Antiphospholipid syndrome and right atrial mass

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Clinical Summary

A 43-year-old woman was urgently admitted to the hospital because of a syncopal episode. In the past she had had repeated spontaneous abortions, venous thrombotic events during pregnancy, and thrombocytopenia associated with lupus anticoagu-

lant positivity. A diagnosis of antiphospholipid syndrome (APS) had been suggested 14 years earlier. Results of cardiac physical examination and a 12-lead electrocardiogram were unremarkable. Two-dimensional transthoracic and transesophageal echocardiography showed ventricular and left atrial chambers normal in size and function, whereas a polypus, mobile mass attached to the free wall was visualized in the right atrium (**Figure 1, A and B**). Semilunar and atrioventricular valves were normal, without any evidence of vegetations. The patient underwent surgical removal of the mass, and pathologic examination ruled out a neoplastic or infective nature, by showing a fibrin network entrapping blood cells with calcific deposits at its implantation base (**Figure 2**). The histologic features were in keeping with an organized thrombosis, and a diagnosis of nonbacterial thrombotic endocarditis of the right atrial endocardium in the setting of APS was suggested. At 6 months' follow-up, the patient is doing well with long-term anticoagulant therapy. She has no evidence of intracardiac mass recurrence on 2-dimensional echocardiography (**Figure 1, C**).

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Figure 1. A and B, Two-dimensional transesophageal echocardiogram shows a polypus mass in the right atrium attached to the free wall and free floating during the cardiac cycle so as to occasionally impinge into the tricuspid valve orifice. C, No evidence of intra-atrial mass recurrence at 2-dimensional transthoracic echocardiography at 6 months' follow-up.

Figure 2. A, Gross examination of the resected mass reveals a reddish elongated neoformation with a rough surface, $3.5 \times 0.8 \times 0.9$ cm in size and 2.3 g in weight. B, At histologic examination, the mass consisted of a fibrin network entrapping red cells, platelets, and leukocytes with focal early organization into fibrous tissue.

Discussion

APS is defined according to the clinical and laboratory criteria established by an international consensus, that is, history of thrombosis and/or pregnancy morbidity, associated with the presence in the blood of lupus anticoagulants and/or anticardiolipin antibodies of immunoglobulin G and/or immunoglobulin M isotype, positive

in medium or high titers on 2 or more occasions at least 6 weeks apart.¹ APS usually is classified as primary and secondary, respectively, accordingly to the absence or presence of an underlying disease as well as other autoimmune, malignancy, or drug-induced disorders. In both instances, every organ can be involved by thrombotic events. In a recent study of a cohort of 1000 patients affected by primary and secondary APS, cardiac involvement was observed in 27% of cases,² mostly characterized by valvular and coronary artery disease. The most frequent echocardiographic findings are either valve vegetations or leaflets thickening. The literature reports only a few cases of intracardiac thrombosis³ associated with primary APS, observed in all cardiac chambers, provoking either pulmonary or systemic embolism.

The clinical history, along with the stoppage of anticoagulant therapy for many years, led to a diagnosis of atrial thrombosis. The treatment of patients affected by APS is usually based on a healthy life-style, by abstaining from smoking and intake of oral contraceptives, by reducing the risk for vein stasis and vascular injuries, and by performing thromboprophylaxis during pregnancy. Moreover, in patients who have already experienced thrombotic events, it is mandatory to prevent recurrent thrombosis by intermediate to high-intensity warfarin therapy.⁴ Whether thrombolysis, with a high-intensity anticoagulation protocol based on warfarin, or a surgical excision is the better first therapeutic approach remains unknown. However, the great size of the mass and the free-floating appearance at 2-dimensional echocardiogram, which seemed at high risk for pulmonary embolism, led us to choose surgical therapy. Moreover, the pathologic features of an organized thrombosis with early calcific deposits, which could constitute the nucleus for further thrombus deposition, further support the surgical approach.

Since relapsing thrombosis has been reported in both the arterial and venous bed despite long-term warfarin treatment, a strict follow-up by 2-dimensional echocardiogram should be recommended in patients affected by APS syndrome.

References

1. Wilson WA, Gharavi AE, Koike T, et al. International consensus statement on preliminary classification criteria for definite antiphospholipid syndrome. *Arthritis Rheum.* 1999;42:1309-11.
2. Cervera R, Piette JC, Font J, et al. Antiphospholipid syndrome: clinical and immunologic manifestations and patterns of disease expression in a cohort of 1,000 patients. *Arthritis Rheum.* 2002;46:1019-27.
3. Plain D, Van Camp G, Efra A, et al. Intracardiac thrombi associated with antiphospholipid antibodies. *J Am Soc Echocardiogr.* 1996;9:891-3.
4. Roubey RAS. New approaches to prevention of thrombosis in the antiphospholipid syndrome: hopes, trials, and tribulations. *Arthritis Rheum.* 2003;48:3004-8.