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RESEARCH ARTICLE

ATYPICAL PRESENTATION OF ANTIPHOSPHOLIPID SYNDROME

[1*]Gouranga Sarkar and [2]Mousumi Dutta

[1]Cardiology, Assistant Professor, Department of Medicine, I.P.G.M.E.&R., SSKM Hospital, Kolkata, India
[2]Gynaecology, Assistant Professor, Department of Gynaecology, Calcutta National Medical College, Kolkata, India

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ABSTRACT

Antiphospholipid syndrome (APS) is presented with thrombocytopenia, recurrent foetal loss, arterial & venous thrombosis. Primary APS is common compared to secondary one. Here we presented a 25 years old male with progressive exertional chest heaviness over last one year and diagnosed as chronic stable angina. Coronary angiography (CAG) revealed acute thrombotic occlusion of proximal left anterior descending (LAD) artery and underwent primary percutaneous transluminal angioplasty (PTCA) and stenting. Subsequently he received dual antiplatelet and anticoagulant.

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INTRODUCTION

Antiphospholipid syndrome is a systemic autoimmune disorder characterized by thrombocytopenia, arterial & venous thrombosis, recurrent foetal loss (Hughes, 1986) and often associated with elevated titre of Antiphospholipid antibodies (aPL), mainly the lupus anticoagulant (LA) and/or anticardiolipin antibodies (aCL). It may be primary or secondary, secondary to other connective tissue disease particularly Systemic lupus erythematosus (SLE) (Miyakis *et al*., 2000). According to revised Sapporo criteria (Asherson, 1991) for diagnosis of APLA requires a clinical manifestation of thrombosis or thrombotic related events and laboratory confirmation of an antiphopholipid antibody mainly lupus anticoagulant or anticardiolipin antibodies on two or more occasions at least 12 weeks apart. Cardiac manifestation is rare in APLA and occur in approximately 5% of patients under 45 years of age (Asherson, 1991). Cardiac manifestation include myocardial infarction, angina, valvular involvement, cardiomyopathy, vegetations, coronary artery bypass graft or stent thrombosis, intracardiac thrombus and pulmonary embolism/ hypertension (Cervera *et al*., 2002). Here a 25 years old male admitted for evaluation of chest pain and CAG reveals thrombotic occlusion of left anterior descending artery (LAD).

Corresponding author: **Gouranga Sarkar,**
Cardiology, Assistant Professor, Department of Medicine, I.P.G.M.E.&R., SSKM Hospital, Kolkata, India.

Case report: A 25 years old male non-smoker, nondiabetic, hypertensive patient referred from Nephrology unit for evaluation of chest pain. He was complained of progressive exertional chest heaviness over last one year. He was diagnosed as lupus nephritis leading to CKD stage – IV and now on immunosuppressants, antihypertensive and statin. His routine blood tests including blood sugar and fasting lipids were normal except for prolonged activated partial thromboplastin time (aPTT) 53.1 sec (n- 35.08-43.81) which did not correct after mixing with platelet poor plasma from a normal donor. 12 lead electrocardiogram revealed ST-segment depression and T wave inversion in leads V1, V2, V3, and V4. Echocardiogram showed concentric left ventricular hypertrophy along with hypokinetic anterior wall with reduced ejection fraction of 45%, normal valves and no thrombus. We planned for coronary angiography (CAG) after discussing with nephrologist about the possible need of haemodialysis. Coronary angiography performed by right femoral approach revealed thrombus containing significant lesion of proximal left anterior descending artery (LAD) with distal TIMI I flow (Fig. 1, 2). He was diagnosed as lupus nephritis three years ago and antinuclear antibody (ANA) and antiphospholipid antibody (APLA) were positive. His lupus anticoagulant (LA) was positive along with elevated IgG anticardiolipin antibody (aCL) (above 120 GPL U/ml) (n- <10) with normal IgMaCL(2.42 MPL U/ml) (n- <7) and anti b-2 glycoprotein-1 IgG/IgM antibodies. Anticardiolipin antibody was still positive at the end of three months satisfying the revised Sapporo criteria

