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LETTERS TO THE EDITOR

CLINICAL-SCIENTIFIC NOTES

Intracardiac thrombosis complicating antiphospholipid antibody syndrome

A 40-year-old woman with a 5-year history of systemic lupus erythematosus (SLE) presented with dyspnoea and haemoptysis. The diagnosis of SLE was based on the presence of a typical rash, polyarthralgia and serology (antinuclear, double-stranded DNA and Sjogren's syndrome antigen A (SS-A) antibodies). At presentation, computer tomography pulmonary angiography demonstrated a pulmonary embolus and also showed a right atrial mass. *Trans*-oesophageal echocardiography confirmed a frond-like right atrial mass, but could not differentiate endocardial thrombus from myxoma (Fig. 1). Other laboratory abnormalities included marked thrombocytopenia (19×10^9/L), an elevated activated partial thromboplastin time (56 s) and raised IgG anticardiolipin (aCL)-type antiphospholipid (aPL) antibodies (656 IgG antiphospholipid antibody units (GPL), normal range 0–5 GPL). Tests for other procoagulant conditions were negative.

Anticoagulation with heparin followed by warfarin was commenced and prednisolone (1 mg/kg per day) was given for autoimmune thrombocytopenia. At 6 weeks, the platelet count was 45×10^9/L; however, echocardiography showed the mass was unchanged in size.

Excisional biopsy of the cardiac mass as well as tricuspid valve replacement were performed. Histology showed mural thrombus associated with endocardial fibrosis. Recovery was uneventful. Prednisolone was tapered and warfarin anticoagulation was recommenced. At 6-month follow up, she was symptom-free with a platelet count of 80×10^9/L.

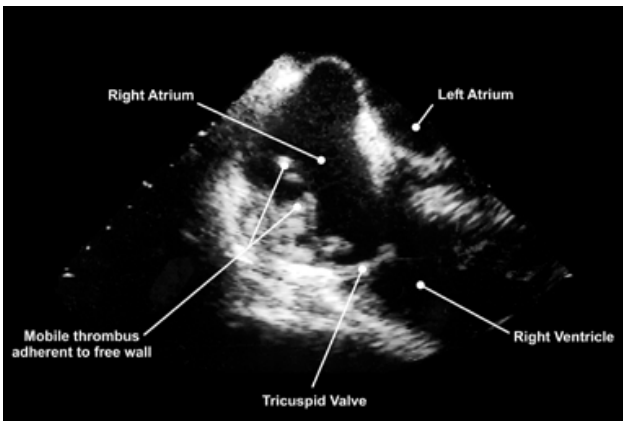

Figure 1 *Trans*-oesophageal echocardiogram of right atrial thrombi.

Common clinical syndromes associated with the antiphospholipid syndrome (APS) include venous and arterial thrombosis, thrombocytopenia and recurrent mid-trimester foetal miscarriage.[1,2] High titre IgG-aCL antibody carries the greatest thrombosis risk, however, many patients with aPL antibodies are asymptomatic.[3,4] Cardiac abnormalities described in association with aPL antibodies include valvular abnormalities, pericardial effusion, myocardial dysfunction and coronary artery occlusion.[1,2] Of these, valvular abnormalities appear to be the most common finding, ranging from 11.6 to 32.0%.[2,3,5]

Intracardiac thrombosis is a potentially life threatening but treatable manifestation of APS. Thrombus formation has been described in all cardiac chambers and can cause pulmonary and systemic embolic events. Excisional biopsy is often required to differentiate thrombus from intracardiac myxoma. Management usually requires surgery and lifelong anticoagulation. APS-associated thrombocytopenia requires caution with anticoagulation.[1] Corticosteroid and immunosuppressive therapy should be considered in underlying connective tissue disease, thrombocytopenia and recurrent intracardiac thrombosis. The presentation and management of 19 previous case reports of intracardiac thrombi in APS have been summarised in Table 1. Key findings included moderate thrombocytopenia ($<75 \times 10^9$/L) in half of the cases, underlying structural cardiac abnormalities in 20%, and multiple intracardiac thrombi in 10%. Thirty per cent of patients were successfully managed without excisional biopsy.

