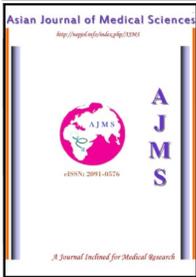


ASIAN JOURNAL OF MEDICAL SCIENCES



Primary Coronary Sinus Thrombosis in Secondary Antiphospholipid Antibody Syndrome

Joseph Theodore^{1*}, P Chitrambalam¹, K Pradeep² and S Visvakumar²

¹Department of Internal Medicine and ²Department of Cardio-Thoracic Surgery, Madras Medical College and Government General Hospital, Chennai, India.

Abstract

Antiphospholipid antibody syndrome (APLA) is a non-inflammatory autoimmune disease characterised by spontaneous abortion, thrombocytopenia and thrombosis (arterial and venous). Intracardiac thrombosis is a rare complication of APLA, but coronary sinus thrombosis in APLA has hitherto not been reported. We recently treated a young woman with secondary APLA and systemic lupus erythematosus in whom coronary sinus thrombosis was detected in association with recurrent pulmonary embolism.

Key Words: intracardiac thrombosis; antiphospholipid antibody syndrome; systemic lupus erythematosus; coronary sinus thrombosis

1. Introduction

The antiphospholipid antibody syndrome (APLA) is an autoimmune disorder presenting with tissue injury in various organs attributable to large or small vessel thrombosis or, in some instances, possible nonthrombotic inflammatory mechanisms, associated with in vitro evidence of antibodies (lupus anticoagulant, IgG or IgM anticardiolipin, or IgG or IgM anti-beta2-glycoprotein I) to certain proteins, or proteinphospholipid complexes.¹ The frequency of thrombosis is reported to be greater in APLA associated with systemic lupus erythematosus (SLE) than in primary APLA; risk factors have also been found to differ for venous and arterial thrombosis in APLA.²

Coronary sinus thrombosis usually occurs as a complication of cardiac transplantation and right heart catheterization in non-infected patients, and rarely in the absence of such procedures.³ Coronary sinus thrombosis in APLA has hitherto not been reported. We recently treated a young woman with secondary APLA and SLE in whom coronary sinus thrombosis was detected in association with recurrent pulmonary embolism.

2. Case Report

In May 2009, a 31 year old woman presented at the emergency room of this hospital with sudden-onset chest discomfort and exertional dyspnea of class III severity (New York Heart Association criteria). The patient was known to be suffering from hypothyroidism, for which she had been on a supplementary dose of thyroxine (100µg/day) from June, 2007. She had recently been diagnosed to have hypertension, for which she was receiving amlodipine (5 mg/day).

At the time of presentation, physical examination revealed a blood pressure of 160/100 mm Hg, heart rate of 100/min and respiratory rate of 24/min. The jugular venous pressure was normal. All heart sounds were regular, but the pulmonic component of the 2nd heart sound (P₂) was accentuated. The lungs were clear and the abdomen was soft; no peripheral edema was found. Acanthosis nigricans and livedo reticularis, markers of autoimmunity, were noted. A chest radiograph showed a normal-sized heart with a relative decrease in right-sided bronchovascular markings, a finding suggestive of segmental pulmonary arterial occlusion. An electrocardiogram showed sinus tachycardia with right-axis deviation and symmetrical 'T' inversion in leads V₁, V₂, and V₃. A colour Doppler and duplex scan ultrasonogram of the lower limbs excluded deep vein thrombosis.

*Correspondence:

Dr. Joseph Theodore, Thomas Diagnostic & Research Centre, 50-A Melapudur Main Rd., I Floor, Tiruchirapalli 620001, Tamilnadu, India. Mobile: +91-9940863963 e-mail: philipthomas@sify.com

