

An Unusual Cause of Multiple Left Ventricle Thrombi in a Young Woman

Sofien Zayed¹, Rami Tlili¹, Fares Azaiez¹, Rim Hentati¹, Fatma Zouari¹, Rim Ben Romdhane¹, Rania Hammami², Youssef Ben Ameer¹

¹Department of Cardiology, Mongi Slim University Hospital La Marsa, Tunis, Tunisia; ²Department of Cardiology, HediChaker University Hospital, Sfax, Tunisia.

Corresponding Author:

Dr. Zayed Sofien
Email: sofienzayed91@gmail.com

This is an Open Access article distributed under the terms of the Creative Commons Attribution License (creativecommons.org/licenses/by/3.0).

Received : January 7, 2020
Accepted : July 28, 2020
Published : September 15, 2020

Abstract

Background: Antiphospholipid syndrome (APS) is a rare autoimmune disease with an increased risk of vascular occlusion and pregnancy complications. As a multisystemic disease, the heart is usually affected by direct (autoimmune action) or indirect (thrombosis) pathological mechanisms. **Case Report:** A 40-year-old diabetic and hypertensive woman presented with congestive heart failure. The echocardiographic exploration showed reduced left ventricle ejection fraction (20%) and diffuse wall motion abnormalities with multiple large thrombi in the left ventricle. The coronary angiography was normal. The APS was confirmed by positive laboratory tests (La, aCL and aβ2GPI). **Conclusion:** The revelation of APS by cardiac manifestations, mainly cardiac thrombosis, is rarely reported in the literature, and evolution is usually unfavorable..

Keywords: Coronary Angiography, Echocardiography, Heart Diseases, Lupus Coagulation Inhibitor, Thrombosis.

Introduction

Antiphospholipid syndrome (APS) is an autoimmune disorder characterized by the persistent presence (more than 12 weeks apart) of antiphospholipid antibodies (aPLs) leading to a state of hypercoagulability associated with vascular thrombosis and loss of pregnancy [1]. Because of the vascular nature, several organs are affected, among which the cardiovascular system and more particular is the cardiac involvement which can be valvular, coronary or myocardial.

Cardiac manifestations may be found in up to 40% of patients with the APS, but significant morbidity appears in only 4% to 6% of these patients [2]. In fact, if arterial or peripheral venous thromboses are common in the APS, intracardiac thromboses are more rarely described [3,4]. Indeed, this is a rare event, reported in only 0.4 % of cases in the “Euro-APS” study [5]. The multiple mechanisms causes cardiac damage and are still

poorly understood. They are linked not only to thrombosis [6], but also to immune lesions [7]. We report the case of APS revealed by multiple cardiac thrombi.

Case Report

A 40-year-old woman, with history of hypertension and diabetes, presented to the emergency department for congestive heart failure. She was non-smoker, obese (BMI = 30 kg/m²), with no history of addiction and similar prior episode. The patient was married and she had two children.

Echocardiographic exploration showed a normal sized left ventricle with a reduced left ventricle ejection fraction (LVEF=20%) and hypokinesia with multiple large thrombi in the left ventricle [Fig.1]. There were no significant valvulopathies other than a minimal secondary mitral regurgitation. The systolic function of the right ventricle was reduced. The coronary

angiography didn't show any abnormality. Faced with this clinical picture in a young woman, thrombosis in the cardiovascular system, heart in this case, a strong clinical suspicion of APS was raised. This was confirmed with all three positive tests (La, aCL and β 2GPI). The patient was treated by congestive heart failure treatment [intravenous diuretics, angiotensin converting enzyme inhibitors (ACE) and effective anticoagulation]. Corticosteroid therapy was not initiated because the patient was congestive. Evolution was marked by the death of patient 3 months later because of refractory heart failure.

