Cardiac Manifestations on Anti-Phospholipid Syndrome

Faisal AH*
Department of Medicine, University Kebangsaan Malaysia Medical Centre, Malaysia

SHORT COMMUNICATION

Antiphospholipid syndrome may present in various ways from cutaneous manifestation, obstetric complications, neurological manifestation, and cardiac manifestation to renal involvement. There are many cardiac complications of anti-phospholipid syndrome, among them are valvular dysfunction, pulmonary hypertension, myocardial infarction, intracardiac thrombi, and ventricular dysfunction [1]. The most common cardiac manifestation is valvular abnormalities ranging from 11.6-32% [2-5].

Cardiac thrombus is an important sequela of APLS. In general, the exact mechanism of intracardiac thrombus formation in APLS is unclear. Julio and Carmen suggested that the endocardial surface might be an important site for thrombus formation in patients with circulating aPL, because these antibodies, in the presence of other hemostatic defects, will abolish the balance between thrombosis and fibrinolysis, and might change the endocardial surface factors so that clot formation is promoted [6]. Coppock, et al. Speculated that an abnormal intracardiac blood flow pattern might contribute to thrombosis [7], and Kaplan, et al. hypothesized that diffuse ventricular dysfunction might predispose to the formation of intracardiac thrombus [8].

Intracardiac thrombosis is a potentially life threatening as it can cause pulmonary and systemic embolic events; however, it is treatable condition. 50% of APLS patients are at risk of developing recurrent embolic events [9], so there must be a delicate balance between this event as well as risk of bleeding from anticoagulation.

Other cardiac manifestations that has been reported to be associated with APLS are valvular lesions, pseudoinfective endocarditis, myocardial infarction, cardiomyopathy and pulmonary hypertension [10].

Valvular lesions has been reported in SLE and APLS patients. Recently; a prospective cross sectional study done showed that positive anticardiolipin, lupus anticoagulant and anti-beta 2 glycoprotein were associated with mitral valve regurgitation; thus the need to perform transthoracic echocardiogram for early detection and management plan even in asymptomatic patients [11]. APLS may also be presents with Libman-Sacks endocarditis which may occur with a prevalence of 11% in SLE. Recently, Kotkar reported on a APLS patient with pulmonary edema with Libman-Sacks endocarditis who required mitral valve replacement [12].

Myocardial infarction occurs in 5% of aPL positive patients [10]. The presence of LA and IgG anticardiolipin antibodies at medium or high titers helps to identify APLS patients at risk for thrombosis. A research done in Tel Aviv University among 214 patients showed that 6.9% with acute myocardial infarction has elevated
antiphospholipid antibodies. Three out of 7 of them did not have risk factors of Ischaemic heart disease [13]. Young patients with vascular thrombosis and myocardial infarction with normal coronaries should be investigated for APLS [14].

Cardiomyopathy as a primary cause from APLS may be difficult to diagnose in a patient with multi-organ involvement. However, Antiphospholipid antibodies may be a useful marker. Leung et al reported that 4 out of 5 patients with isolated left ventricular dysfunction had +ve aPL. The presence of aPL were significantly associated with left ventricular dysfunction [15].

With regards to the management, aggressive treatment of all risk factors and liberal use of folic acid and vitamin B has been advocated. The use of hydroxychloroquine has been shown to have cardioprotective effect as it has suggested that it has anti-atherogenic effects [16,17]. Generally, the treatment for myocardial infarction, pulmonary hypertension and valvular disease are similar as those without APLS.

For APLS patients with thrombotic events, the duration of treatment is according to the patient’s clinical situation and the size, shape, and location of the thrombus. Lifelong warfarin may be an option in the presence of underlying cardiomyopathy which may predispose to further intracardiac thrombosis. A randomized clinical trial done comparing high intensity warfarin (INR 3.0-4.0) versus conventional antithrombotic therapy for the prevention of recurrent thrombosis in patients with antiphospholipid syndrome showed that high intensity warfarin was not superior and is associated with increased risk of minor hemorrhagic complications [18]. A current review states that for patients with definite APLS with first venous thrombotic event, prolonged use of anticoagulation to maintain target INR of 2.0-3.0 is recommended. For those with arterial thrombosis, proposed therapies include: anticoagulation with target INR 3.0-4.0; antiplatelet therapy alone; anticoagulation with target INR 2.0-3.0, or combination of antiplatelet or anticoagulation therapy with target INR 2.0-3.0 [19].

This manuscript illustrates the various cardiac presentations of APLS. The occurrence of thrombotic phenomena, especially in young patients without risk factors, should prompt further investigation towards APLS, i.e. aPL antibodies. Estimation of aPL in cardiological practice assumes considerable importance today.

