Abstract

In this case, we describe a 33-year-old man presenting with acute mesenteric ischemia. When we searched for a source of embolism, a giant right atrial mass and patent foramen ovale was discovered. Standard electrocardiography showed signs of an old, silent anteroseptal wall myocardial infarction, confirmed by echocardiography and left ventriculography. Coronary angiography revealed complete occlusion of the left anterior descending artery. The diagnosis of primary antiphospholipid syndrome was confirmed by anticardiolipin antibodies test. Surgical myocardial revascularization along with the resection of the mass and the closure of the patent foramen ovale were performed. Histological examination of the operative specimen showed a thrombus. This is the first reported case presenting with acute paradoxical mesentery embolism accompanying an old myocardial infarction in a young patient with primary antiphospholipid syndrome.

Introduction

The primary antiphospholipid syndrome (PAS) is a thrombophilic disorder in which the combination of recurrent venous or arterial thrombosis and positive antiphospholipid antibodies (anticardiolipin antibodies and/or lupus anticoagulant) occurs in patients without presence of collagen vascular disorder, certain infection, or medications [Gezer 2003]. We report an unusual case with a documented giant right atrial thrombus and patent foramen ovale (PFO) presenting with acute paradoxical mesentery embolism accompanying an old myocardial infarction in a young patient with PAS, which has not been reported before.
A friable 4 cm mass was seen attached to the atrial site of the posterior tricuspid leaflet. Transesophageal echocardiography revealed a shunting of a few air microbubbles (double arrows) through a patent foramen ovale into the left atrium from the right atrium. Coronary angiography (D) shows complete occlusion of proximal left anterior descending coronary artery (arrow). LA indicates left atrium; LV, left ventricle; RA, right atrium; PTL, posterior tricuspid leaflet.

ultrasonography of the abdomen and lower limb veins were normal.

The patient was commenced on subcutaneous low molecular weight heparin when PAS was suspected, prior to starting warfarin. High-intensity anticoagulation with warfarin (international normalized ratio [INR] 3-3.5) was achieved. He was discharged after a 10-day stay in the hospital, as he felt remarkably better. Nevertheless, the right atrial mass persisted on transthoracic echocardiography, which was repeated 3 weeks later.

In a treadmill exercise stress test, performed using the Bruce Protocol, an approximately 2-mm down-sloping ST segment depression was observed in leads V3, V4, V5, and V6 at the end of the second stage, although the patient did not complain of chest pain. Coronary angiography revealed complete occlusion of the left anterior descending artery (LAD) after the first diagonal branch (Figure, D). The other coronary arteries were normal. Left ventriculography revealed anterosetal akinesia consistent with a prior myocardial infarction. The patient denied any past history of angina or hypertension and had no family history of ischemic heart disease.

Surgical treatment was recommended. A coronary artery bypass graft operation using the left internal mammarian artery was performed. A friable 4 × 4 cm mass was seen attached to the atrial site of the posterior tricuspid valve, extending down to the inferior vena cava. Subsequently, the mass was excised and foramen ovale was closed by direct suture during the same surgical procedure. Histological examination of the operative specimen showed a well-organized and calcified thrombus. High-intensity anticoagulation with warfarin was restarted and the patient was discharged without symptoms. He was followed-up at the outpatient clinic where he remained well and INR was maintained at a level between 3.0 and 3.5.

DISCUSSION

In this unusual case, there was a giant right atrial thrombus and paradoxical mesentery and, presumably, a coronary artery emboli and a high titer of anticardiolipin (ACL) antibodies. Based on these findings, the patient was diagnosed with PAS, as defined by Asherson and Cervera’s criteria [Asherson 1994]. A firm diagnosis of systemic lupus erythematosus was not made by the rheumatologists, as the patient did not fulfill the American College of Rheumatology criteria.

The case described above is noteworthy in 2 important aspects. Firstly, the acute mesenteric ischemia together with a giant right atrial thrombus and PFO has not hitherto been described in PAS. The second unusual feature of this case is the concomitance with silent myocardial infarction in a young patient with PAS who has no risk factors for ischemic heart disease.

Although several cases with intestinal infarction have been reported in PAS, association of the right atrial thrombus and PFO has not been described [Sanchez-Guerrero 1992; Kurz 1997].

Sanchez-Guerrero et al reported 2 patients with PAS who developed intestinal infarction [Sanchez-Guerrero 1992]. They explained that the cause of mesentery artery occlusion in PAS may be related to proliferative lesions of the artery or the intra-arterial thrombosis. In our case, we clearly demonstrated the right atrial thrombus and PFO, which were highly suggestive for paradoxical mesentery artery embolism.

Several case reports have described acute myocardial infarction associated with PAS in young patients [Kattwinkel 1992; Lagana 2001; Baudouy 2003]. Possible mechanisms for the development of myocardial infarction in PAS were explained in these reports as increased susceptibility to coronary artery disease due to antiphospholipid antibodies, localized thrombosis [Baudouy 2003], and myocardial microvascularopathy [Kattwinkel 1992; Lagana 2001]. The association of PAS with an old, silent myocardial infarction was described only in 1 case whose coronary angiography showed no abnormality [Lagana 2001]. It was concluded that this silent myocardial infarction could be caused by a cardiac microvascular disease accompanying the PAS. In our case, we reported a young PAS patient without any risk factors for coronary artery disease who was diagnosed with silent myocardial infarction. In his coronary angiography, the LAD was occluded completely; nevertheless there was no evidence of atherosclerotic disease in the coronary arteries. Additionally, we clearly demonstrated a giant right atrial mass and PFO. These findings are consistent with embolus of thrombus. In our opinion, this occlusion was caused by a paradoxical embolism coming from the right atrial mass, although localized coronary thrombosis could not be excluded.

Pathogenesis of the PAS is not completely understood.
Antiphospholipid antibodies have been implicated in thrombus formation, which is considered central to its clinical complications [Gezer 2003]. Antiphospholipid antibodies are directed against several phospholipid-binding proteins involved in coagulation pathways and may activate endothelial cells in the presence of β2-glycoprotein I, thus triggering thrombus generation [Roubey 1997]. High titers of anticardiolipin antibodies and, more specifically, anti-β2-glycoprotein I antibody correlate with the risk of thrombosis [Tsutsumi 1996]. Furthermore, IgG anticardiolipin antibodies are more specific for cardiac valve involvement than IgM antibodies [Hojnik 1996]. In our case, markedly elevated levels of IgG anticardiolipin antibody placed the patient at increased risk of cardiac thrombus.

Management of cardiac disease in PAS remains controversial. Recently, a committee consensus report for the treatment of cardiac disease in antiphospholipid syndrome was published [Lockshin 2003]. According to this consensus report, anticoagulation with warfarin titrated to INR of 3.0 to 3.5 and consultation with the cardiac surgeons were recommended for symptomatic patients with valvulopathy and/or intracardiac thrombus. In asymptomatic patients, prophylactic antiplatelet therapy such as aspirin and clopidogrel may be used, but this therapy is not evidence based. Previous reports indicate that valvular lesions may disappear after intense anticoagulation [Agirbasli 1997]. In our case, we preferred surgical therapy because of the persisting right atrial thrombus despite 3 weeks of high-intensity anticoagulation with warfarin.

This case emphasizes the need to carefully search for a source of embolism in a young patient presenting with an acute thrombo-embolic event, such as acute mesentery artery embolism. Immunological tests (lupus anticoagulant, anticardiolipin antibodies, etc) and meticulous evaluation of the cardiovascular system may be crucial for diagnosis and treatment options.

REFERENCES


