

An unusual and under-recognised cause of myocardial infarction

Bo Xu and John Hounsell

Clinical record

A 57-year-old woman underwent an uncomplicated vaginal hysterectomy for vaginal prolapse. Three days later, she re-presented with ischaemic chest pain associated with inferior ST-segment elevation. Thrombolysis was not initiated, due to her recent operation. The patient was transferred to a tertiary hospital for an urgent coronary angiogram, which showed normal, smooth coronary arteries and a thrombus in the right posterolateral branch. The patient was conservatively managed. Investigations for a paradoxical embolus were unrevealing: the results of a bilateral lower limb Doppler ultrasound examination were negative, and a transthoracic echocardiogram showed no evidence of a patent foramen ovale. The patient was discharged home. Three days after discharge, she presented to our hospital with an acute, painless loss of vision in the superior medial quadrant of her right eye. An urgent ophthalmological consultation led to a diagnosis of retinal artery thrombosis. A bilateral carotid Doppler ultrasound result was unremarkable. This second thrombotic event within a short time prompted an examination for underlying thrombophilia. A screening test result for antiphospholipid antibodies was strongly positive: anticardiolipin IgG antibody, 143 units (reference range [RR], < 20 units); β_2 -glycoprotein IgG antibody, 185 units (RR, < 20 units); lupus anticoagulant, 1.5 (RR, < 1.3). Results of tests for factor V Leiden (R506Q) and prothrombin gene (20210) mutations and antithrombin

III, protein C and protein S deficiencies were negative. On further questioning, our patient revealed that she had experienced intermittent visual blurriness in the right eye since 1992. She had previously been treated for a lupus-like condition with hydroxychloroquine. She had Raynaud's phenomenon affecting her fingers, and chronic shoulder and limb pain. Interestingly, the patient also had Dupuytren's contracture bilaterally in her hands and feet; this may have been part of her connective tissue disorder. While she was being treated, the patient's visual symptoms resolved. Her hydroxychloroquine treatment had been discontinued several years previously because her test results for serum antinuclear antibody (ANA) became negative. A screen for autoimmune disease at our hospital showed a strongly positive ANA titre of 1/1280 (RR, < 1/160) with a finely speckled homogeneous pattern, consistent with a lupus-like connective tissue disorder. Additional screening test results for autoimmune disease (anti-Ro, anti-La, anti-RNP, anti-Sm, anti-Scl-70, anti-Jo 1, anti-PCNA, antiribosomal P, anti-PM-Scl, and antismooth muscle) were negative. To prevent recurrent thrombotic events, warfarin treatment was commenced, aiming to achieve a target international normalised ratio of 2.5–3.5, and therapeutic enoxaparin was administered during the warfarin titration period. Hydroxychloroquine treatment was also begun, at a dose of 200 mg orally twice a day. ♦

We hypothesise that our patient had longstanding untreated connective tissue disease associated with antiphospholipid syndrome (APS). Her previous intermittent visual blurriness may have been caused by microthrombotic events. Her recent hysterectomy was the only major operation our patient had undergone (she had previously had a repair of Dupuytren's contracture in her right foot). The recent surgery and associated release of inflammatory mediators may have been the trigger that caused endothelial activation in her coronary and retinal arteries, causing myocardial infarction and visual loss, respectively. APS can have multiorgan manifestations, and is commonly associated with a connective tissue disorder such as systemic lupus erythematosus (SLE).¹ Unrecognised APS can have life-threatening consequences. It is a rare cause of myocardial infarction and, conversely, myocardial infarction is not an uncommon presentation of APS. In a cohort of 1000 patients with APS, 2.8% first presented with myocardial infarction.² Recurrent coronary

arterial thromboses have also been reported in patients with primary APS.^{3,4} Myocardial infarction due to APS is often undiagnosed initially, because the association between myocardial infarction and APS is under-recognised. Our case illustrates the importance of screening patients with SLE for APS, and highlights the importance of recognising and treating APS in patients with unusual recurrent thrombotic events.

The European League Against Rheumatism (EULAR) recommends screening patients with SLE at baseline for antiphospholipid (aPL) antibodies, and to repeat screening in previously negative patients when there are new risk factors for thromboembolism, such as pregnancy, surgery, transplantation and use of oestrogen-containing treatments.⁵ The other indications for screening patients for aPL antibodies are recurrent venous and/or arterial thromboses, especially in patients younger than 50 years, recurrent miscarriages or fetal loss and early or severe pre-eclampsia.¹ SLE and a positive test result for aPL antibodies, especially persistent anticardiolipin antibodies and lupus anticoagulant, increase the risk of thrombosis.¹ Data on primary prevention in asymptomatic patients are limited. EULAR recommends consideration of low-dose aspirin in patients with SLE for primary prevention of thrombosis and pregnancy loss.⁶ However, asymptomatic individuals testing positive for aPL antibodies do not appear to benefit from aspirin.⁷ For patients with SLE or a lupus-like connective tissue disease who test positive for aPL antibodies, we recommend making a case-by-case decision on primary prevention, based on an assessment of each patient's thromboembolic risk profile.

Lessons from practice

- Myocardial infarction associated with antiphospholipid syndrome is under-recognised.
- Antiphospholipid syndrome should be suspected in patients with unexplained, recurrent thrombotic events.
- Patients with systemic lupus erythematosus should be screened for antiphospholipid antibodies at baseline, with repeat screening when new thromboembolic risk factors arise.
- Low-dose aspirin should be considered in patients with systemic lupus erythematosus who test positive for antiphospholipid antibodies, as primary prevention of thrombosis and pregnancy loss. ♦

Competing interests

None identified.

Author details

Bo Xu, MB BS(Hons), Medical Registrar

John Hounsell, FRACP, Haematologist and Physician

Warrnambool Base Hospital, Warrnambool, VIC.

Correspondence: bo.xu@svhm.org.au

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