Although it is not common, right heart thrombus should be considered in patients with APS presenting with pulmonary embolism. *Trans*-oesophageal echocardiogram may be required for diagnosis since thrombi arising from the right atrial appendage are poorly visualised on *trans*-thoracic echocardiography.

Treatment of intracardiac thrombosis complicating APS requires a careful balance between anticoagulation and bleeding risk. Complete resolution of thrombi with anticoagulation alone is reported, but excisional biopsy is usually required to exclude the diagnosis of cardiac myxoma and to remove the thrombus. Following initial treatment, lifelong prophylactic anticoagulation would seem prudent given the risk of recurrence for APS patients who cease warfarin.[2,6] International normalised ratios should be maintained above 2 and possibly higher for arterial thrombosis or recurrent disease.[2]

Table 1 Clinical features and management of patients with antiphospholipid syndrome and intracardiac thrombi

Age (sex)	Heart chamber	Association	Presentation	aCL	LAC	Treatment	Outcome	Reference
28F	RA	Nil	Asymptomatic	+	+	S+AC	NA	Granel *et al.*[6]
32F	RA	DVT	Asymptomatic	IgG	+	S+AC	Resolved	Leventhal *et al.*[7]
17F	RA	APS	PE	IgM, IgG	+	S+AC	Resolved	Gertner & Leatherman[8]
36F	RA	APS	PE	IgM, IgG	+	S+AC	Recurrence	Gertner & Leatherman[8]
49M	RA	APS	Asymptomatic	+	NA	AC	NA	Amato *et al.*[9]
58F	RA	APS	CCF	NA	+	S+AC	NA	Tamura *et al.*[10]
40F	RA	SLE	PE	IgG	+	S+AC	Resolved	†
36F	RA	SLE	PE	IgG	+	S+AC	Resolved	Gertner & Leatherman[8]
NA	RA	SLE	SE	+	+	AC+IS	NA	Ramdane *et al.*[11]
19F	RA	SLE	DVT	IgG	+	S+AC	NA	Ghirarduzzi *et al.*[12]
50F	RA	cutaneous SLE	NA	+	+	S	NA	Kjernsmo[13]
18M	RV	Nil	PE	IgG	–	S+AC+IS	Recurrence	O'Hickey *et al.*[14]
34F	RV	APS	Asymptomatic	IgG	+	S+AC	NA	Coppock *et al.*[15]
26F	LA	SBE/MVR	CCF	NA	+	S+AC	Died	Lubbe & Asherson [16]
58F	LV	Nil	SE	IgG	+	S+AC	NA	Baum & Jundt [17]
19F	LV	APS	SE	IgM, IgG	+	S+AC	Resolved	Aguilar & Summerson [18]
41F	LV	APS/TTP	SE	+	+	AC	Resolved	Bruce *et al.*[19]
38F	LV	SLE	SE	IgM, IgG	+	AC	NA	Kaplan *et al.*[1]
25F	LV	SLE	SE	NA	NA	S+AC	Died	Gorelick *et al.*[20]
16F	LV	SLE	SE, CCF	NA	+	AC+IS	Died	Gur *et al.*[21]

AC, anticoagulation; aCL, anticardiolipin; APS, antiphospholipid syndrome; CCF, congestive cardiac failure; DVT, deep vein thrombosis; F, female; IS, immunosuppression; LA, left atrium; LAC, lupus anticoagulant; LV, left ventricle; M, male; MVR, mitral valve replacement; NA, not available; PE, pulmonary embolus; RA, right atrium; RV right ventricle; S, surgery; SBE, subacute bacterial endocarditis; SE, systemic embolus; TTP, thrombocytopenic thrombotic purpura; +, positive; –, negative. †Patient described in the present paper.

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