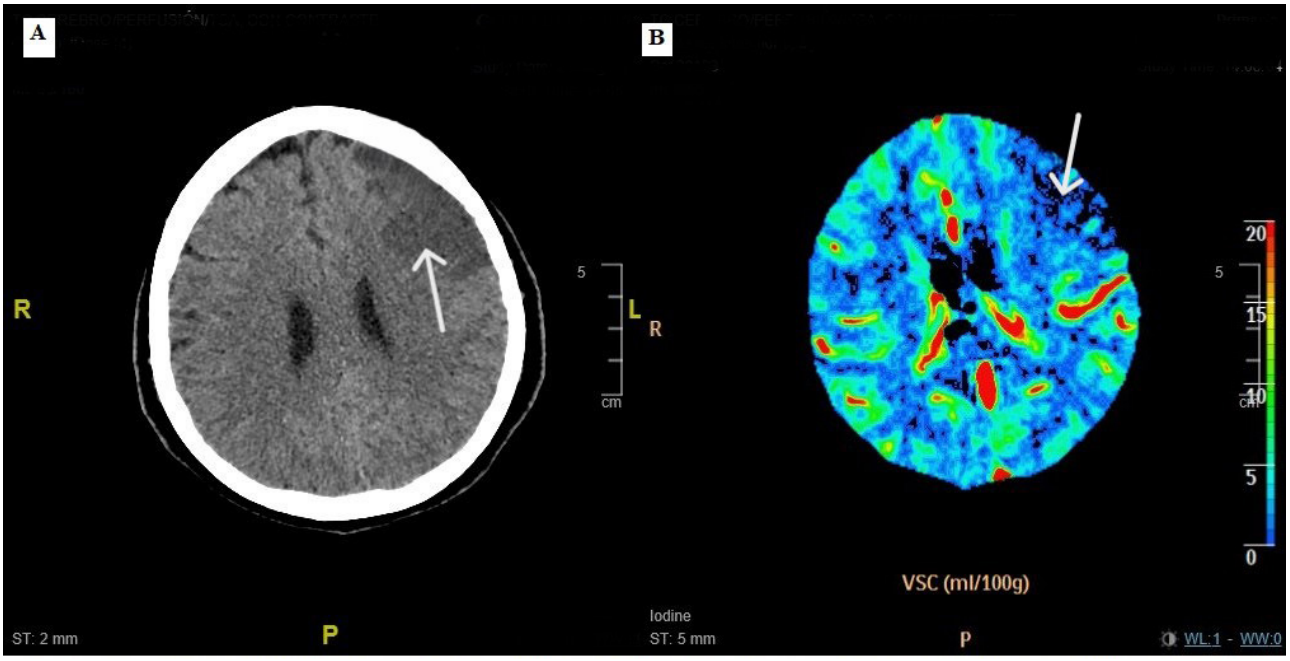

Figure 4 *Cranial CT angiography revealed an area of marked hypodensity in the left frontal territory (A), while cranial CT with perfusion showed occlusion of the second branch of the left middle cerebral artery with penumbral volume (B).*

chambers (mitral followed by aortic), but other valves can also be involved. A cerebrovascular embolism can present as a stroke or a transient ischemic attack with the presence of cerebromicroembolism, neurocognitive dysfunction, and focal brain lesions[4], as the present case with two thrombotic-cardioembolic strokes. Systemic thromboembolism can manifest as peripheral embolism, presenting as Janeway lesions, as described in patients with systemic lupus erythematosus, bacteremia without endocarditis, gonococcal sepsis, typhoid fever, hemolytic anemia and marantic endocarditis[5].

SLE patients show constitutional symptoms (fever), malar rash, discoid rash, arthritis, oral ulcers, serositis (pleuritis, pericarditis), renal disorders, neuropsychiatric symptoms, hematological issues (pancytopenia), or abnormal laboratory values (ANA, Anti-dsDNA, Anti-SM). Antiphospholipid syndrome presents with recurrent miscarriages, abortions, and venous/arterial thrombosis, with the presence of lupus anticoagulant and IgG or IgM for anti-cardiolipin antibodies[6].

In patients suspected of LSE, a full assessment should include complete blood count, complete metabolic panel, and blood cultures to differentiate it from other etiologies such as infective endocarditis. A hypercoagulable workup is also necessary in each case suspected of LSE, including lupus anticoagulant and antiphospholipid antibodies[7,8].

Transesophageal echocardiography has greater sensitivity and specificity than transthoracic echocardiography. Irregular borders, heterogeneous echo density, and the absence of independent motion characterize the masses found on the cardiac valves and endocardium. These are usually small, sessile masses, but they can be as large as 10 mm in diameter. The basal and mid-portion of the mitral and aortic valves are most commonly involved. Diffuse or focal leaflet thickening of the mitral and aortic valves may be observable. The involved valves may exhibit regurgitation[9].

The optimal treatment of LSE is not well established, but anticoagulation is recommended. The underlying disease (SLE or APS) also requires treatment. Prevention of recurrent embolization is the most common reason for surgery[10].

CONCLUSIONS

LSE is not a common condition, but it is often associated with high morbidity and mortality. The healthcare team should be aware that the vegetations can embolize and leading to ischemia and necrosis of the affected organs. Sometimes, anticoagulant treatment alone is not sufficient, and surgery may be necessary. Multidisciplinary follow-up is required, including a cardiologist, a cardiac surgeon, a rheumatologist, an infectious disease specialist and a neurologist.

Conflict of interest

The authors declare no conflicts of interest.

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