Superinfected Libman-Sacks endocarditis revealing a systemic lupus erythematosus case report with review of the literature

Abstract

We present the case of isolated mitral regurgitation detected in a 24 years old patient who was presented to the cardiology department of Ibn Rochd University Hospital complaining of dyspnea on moderate exertion. The present case report describes a 24 years-old Moroccan lady, single who were complaining of dyspnea on moderate exertion since 3 months. Parenteral probabilistic antibiotic therapy combining gentamicin (3 mg/kg/day) and ceftriaxone (2 g/d) was initiated and well followed. However, the clinical and biological course was stationary until the 7th day. This had motivated the realization of an immunological assessment which showed a high titer of anti-nuclear antibodies, positive native anti-DNA antibodies. Valvular abnormalities are often clinically silent, without significant valvular dysfunction. Valvular regurgitation is more common than stenosis, which is rare. Valvular dysfunction can result in cardiac failure. There is no consensual therapeutic protocol in the literature, hence the interest of multidisciplinary consultation meetings in which the place of the internist is essential.

Keywords: anti-DNA antibodies, cardiology, anti-nuclear antibodies, Libman-Sacks endocarditis

INTRODUCTION

Libman-Sacks endocarditis is rare as a first manifestation of systemic lupus erythematosus. Currently, sterile verrucous lesions of Libman-Sacks endocarditis are recognised as a cardiac manifestation of both systemic lupus erythematosus and antiphospholipid syndrome. They are clinically silent in a majority of the cases. The presence of antiphospholipid antibodies in systemic lupus erythematosus is associated with three times higher prevalence of mitral valve nodules and significant mitral regurgitation. We present the case of isolated mitral regurgitation detected in a 24 years old patient who was presented to the cardiology department of Ibn Rochd University Hospital complaining of dyspnea on moderate exertion.

Case report

The present case report describes a 24 years-old Moroccan lady, single who were complaining of dyspnea on moderate exertion since 3 months. She was presented to the cardiology department of Ibn Rochd University Hospital to ask for medical advice.Clinical examination showed her BP = 100/70 and HR 90 bpm. Peripheral pulse was well felt. Cardiac examination revealed accentuated S2, and S4 on the apex. Normal chest/abdominal examination. No skin or oropharyngeal lesions, as well as no arthritis or any joint deformities were observed. The electrocardiogram was normal. Transthoracic echocardiography was done for the patient showing a vegetation attached to the ventricular surface of the anterior mitral valve leaflet

measuring 10 * 7 mm, and causing mild mitral regurgitation. Three pairs of blood culture samples were collected from different sites and all had negative results (3 series made at the time of febrile peaks and remained negative even after 7 days of culture). The serodiagnosis of certain microorganisms (e.g. Coxiella burnetii, Bartonella spp, Chlamydia psittaci, Brucella spp) could not be carried out due to the unavailability of special media within our department. Hemoglobin = 11.8 mg/dL, platelets = 270,500 and leukocytes = 3600, creatinine = 0.6 mg/dL, C-reactive protein (CRP) = 54 mg/L normal urinary sediment. In front of this table, the diagnosis of probable infective endocarditis was then evoked according to the modified Duke criteria (a major criterion: vegetation and a minor criterion: fever).

Parenteral probabilistic antibiotic therapy combining gentamicin (3 mg/kg/day) and ceftriaxone (2 g/d) was initiated and well followed. However, the clinical and biological course was stationary until the 7th day. This had motivated the realization of an immunological assessment which showed a high titer of anti-nuclear antibodies, positive native anti-DNA antibodies. In front of this table, the diagnosis of superinfected Libman-Sacks endocarditis had been retained; thus motivating the addition by internists of corticosteroid therapy at 1 mg / kg / day to antibiotic therapy.

Discussion

Libman–Sacks endocarditis (otherwise known as verrucous, or nonbacterial endocarditis) is the characteristic cardiac manifestation of the autoimmune disease systemic lupus erythematosus (SLE). 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).

