Disseminated intracardiac thrombosis: a rare manifestation of antiphospholipid syndrome

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A 39-year-old severely obese male (body mass index 39 kg/m²) patient was referred to our department with suspected endocarditis. He presented with dyspnoea and recurrent fever of unknown origin. Infection parameters were found to be positive (C-reactive protein 24 mg/dL, reference <0.5 mg/dL; white blood cells 12 900/μL) and platelet count was low (16 000/μL). Other blood tests including troponin and blood cultures were normal. Transthoracic echocardiography showed a complex and irregular mass adherent to the endocardial surface of the right outflow tract and smaller floating structures of the right atrium (RA) and right ventricle (RV). Transoesophageal echocardiogram certified three masses: one was connected with the RA septum near the vena cava superior (Panel A), hyperechogenic and extending to the RV in diastole; another one was attached to the free RV wall; and the third one was adherent to the wall of the RV outflow tract and intermittently prolapsed through the pulmonary valve. A cardiac computed tomography confirmed the pseudotumours with partial calcification and the absence of contrast enhancement (Panel B). Hereditary thrombophilia and heparin-induced thrombocytopenia as well as rheumatological and serological testings were negative. A coronary angiography was performed due to intermittent chest pain and in preparation for a planned surgical treatment. The diagnostic procedure ruled out coronary heart disease and verified normal systolic left ventricular function. Cardiothoracic surgery was then performed due to the obstruction of RV outflow tract and to prevent further embolic complications. Surgical resection confirmed the thrombotic material with focal regions of calcification (Panel C). The diagnosis of antiphospholipid syndrome was confirmed by an anticardiolipin antibody test. Despite of continuous effective anticoagulation therapy, the patient died 2 weeks after surgical resection. An autopsy demonstrated disseminated intracardial thrombosis in all four chambers that led to multiple organ infarctions and vascular occlusive events (Panel D).

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.

Panel A. Transoesophageal echocardiography showing the right (RA) and left atrium (LA). Intracardiac mass with partial calcification was detected in the RA.

Panel B. Cardiac multislice computed tomography demonstrating intra-atrial mass (arrowheads) with partial calcification and lack of contrast enhancement.

Panel C. Resected right atrial thrombus.

Panel D. Disseminated intracardiac thrombosis (asterisk) with extensive intracardiac thrombosis in the right atrium (RA) and right ventricular (RV) apex and destruction of the tricuspid valve with disseminated thrombosis.