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Case Report

Paradoxical coronary embolism in Erdheim-Chester disease: invasive assessment and multidisciplinary management

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Abstract

We report a case of non-ST elevation myocardial infarction in a 36-year-old man with Erdheim-Chester disease (ECD). Multimodality assessment revealed acute coronary thrombus with simultaneous recurrent pulmonary embolism in spite of compliance with a direct oral anticoagulant. Prior case reports of acute myocardial infarction in this population have not outlined the role of catheter based intravascular assessment and treatment in this rare clinical entity.

Keywords: coronary embolism; Erdheim-Chester disease; non-ST elevation myocardial infarction

Introduction

Erdheim-Chester Disease is a rare non-Langerhans cell histiocytosis with multi-system involvement, characterised by production and accumulation of CD68(+), CD1a(−) non-Langerhans foamy histiocytes [1]. Fewer than 1000 cases are described. Cardiovascular involvement occurs in nearly half of ECD patients [2].

Our case shows intracoronary imaging plays a key role, guiding the management via a multidisciplinary approach when ECD presents with myocardial infarction.

Case report

A 36-year-old white British male presented to the emergency department with a three-weeks of progressively worsening exertional central chest pain, associated with breathlessness. Examination was unremarkable.

He was diagnosed with ECD ten years previously which had been controlled with pegylated interferon. Nine months earlier he acquired COVID-19, which was complicated by deep venous thrombosis and subsequently pulmonary embolism (PE). Direct oral anticoagulant (DOAC), Apixaban 5 mg twice daily, was started. Subsequent investigations revealed ECD no longer in remission. Interferon dosing was increased with an additional course of prednisolone. He has no classic risk factors for atherosclerosis.

Admission electrocardiogram (ECG) showed sinus rhythm with <1 mm convex ST segment elevation V1-V6, with T-wave inversion in I and aVL. Presenting high-sensitivity troponin T was 51 ng/L (0–14 ng/L) rising to 132 ng/L 6 h later. His D-dimer was raised at 608 ng/mL (0–500 ng/mL).

The differential diagnosis on presentation included acute myocardial infarction, myocarditis/pericarditis and recurrent PE.

Given the possible diagnoses, transthoracic echocardiography (TTE) and a contrast enhanced gated CT thorax to assess the coronary arteries (CTCA), with acquisition extended to capture pulmonary vasculature, was arranged. TTE was unremarkable. Although the coronary calcium score was 0, an ostial left anterior descending (LAD) artery non-calcified plaque 1.5 cm in length, with high-risk plaque features of positive remodelling and a low-density lipid rich core, caused severe stenosis (Fig. 1). A small pulmonary infarct suggestive of acute pulmonary embolism was also seen.

Ahead of invasive coronary angiography Apixaban was discontinued, and therapeutic subcutaneous low-molecular weight heparin (LMWH) initiated alongside dual antiplatelet therapy (aspirin 75 mg, clopidogrel 75 mg). This confirmed an ostial filling defect in the LAD with distal TIMI 3 flow (Fig. 2). Left main stem, circumflex, and non-dominant right coronary artery were normal.

In the context of prior, and now recurrent, central venous thromboembolism due to an underlying pro-thrombotic condition, in the absence of atherosclerotic risk factors, young age and a CT calcium score of 0, the presentation was felt consistent with coronary embolism. Distal flow was preserved, and in the absence of ongoing symptoms, further assessment by intracoronary imaging was deferred due to risk of clot embolization. Following multi-disciplinary discussion with specialist haematologist input a bolus and infusion of glycoprotein IIb/IIIa (Gp2b/3a) inhibitor Tirofiban was administered until repeat angiography 72 h later. The LMWH was reduced to a prophylactic dose to limit bleeding risk while on quadruple therapy.

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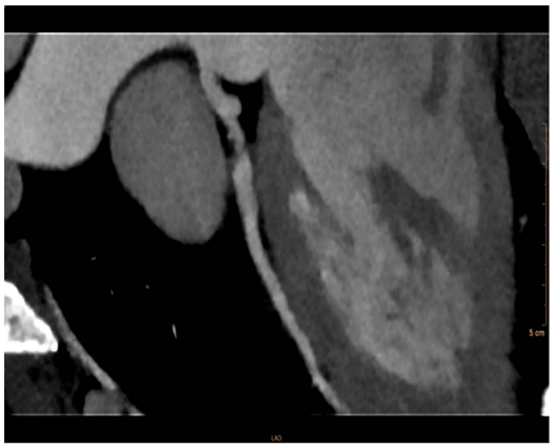

Figure 1. Computerized tomography (CT) coronary angiogram. 15 mm non-calcified lesion from ostial to proximal left anterior descending (LAD) coronary artery causing moderate to severe stenosis.