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).Anatomopathological examination of vegetations shows fibrin deposits, an infiltrate of mononuclear inflammatory cells, fibrosis, neovessels and sometimes immunoglobulin and complement deposits (4). In our case, the patient did not have surgery, so we did not have an anatomopathological result.

Valvular abnormalities are often clinically silent, without significant valvular dysfunction. Valvular regurgitation is more common than stenosis, which is rare. Valvular dysfunction can result in cardiac failure. Roldan et al. found a 43% prevalence of Libman-Sacks endocarditis in a series of 69 lupus patients by routine transesophageal ultrasound. For other authors, Libman-Sacks endocarditis accounts for less than 10% of lupus patients. This difference in prevalence is probably explained by the lack of systematic transesophageal cardiac ultrasound. After 4 years of follow-up, valve failure and/or stenosis were frequent, and two patients who were candidates for heart surgery died (5).

Thus, the diagnosis of LSE is important and the echocardiogram is currently the best imaging procedure for diagnosis.6 One must also recall that the infectious endocarditis is not unusual in

SLE patients with LSE and a differential diagnosis is mandatory. In this aspect, three laboratory data are important: leukocyte count, CRP levels, and blood cultures (6) .The leukocytes tend to decrease during lupus activity and the opposite occurs in infectious endocarditis. Very high CRP levels suggest an infectious cause, as lupus patients are less capable of presenting an exuberant response of this protein; however, for a definitive differential diagnosis, the blood cultures are more important (7). The combined rate of heart failure, valvular replacement, thromboembolism, and secondary infective endocarditis has been reported to be as high as 22% in lupus patients with valvular disease, compared with 8% of patients without valvular disease. Most patients do not have clinically significant valvular dysfunction. Regurgitation is noted on echocardiography images in 25–61% of lupus patients. The reported patients who need valve replacement varies from 1% to 8% of cases(8) .The occurrence of clinically significant embolic phenomena is thought to be low. Although stroke rates are higher in patients with lupus and antiphospholipid syndrome, multifactorial etiologies for neurological events are often present, making the specific contribution of valvular abnormalities difficult to determine.

There is no consensual therapeutic protocol in the literature, hence the interest of multidisciplinary consultation meetings in which the place of the internist is essential. Many studies show that the use of corticosteroids and immunosuppressants appears to have no effect on the regression of valve lesions (9).According to other studies, corticosteroids accelerate healing of autoimmune myocardial injury by reducing inflammation and disease activity, but may aggravate bacterial infection and are therefore not recommended if infective endocarditis is suspected (10) or bacterial superinfection.With regard to anticoagulation, it is necessary as a preventive measure when the thromboembolic risk is high. Indeed, the presence of valvular involvement in a context of Antiphospholipid Syndrome (APS), is associated with a high risk of occurrence of arterial thrombosis and cerebral embolic events. However, the occurrence of bleeding complications may limit the prescription and therefore expose patients to more thromboembolic risk.In case of valve dysfunction with significant hemodynamic impact, control can be done by conservative treatment: immunosuppressants, anticoagulants, and specific treatment of heart failure (diuretics, beta-blockers and ACE inhibitors). If the picture is severe and refractory, surgical correction is necessary. APS is the most common autoimmune cause of non-infectious endocarditis requiring valve replacement (11). . As a result, a mechanical prosthesis may provide better outcomes for mitral valvular heart disease due to Libman-Sacks endocarditis (12).Plasty is preferred whenever possible. When valve replacement is unavoidable, the choice between a bio prosthesis or a mechanical prosthesis will depend on the age of the patient, his wishes and, in the woman, the desire to procreate. It should be remembered that in case of SLE, the risk of calcification of the bioprosthesis must be taken into consideration and anticoagulation may be necessary.

Conclusions: Libman-Sacks vegetations can be found in approximately 1 of 10 patients with systemic lupus erythematosus, and they are associated with lupus duration, disease activity, anticardiolipin antibodies, and antiphospholipid syndrome manifestations. A progression of valve lesions may occur during long-term follow-up.

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