(Miyakis *et al*., 2000). In our case it was diagnosed as secondary antiphospholipid syndrome. Therefore, percutaneous transluminal coronary angioplasty (PTCA) with stenting to LAD was successfully performed. After stent implantation TIMI III flow was achieved (Fig. 3, 4). Intravenous heparin infusion was given for next 24 hours followed by warferin with target international normalised ratio (INR) 3-4. Aspirin, clopidogrel, atorvastatin, metoprolol and ramipril were also administered following stent implantation.
His repeat echocardiography at 3 months showed improvement in ejection fraction 60%, no evidence of regional wall motion abnormality and left ventricular chamber dimension is also reduced. Patient was under follow up at 3 weeks, 6 weeks, 3 months and 6 months and now asymptomatic. Repeat coronary angiography (CAG) performed three months later showed normal coronary arteries. Stress test performed after six month is negative.

DISCUSSION

Antiphospholipid antibody syndrome (APS) are associated with autoimmune diseases such as systemic lupus erythematosus (SLE) and known as secondary APS. Sometimes they occur in isolation known as primary APLA. Primary APS is more common in young woman with female to male ratio 3.5: 1 (Cervera *et al*., 2002). Approximately, one third of patients with SLE have an antiphospholipid antibody and not all antiphospholipid antibodies suffer from thrombotic complication. The main antiphospholipids that are linked to higher thrombotic complication and atherosclerotic are anticardiolipin antibody, lupus anticoagulant and IgG antibodies against plasma phospholipid binding protein such as b2 glycoprotein I and prothrombin (Miyakis *et al*., 2006; Wilson *et al*., 1999; Park, 2004; Shi, 1990; Levine, 2002; Schultz, 1997). Lupus anticoagulant was strong risk factors for both arterial and venous thrombosis and anticardiolipin antibodies were associated with stroke and myocardial infarction. The most common clinical features of APLA are recurrent venous and arterial thrombosis, pulmonary thromboembolism and stroke.

The major cardiac events associated with APS include valve disease, myocardial infarction, intracardiac thrombus and myocardial microthrombosis (George, 2009). Actual mechanism of thrombosis of APS is not yet known, but numerous mechanism have been proposed. In APS, aPL antibodies cause thrombosis and accelerated atherosclerosis in normal vessels. The possible mechanisms of thrombosis in APS include effects of aPL antibodies on platelet membranes, on endothelial cells and on clotting components such as prothrombin, protein C and protein S leading to platelet aggregation and clot formation. In patients with APS, the aPL antibodies persist for years, possibly for a lifetime (Davies *et al*., 2007). Recently, circulating procoagulant microparticles were found to contribute to thrombotic propensity in patients with APS (Morel *et al*., 2005). The diagnostic criteria include a combination of clinical criteria(vascular thrombosis or fetal loss) and laboratory criteria (LA and/or aCL IgG or IgM, antibodies or anti-2glycoprotein-1 IgG or IgM antibodies) at least 12 weeks apart.[2] In our study patient, antibodies were persistently high even six months after the PTCA fulfilling the revised Sapporo criteria (Miyakis *et al*., 2000). Currently, angioplasty followed by stent implantation is an effective way of revascularization for thrombotic occlusion of coronary artery.

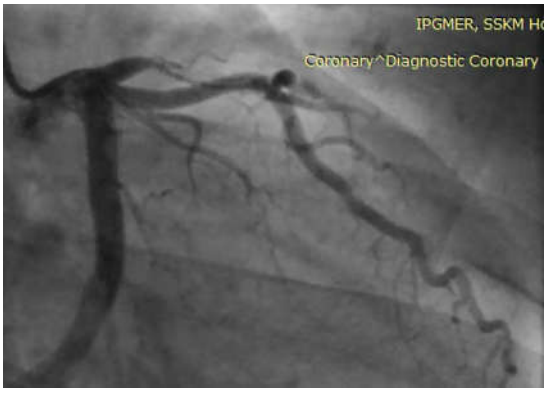

Figure 1. Coronary angiogram in RAO caudal view showing thrombus containing significant proximal LAD lesion