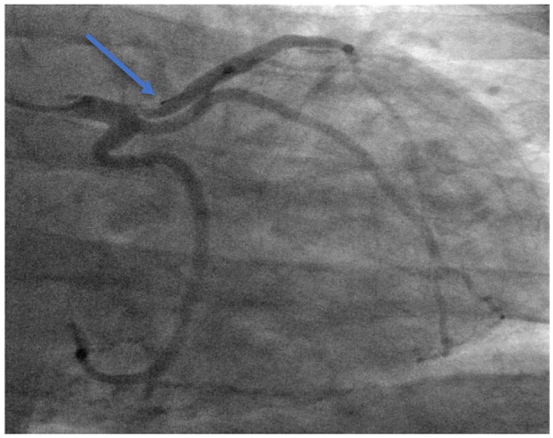

Figure 2. Initial angiogram. Ostial filling defect in the left anterior descending (LAD) coronary artery (arrow).

Repeat invasive assessment of the LAD angiography showed improvement in luminal calibre enabling intracoronary imaging (Fig. 3). Optical coherence tomography (OCT) showed features of residual thrombus and fibroatherosclerotic plaque, however images were suboptimal due to poor clearance of blood in a large calibre vessel. Intravascular ultrasound (IVUS) was therefore performed which confirmed the OCT findings (Fig. 4). Intra-procedurally the patient developed chest pain. Angiography showed worsening ostial LAD stenosis. Balloon dilatation was performed with a 3.5 mm non-compliant balloon to relieve pain and allow time for decision making.

Direct stenting was discounted due to the large calibre LAD, and the requirement to stent into the left main stem across both a large ramus branch and the left circumflex artery. A 4.0 × 20 mm Sequent drug-eluting balloon was inflated, following which symptoms resolved. Recurrent thromboembolic disease whilst taking DOAC triggered a switch to a vitamin K antagonist with a target INR of 2.5. As the presentation was consistent with paradoxical embolization a bubble study was arranged. This suggested presence of a patent foramen ovale (PFO). There is no guideline for this situation, however the multidisciplinary meeting (MDT) felt offering closure of the PFO was preferable given the propensity to recurrent venous thromboembolism with presumed systemic embolization despite anticoagulation therapy.

Six weeks post discharge combined PFO closure and repeat LAD assessment with intravascular imaging occured. PFO closure was performed with a 25 mm Amplatzer PFO occluder using an

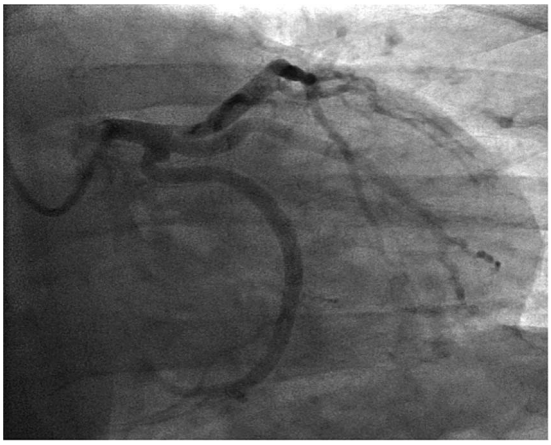

Figure 3. Repeat angiogram. Improvement to previous ostial left anterior descending (LAD) coronary artery lesion.

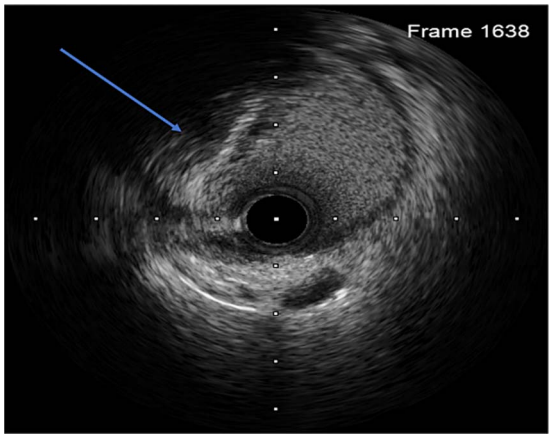

Figure 4. Intravascular ultrasound (IVUS). Features of thrombus (arrow).