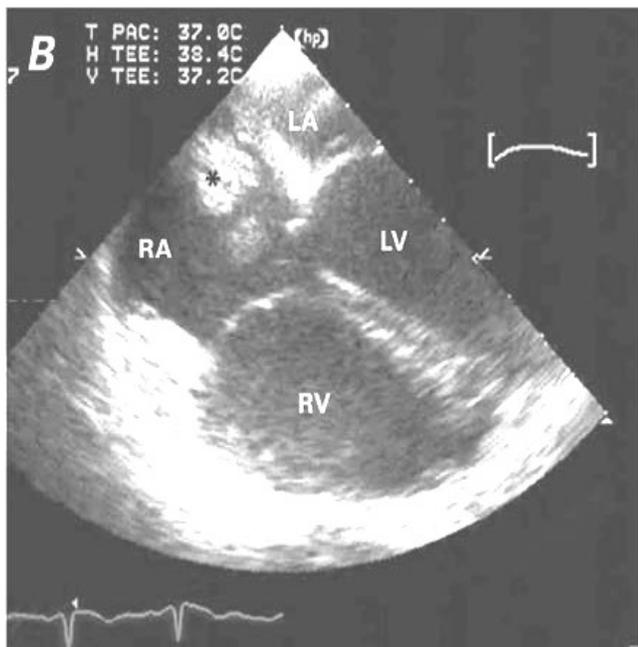
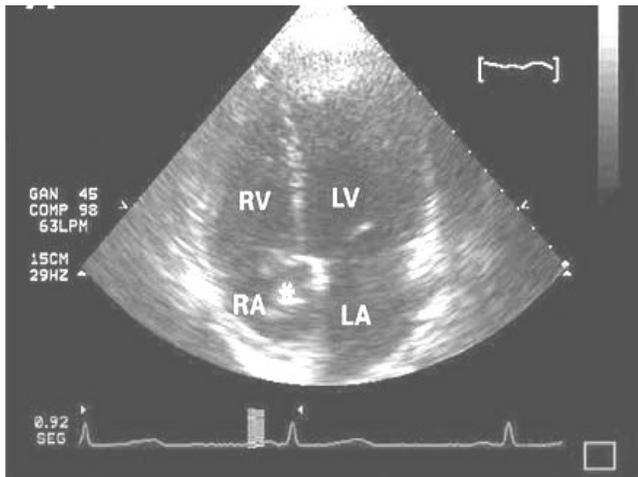


Figure-1: A. Transthoracic echocardiography showing an hyperechoic signal at the point of entry of the coronary sinus into the right atrium B. Transesophageal echocardiography confirming the transthoracic echocardiography finding. RA=right atrium; RV=right ventricle; LV= left ventricle; LA= left atrium

Two-dimensional transthoracic echocardiography revealed normal dimensions of the left ventricle and left atrium. Doppler echocardiography of the heart revealed moderate pulmonary hypertension, with the pulmonary arterial systolic pressure exceeding 50 mmHg. A hyperechoic signal was obtained at the site of entry of the coronary sinus into the right atrium (Fig. 1A). In view of these findings, transesophageal echocardiography was performed and this confirmed the presence of a mobile, friable mass in the coronary sinus (Fig. 1B). A spiral computerized tomograph of the chest showed occlusion of multiple right lower lobe segmental arteries with a calcified thrombus in the coronary sinus at the site of entry into the right atrium (Fig. 2). Further

investigations revealed that the patient was suffering from systemic lupus erythematosus with secondary antiphospholipid antibody syndrome (APLA), which was the possible cause of the thrombosis. Oral anticoagulant therapy was commenced (warfarin sodium, 3 mg once daily), and the prothrombin time (INR) was maintained at 2.5. The patient underwent surgery. During cardiopulmonary bypass, the coronary sinus was opened and the mass was removed. Histological examination of the mass revealed an organized thrombus with a fibrinous network and calcification.

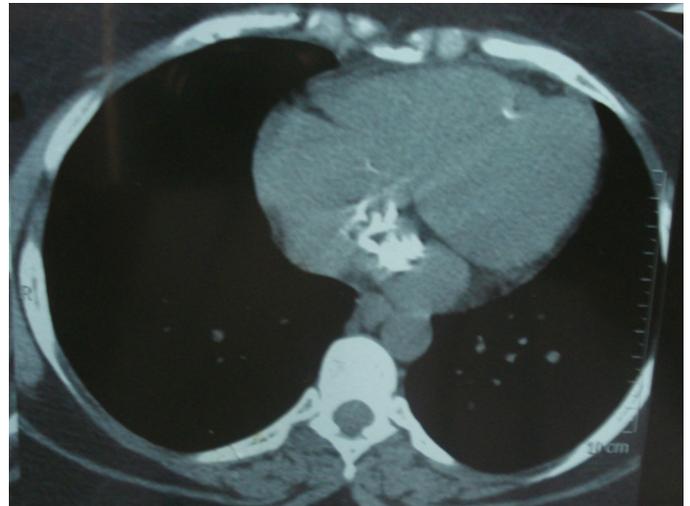


Figure-2: A 64- slice computerized tomogram with contrast revealed the presence of a calcified thrombus in the coronary sinus jutting into the right atrium.

The patient experienced no post-operative complications. Echocardiogram was repeated after two weeks; this showed a considerable decrease in pulmonary systolic pressure and a patent coronary sinus. The patient is currently being reviewed on a regular basis, and continues to receive warfarin, steroids, amlodipine and thyroxine

3. Discussion

The coronary sinus is the conduit at the posterior atrioventricular groove which drains into the right atrium. Coronary sinus thrombosis is a rare acquired anomaly. Although cases of primary coronary sinus thrombosis have been reported, most cases occur due to endothelial damage following invasive cardiac procedures involving the right atrium, such as central venous line placement, right ventricle pacing, cardiac resynchronising therapy and cardiac transplant.^{4,6} *Aspergillus fumigatus* thrombi causing total occlusion of both coronary arterial ostia, all four major epicardial

coronary arteries and coronary sinus and associated with purulent pericarditis has also been reported.⁷