Discussion

Ventricular dysfunction in APS may result from the direct effect of antibodies in the myocardium or microvascular thrombosis [8-10]. Of these, microvascular injury is probably the most feared complication and may not be recognized because its detection requires specific diagnostic techniques [11]. In fact, the true prevalence of myocardial ischemia in APS is unknown. In one study, the prevalence of unrecognized myocardial scars detected by cardiac MRI was 30% [12]. Limited data are available for abnormalities of ventricular function in APS. Several reports have demonstrated the presence of aPL with otherwise unexplained ventricular dysfunction (by conventional risk factors), especially in young people [8,10,13]. Some studies have found abnormalities of ventricular relaxation in patient with primary APS compared to controls [14].

The only effect of APS on left ventricular function is difficult to extrapolate, as patients included in the studies had other known comorbidities to cause left ventricular dysfunction. The pathogenesis of ventricular dysfunction in APS in the absence of valve dysfunction or coronary artery disease is unclear. The diagnosis of APS should be discussed in patients with intracardiac thrombi without underlying cardiac disease. Histological findings range from microvascular thrombosis to

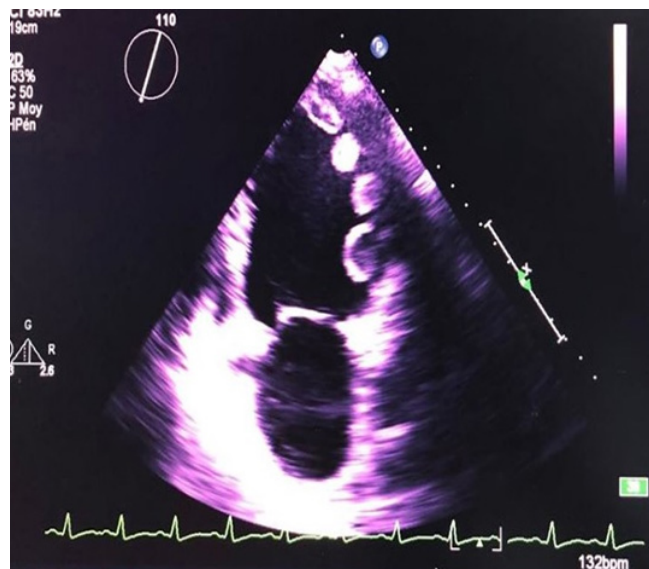


Fig.1: Echocardiographic evidence of multiple large thrombi in the left ventricle.

endomyocardial fibrosis. Myocardial involvement has also been documented in patients with APS and could be explained by repeated myocardial microthrombosis or endothelial lesions [8-10]. Regional movement abnormalities in the left ventricle have been correlated with the presence of high levels of aPL [15]. In addition, the right ventricle appears to be more involved than the left one [16].

Strategies for treating cardiovascular disease associated with APS include aggressive control of all traditional risk factors, both through lifestyle changes and drug therapy [17]. Therapeutic options in APS associated cardiovascular diseases include antiplatelet agents and anticoagulants, angiotensin converting enzyme inhibitors, beta blockers, and statins. Specific drugs may be beneficial, including folic acid, B vitamins and possibly hydroxychloroquine [17]. Evidence has recently emerged indicating that the effects of anti-rheumatic therapy may go beyond controlling inflammation and disease activity, including the endothelial effects of anti-tumor necrosis factor (TNF) α agents and the effects of conventional therapy and biological drugs on vascular function [11].

Conclusion

The APS is a multi-systemic disease and the cardiac manifestations and cardiac involvement is not a benign pathology. Although cardiac events are not common in APS, they can rapidly move from asymptomatic to severe life-threatening events. Timely diagnosis and knowledge of the pathophysiology of APS and its manifestations can help therapeutic decision-making, in an area where management is based on consensus and on a fundamentally empirical therapy.

Contributors: ZS, TR: manuscript writing, literature review, and patient management; AF, HR: manuscript editing, references and patient management. ZF, BRR, HR, BAY: critical inputs into the manuscript. ZS will act as a study guarantor. All authors approved the final version of this manuscript and are responsible for all aspects of the study.

Funding: None; *Competing interests:* None stated.

