Libman–Sacks endocarditis and cerebral embolization in antiphospholipid syndrome

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In antiphospholipid syndrome (APS), there is a high prevalence of valvular heart disease which leads to increased risk of thrombo-embolic events, in particular, cerebrovascular events. We present a patient with cerebral infarction, previous deep-vein thrombosis, and miscarriages with positive lupus anticoagulant and anticardiolipin antibodies. Echocardiographic examination revealed mitral valve leaflet thickening and verrucous vegetations consistent with Libman–Sacks endocarditis, which is commonly associated with APS. In patients with combined Libman–Sacks endocarditis and antiphospholipid antibodies, anticoagulation therapy with warfarin is indicated due to high risk of valvular thrombus formation and subsequent embolization.

KEYWORDS
Antiphospholipid syndrome; Libman–Sacks endocarditis; Cerebral embolism

Introduction

Antiphospholipid syndrome (APS), either primary or secondary, is characterized by vascular thrombosis and pregnancy morbidity in addition to the presence of antiphospholipid antibodies. Several studies have reported high prevalence of valvular heart disease in APS. Echocardiographic valvular involvement is characterized by thickening of the leaflets and/or verrucous vegetations (Libman–Sacks endocarditis) and it can be associated with valvular dysfunction. Valve involvement increases the risk of thrombo-embolic complications, mainly cerebrovascular embolization.

Case report

A 46-year-old woman presented with left-sided hemiparesis. Computer tomography and magnetic resonance imaging of the brain demonstrated a right media infarction and an occlusion of a posterior branch of the right median cerebral artery.

There was an increased prevalence of thrombo-embolic disease in the family. Father and brother had deep-vein thrombosis (DVT) and a younger sister had a cerebral infarction some years ago. The medical history included two spontaneous abortions and two premature deliveries because of pre-eclampsia. In association with one of the pregnancies, she had a DVT. Except for migraine attacks, she had otherwise been healthy and did not use any regular medication.

Laboratory tests showed positive lupus anticoagulant and anticardiolipin antibodies. Protein S level was slightly reduced 39% (<65%) but other thrombophilia tests including Leiden and protrombin mutation, protein C, and antithrombin were normal. Infectious parameters (C-reactive protein 1 mg/L, leucocyte count 6.4 × 10^9/L, and sedimentation ratio 15 mm/h) and thrombocyte count (192 × 10^9/L) were also normal.

Transthoracic echocardiography demonstrated normal left ventricular geometry and function. There was a mild mitral regurgitation without dilatation of the left atrium. Thickening of the mitral valve and vegetations on the anterior and posterior mitral valve leaflets were demonstrated by transoesophageal echocardiography (Figures 1 and 2).

Anticoagulation therapy with warfarin was initiated. Clinical regression of neurological symptoms was reported during rehabilitation.

Discussion

This patient presented a classical medical history of APS, including spontaneous abortion, premature delivery because of pre-eclampsia, migraine, and past and present thrombo-embolic disease. Laboratory test with positive lupus anticoagulant and anticardiolipin antibodies confirmed the diagnosis and is consistent with the criteria for definite APS set by an international consensus statement. There
were no additional symptoms or pathology in laboratory tests, suggesting bacterial endocarditis, the main differential diagnosis.

Our patient’s echocardiography demonstrated both valve thickening and verrucous vegetations on the anterior and posterior mitral valve leaflets, suggestive of Libman–Sacks endocarditis, the typical heart valve involvement in APS, most often affecting the mitral valve. Subendothelial deposits of immunoglobulins and complement have been shown in deformed valves from APS patients.

In Libman–Sacks endocarditis, the presence of antiphospholipids antibodies promotes the formation of valve thrombi, giving rise to cardiac thrombo-embolism. Cerebral thrombo-embolic disease is the most common manifestation of APS in patients with Libman–Sacks endocarditis, as in our patient. Even though there were several cases of thrombo-embolic disease in the family, no inherited thrombophilia was revealed by the laboratory tests; except for a slight reduction in protein S, there were no mutations affecting the coagulation system. Thus, it is most likely that APS explains the thrombo-embolic events in our patient.

In conclusion, this patient’s symptoms and echocardiographic findings were consistent with APS and Libman–Sacks endocarditis. It is frequently complicated by cerebral embolism and therefore there is indication for anticoagulation therapy with warfarin.

Immunosuppressive agents should be used only for the treatment of an underlying condition which was not present in our patient.

Conflict of interest: none declared.

Supplementary material
Supplementary data associated with this article can be found in the online version.

References