REFERENCES

1. F Tenedios, D Erkan, MD Lockshin. Cardiac involvement in the Antiphospholipid Syndrome. Lupus. 2005; 14: 691-696. Ref.: https://goo.gl/WduHpp

2. Asherson RA, Khamashta MA, Ordi-Ros J, Derksen RH, Machin SJ, et al. The primary antiphospholipid syndrome: Major clinical and serological features. Medicine (Baltimore). 1989; 68: 366-734. Ref.: https://goo.gl/4ezdoF

3. Leventhal JL, Borofsky MA, Bergey PD, Schumacher HR. Antiphospholipid antibody syndrome with right atrial thrombosis mimicking an atrial myxoma. Am J Med. 1989; 87: 111-113. Ref.: https://goo.gl/6Of0FH

4. Gertner E, Leatherman JW. Intracardiac mural thrombus mimicking atrial myxoma in the antiphospholipid syndrome. J Rheumatol. 1992; 19: 1293-1298. Ref.: https://goo.gl/zTe5DU

5. Love PE, Santoro SA. Antiphospholipid antibodies: Anticardiolipin and the lupus anticoagulant in systemic lupus erythematosus (SLE) and on non-SLE disorders. Ann Intern Med. 1990; 112: 682-698. Ref.: https://goo.gl/i4JSGO

6. Julio AA, Carmen S. Intracardiac thrombus in antiphospholipid antibody syndrome. J Am Soc Echocardiogr. 2000; 13: 873-875. Ref.: https://goo.gl/Ih4SGK

7. Coppock MA, Safford RE, Danielson GK. Intracardiac thrombosis, phospholipid antibodies, and two-chambered right ventricle. Br Heart J. 1988; 60: 455-458. Ref.: https://goo.gl/zzZa4n

8. Kaplan SD, Chartash EK, Pizzarello RA, Furie RA. Cardiac manifestations of the antiphospholipid syndrome. Am Heart J. 1992; 124: 1331-1338. Ref.: https://goo.gl/dHgKls
9. Rosove MH, Brewer PM. Antiphospholipid thrombosis: Clinical course after the first thrombotic event in 70 patients. Ann Intern Med. 1992; 117: 303-308. Ref.: https://goo.gl/lFk1sF

10. Asherson RA, Cervera R. Antiphospholipid antibodies and the heart. Lessons and pitfalls for the cardiologist. Circulation. 1991; 84: 920-923. Ref.: https://goo.gl/cn9W7F

11. Mohammed AG, Alghamdi AA, Aljahan MA, Al-Homood IA. Echocardiographic findings in asymptomatic systemic lupus erythematosus patients. Clin Rheumatol. 2017; 36: 563. Ref.: https://goo.gl/RwEjIo

12. Kotkar KD, Said SM. Libman-Sacks Endocarditis in a Patient With Antiphospholipid Syndrome. Ann Thorac Surg. 2016; 102: e31-e32. Ref.: https://goo.gl/0LDy9Z

13. Adler Y, Finkelstein Y, Zandeman-Goddard G, Blank M, Lorber M, et al. The presence of antiphospholipid antibodies in acute myocardial infarction. Lupus. 1995; 4: 309-313. Ref.: https://goo.gl/wy1i6g

14. Prashanth P, Mukhaimi M, Riyami A. A rare presentation of Primary Antiphospholipid Syndrome. Oman Med J. 2009; 24: 300-302. Ref.: https://goo.gl/ta6tkt

15. Leung WH, Wong KL, Lau CP, Wong CK, Cheng CH. Association between antiphospholipid antibodies and cardiac abnormalities in patients with systemic lupus erythematosus. Am J Med. 1990; 89: 411-419. Ref.: https://goo.gl/Ne21eb

16. Lockshin M, Tenedios F, Petri M, McCarty G, Forastiero R, et al. Cardiac disease in the antiphospholipid syndrome. Lupus. 2003; 12: 518-523. Ref.: https://goo.gl/ILC2d0

17. Molad Y, Gorshtein A, Wysenbeek AJ, Guedj D, Majadla R, et al. Protective effect of hydroxychloroquine in systemic lupus erythematosus. Prospective long-term study of an Israeli cohort. Lupus. 2002; 11: 356-361. Ref.: https://goo.gl/8JBBh5

18. G Finazzi, R Marchioli, Brancaccio V, Schinco P, Wisloff F, et al. A Randomized clinical trial done comparing high intensity warfarin vs. conventional antithrombotic therapy for the prevention of recurrent thrombosis in patients with antiphospholipid syndrome (WAPS). J Thromb Haemost. 2005; 3: 848-853. Ref.: https://goo.gl/dT9qke

19. Espinosa G, Cervera R. Current treatment of antiphospholipid syndrome: lights and shadows. Nat Rev Rheumatol. 2015; 11: 586-596. Ref.: https://goo.gl/0FNsF5