References

- Miyakis S, Lockshin MD, Atsumi T, Branch DW, Brey RL, Cervera R, *et al.* International consensus statement on an update of the classification criteria for definite antiphospholipid syndrome (APS). *J Thromb Haemost JTH.* Févr. 2006;4:295-306.
- Cervera R. Coronary and valvular syndromes and antiphospholipid antibodies. *Thromb Res.* 2004;114:501-507.
- Leventhal LJ, Borofsky MA, Bergey PD, Schumacher HR. Antiphospholipid antibody syndrome with right atrial thrombosis mimicking an atrial myxoma. *Am J Med Juill.* 1989;87:111-113.
- Gertner E, Leatherman JW. Intracardiac mural thrombus mimicking atrial myxoma in the antiphospholipid syndrome. *J Rheumatol. Août.* 1992;19:1293-1298.
- Cervera R, Piette JC, Font J, Khamashta MA, Shoenfeld Y, Camps MT, *et al.* Antiphospholipid syndrome: clinical and immunologic manifestations and patterns of disease expression in a cohort of 1,000 patients. *Arthritis Rheum. Avr.* 2002;46:1019-1027.
- Tripodi A, de Groot PG, Pengo V. Antiphospholipid syndrome: laboratory detection, mechanisms of action and treatment. *J Intern Med Août.* 2011;270:110-122.
- Jara LJ, Medina G, Vera-Lastra O, Amigo MC. Accelerated atherosclerosis, immune response and autoimmune rheumatic diseases. *Autoimmun Rev Mars.* 2006;5:195-201.
- Lauwerys BR, Lambert M, Vanoverschelde JL, Cosyns JP, Houssiau FA. Myocardial microangiopathy associated with antiphospholipid antibodies. *Lupus.* 2001;10:123-125.
- Azeem T, Vassallo M, Samani NJ. Images in cardiology. Endomyocardial fibrosis associated with antiphospholipid syndrome. *Heart Br Card Soc Août.* 2000;84:156.
- Murphy JJ, Leach IH. Findings at necropsy in the heart of a patient with anticardiolipin syndrome. *Br Heart J Juill.* 1989;62:61-64.
- Sitia S, Atzeni F, Sarzi-Puttini P, Di Bello V, Tomasoni L, Delfino L, *et al.* Cardiovascular involvement in systemic autoimmune diseases. *Autoimmun Rev Févr.* 2009;8:281-286.
- Sacré K, Brihaye B, Hyafil F, Serfaty JM, Escoubet B, Zennaro MC, *et al.* Asymptomatic myocardial ischemic disease in antiphospholipid syndrome: a controlled cardiac magnetic resonance imaging study. *Arthritis Rheum Juill.* 2010;62:2093-2100.
- Greisman SG, Thayaparan RS, Godwin TA, Lockshin MD. Occlusive vasculopathy in systemic lupus erythematosus. Association with anticardiolipin antibody. *Arch Intern Med Févr.* 1991;151:389-392.
- Leung WH, Wong KL, Lau CP, Wong CK, Liu HW. Association between antiphospholipid antibodies and cardiac abnormalities in patients with systemic lupus erythematosus. *Am J Med.* 1990;89:411-419.
- Nihoyannopoulos P, Gomez PM, Joshi J, Loizou S, Walport MJ, Oakley CM. Cardiac abnormalities in systemic lupus erythematosus. Association with raised anticardiolipin antibodies. *Circulation Août.* 1990;82:369-375.
- Tektonidou MG, Ioannidis JP, Moysakakis I, Boki KA, Vassiliou V, Vlachoyiannopoulos PG, *et al.* Right ventricular diastolic dysfunction in patients with anticardiolipin antibodies and antiphospholipid syndrome. *Ann Rheum Dis Janv.* 2001;60:43-48.
- Lockshin M, Tenedios F, Petri M, McCarty G, Forastiero R, Krilis S, *et al.* Cardiac disease in the antiphospholipid syndrome: recommendations for treatment. Committee consensus report. *Lupus.* 2003;12:518-523.