APLA is characterized by arterial and venous thrombosis.⁸⁻¹⁰ The frequency of thrombosis and pregnancy loss (another feature of APLA) is greater in APLA associated with SLE than in primary APLA.^{2,10} Intracardiac thrombus has rarely been reported as a complication of APLA.¹⁰ To our knowledge, primary coronary sinus thrombosis due to SLE with secondary APLA has hitherto not been reported. A search of the Pub Med database using the keywords 'coronary sinus thrombosis' and 'antiphospholipid antibody syndrome' only generated two results (neither were relevant in the context of the present case); the use of the keywords 'coronary sinus thrombosis' and 'systemic lupus erythematosus' also generated two references that were not relevant to the present context.

The clinical course of coronary sinus thrombosis is unpredictable. Most cases remain asymptomatic and are detected only incidentally at angiography done for other purposes or at autopsy. Fatal complications, such as sudden cardiac death, venous haemorrhagic myocardial infarction and pericardial tamponade have been reported.³⁻⁶ It can also serve as a nidus for recurrent pulmonary embolism and lead to pulmonary hypertension, as in our patient. In symptomatic patients, heparin therapy followed by oral anticoagulants and open heart thrombectomy may be rewarding.

4. Conclusion

APLA is a non-inflammatory autoimmune disease characterised by spontaneous abortion, thrombocytopenia and thrombosis (arterial and venous). Intracardiac thrombosis is a rare complication of APLA. To our knowledge, primary coronary sinus thrombosis due to systemic lupus erythematosus with secondary APLA has hitherto not been reported. Coronary sinus thrombosis can be a source of recurrent pulmonary embolism as in our patient. Additional cases and reports are needed to confirm that the coronary sinus may serve as another site of thrombosis, which can lead to pulmonary embolism. Oral anticoagulants and resection of the thrombus enabled our patient to recover without major complications.

5. References

1. Eby C. Antiphospholipid syndrome review. Clin Lab Med 2009; 29(2):305-19. [doi:10.1016/j.cll.2009.06.001](https://doi.org/10.1016/j.cll.2009.06.001) PMID:19665680
2. Danowski A, de Azevedo MN, de Souza Papi JA, Petri M. Determinants of risk for venous and arterial thrombosis in primary antiphospholipid syndrome and in antiphospholipid syndrome with systemic lupus erythematosus J Rheumatol 2009; 36(6):1195-9. [doi:10.3899/jrheum.081194](https://doi.org/10.3899/jrheum.081194) PMID:19447935
3. Ramsaran EK, Sadigh M, Miller D. Sudden cardiac death due to primary coronary sinus thrombosis. South Med J 1996; 89:531-3. [doi:10.1097/00007611-199605000-00019](https://doi.org/10.1097/00007611-199605000-00019) PMID:8638186
4. Guindi MM, Walley VM. Coronary sinus thrombosis: a potential complication of right heart catheterization Can J Surg 1987; 30(1):66-7. PMID:3815188
5. Urbanová D. Coronary sinus thrombosis in patients after heart transplantation. Cor Vasa 1989; 3(3):231-7. PMID:2670448
6. Wells CA, Senior AJ. Coronary sinus thrombosis and myocardial infarction secondary to ventriculoatrial shunt insertion. J Pediatr Surg 1990; 25(12):1214-5. [doi:10.1016/0022-3468\(90\)90506-5](https://doi.org/10.1016/0022-3468(90)90506-5)
7. Ross EM, Macher AM, Roberts WC. Aspergillus fumigatus thrombi causing total occlusion of both coronary arterial ostia, all four major epicardial coronary arteries and coronary sinus and associated with purulent pericarditis. Am J Cardiol 1985; 56(7):499-500. [doi:10.1016/0002-9149\(85\)90904-X](https://doi.org/10.1016/0002-9149(85)90904-X)
8. Rosove MH, Brewer PM. Antiphospholipid thrombosis: clinical course after the first thrombotic event in 70 patients. Ann Intern Med 1992; 117(4):303-8. PMID:1637025
9. Finazzi G, Brancaccio V, Moia M, Ciaverella N, Mazzucconi MG, Schinco PC, et al. Natural history and risk factors for thrombosis in 360 patients with antiphospholipid antibodies: a four-year prospective study from the Italian Registry. Am J Med 1996; 100(5):530-6. [doi:10.1016/S0002-9343\(96\)00060-5](https://doi.org/10.1016/S0002-9343(96)00060-5)
10. Soltész P, Veres K, Lakos G, Kiss E, Muszbek L, Szegedi G. Evaluation of clinical and laboratory features of antiphospholipid syndrome: a retrospective study of 637 patients. Lupus 2003; 12(4):302-7. PMID:12729054