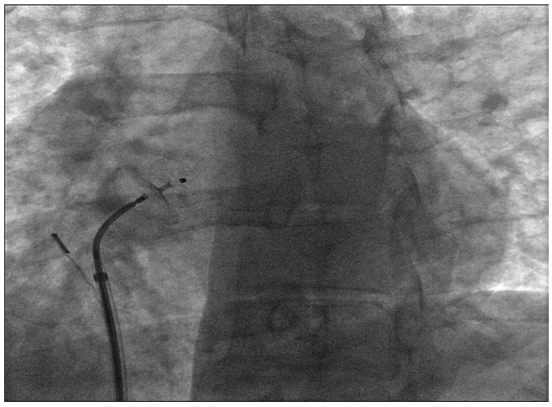

Figure 5. PFO closure. A 25 mm amplatzer PFO occluder using a 8-French Trevision delivery system was used.

8-French Trevisio delivery system under intracardiac echo guidance (Fig. 5). IVUS (Fig. 6) and OCT (Fig. 7) showed eccentric fibrotic plaque with minimal luminal area of 9 mm^2 in the proximal LAD. He was discharged on secondary prevention medication for myocardial infarction, lifelong VKA, aspirin for one month, and clopidogrel for 12 months.

Discussion

Cardiac involvement in Erdheim-Chester Disease can affect coronary blood vessels, myocardium and pericardium [3–5]. Coronary

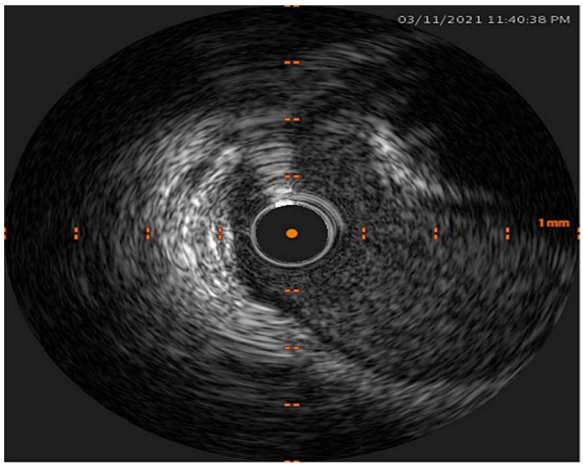

Figure 6. Repeat intravascular ultrasound (IVUS). Eccentric fatty/fibrotic plaque in proximal left anterior descending (LAD) coronary artery with mild calcification.

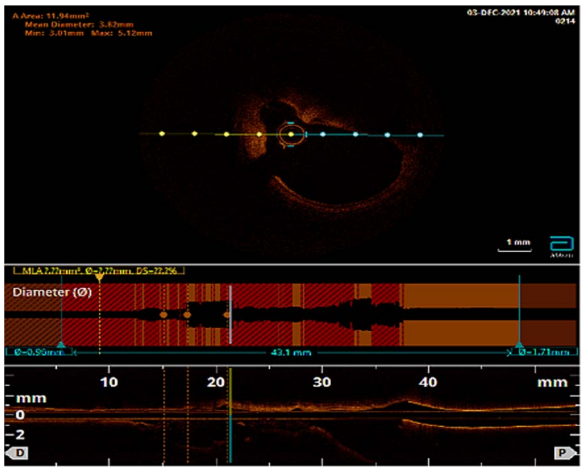

Figure 7. Repeat optical coherence tomography (OCT). Limited by dropout segment in proximal left anterior descending (LAD) coronary artery.

pathology can be extravascular (perivascular infiltration extending into the coronary sulci), vascular (abnormal sheathing of the aorta and coronary arteries), or intravascular due to the elevated rates of thrombus formation [6, 7].

Here both coronary thrombus and atherosclerotic plaque were present. Intravascular imaging allows characterisation, and prognostication, of ambiguous coronary lesions in patients with diagnostic uncertainty [8]. Although IVUS and OCT can assess fibrous plaque, OCT is superior in assessing thrombus [9]. Coronary appearance plus the context of a patent foramen ovale, simultaneous recurrent pulmonary embolism and a prothrombotic state favoured an embolic aetiology. Up to 3% of myocardial infarction may be caused by coronary embolism, however the incidence of paradoxical coronary embolism is unknown [10]. The role of percutaneous device closure to prevent recurrent paradoxical coronary embolism has not been studied. Offering closure in rare cases like this is a balance of judgement of risks and benefits that our MDT on this occasion felt that supported closure of the patent foramen ovale.

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Conflict of interest

No conflict of interest.

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Ethical approval

None required.

Consent

Written consent was obtained from the patient for this publication.

Guarantor

Dr Freidoon Keshavarzi is the guarantor for this case.

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