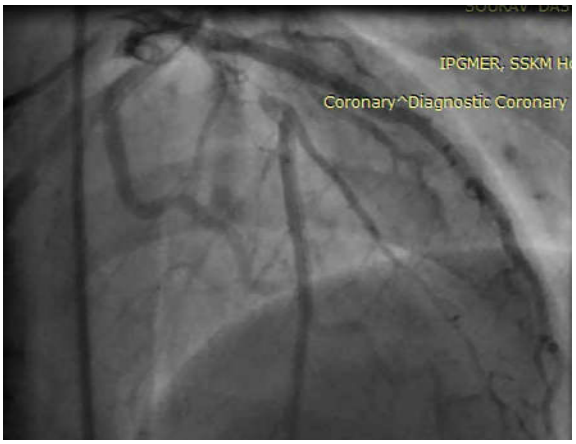

Figure 2. Coronary angiogram in RAO cranial view showing thrombus containing significant proximal LAD lesion

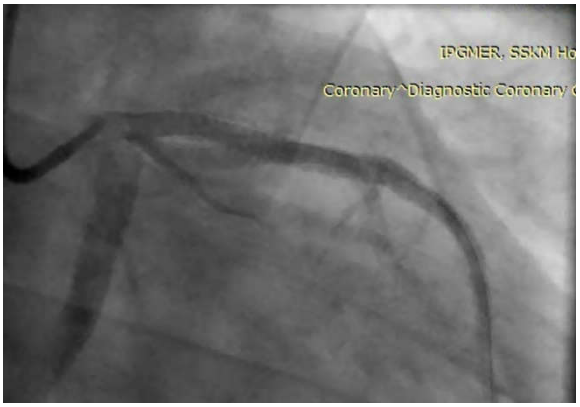

Figure 3. Coronary angiogram in RAO caudal view after successful stent implantation in LAD

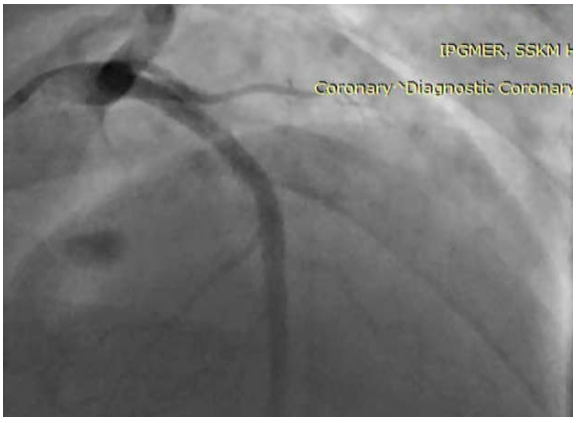

Figure 4. Coronary angiogram in RAO cranial view after successful stent implantation in LAD

In the literature, there are few cases of APS that were treated by primary angioplasty in the setting of acute myocardial infarction. Stent implantation may lead to acute thrombosis in APS patient, so balloon dilatation may be an alternative approach. Thrombotic aspiration may be an alternative approach, but it has got limited role and may be an adjunctive for balloon angioplasty without stenting. In our case after balloon angioplasty we deployed a stent in order to prevent restenosis. Therefore, long term oral anticoagulant and antiplatelet to be continued where chance of recurrent thrombosis is more. Glycoprotein IIb/IIIa inhibitor may be used as an adjunctive therapy. In our case we initiated aspirin, clopidogrel and heparin followed by warferin in order to prevent acute stent thrombosis. This type of patient should be managed with proper antiplatelet and anticoagulation as chance of recurrent coronary artery thrombosis is more. So, In our case we initiated high dose oral anticoagulation with target INR of 3-4 (Khamashta *et al*., 2005).

Conclusion

Young patient presented with chest discomfort in the setting of SLE nephritis should be evaluated for secondary APS and it is recommended to initiate and maintain long term anticoagulation and antiplatelet therapy following successful coronary angioplasty and stent implantation. Due to high rate of mortality and thrombotic burden these patients required early diagnosis and effective multimodal treatment.

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