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BRIEF COMMUNICATION

Recurrent ST Elevation Myocardial Infarction: What is the Aetiology?

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Coronary artery disease is a leading cause of morbidity and mortality in the western world. Plaque rupture in an atherosclerotic lesion is the most commonly encountered underlying pathophysiology. Spontaneous coronary dissection can have similar presentation, but we as a community of cardiologists may not be aware of uncommon aetiologies, such as vasculitis presenting as ST elevation myocardial infarction (STEMI). Here we present a case report of a lady, who presented with STEMI on three occasions within five days, due to underlying granulomotosis with polyangiitis (GPA).

Keywords Wegener Granulomatosis • Vasculitis • Myocardial infarction • Coronary thrombosis

A 58-year-old lady was brought to the hospital with acute-onset chest pain, and electrocardiogram demonstrated inferior ST elevation myocardial infarction (STEMI) (Figure 1A). Coronary angiography (CAG) demonstrated normal coronaries, except occluded distal part of posterior descending artery (Figure 2, A-C) that was treated conservatively. She was not known to have any cardiac risk factors. Five days prior, she had been admitted to a nearby hospital with documented inferior STEMI, and CAG was reported to be normal. CT pulmonary angiography and echocardiogram ruled out pulmonary embolism and absence of flow across the inter-atrial septum. No arrhythmias were detected on telemetry and after three days of uncomplicated stay, the patient was discharged on dual antiplatelet therapy.

Two hours after catheterisation, the patient complained of chest pain and electrocardiogram demonstrated global ST elevation (Figure 1B). Within minutes, she was in cardiogenic shock and had recurrent ventricular fibrillation, requiring direct-current cardioversion and cardio-pulmonary resuscitation (CPR). The patient was intubated and angiography demonstrated occluded left main stem (LMS), which was wired, ballooned and stented to restore the flow, while undergoing ongoing CPR. Angiography of the right coronary artery demonstrated occluded distal part of the postero-lateral branch, which was a new finding (Figure 2, D-I).

Spontaneous coronary artery occlusion in different segments raised a strong suspicion of vasculitis, as it seemed very unlikely to be thrombo-embolic/atherosclerotic in nature. Inquiry with family revealed that the patient reported dry cough and unexplained weight loss in the preceeding two to three months, but no formal investigations were performed. Rheumatology consultation strongly raised the possibility of vasculitis as the underlying mechanism. The patient was treated with pulsed methylprednisolone, with planned plasmapharesis but unfortunately the patient died within 12 hours from her presentation.

Laboratory results demonstrated elevated level of cytoplasmic anti-neutrophil cytoplasmic antibody (C-ANCA) (>1:320); with strong positivity against proteinase-3 antigen, but negative for myeloperoxidase. Antibodies against nuclear, double stranded-DNA, cardiolipin and beta-2-Glycoprotein antigens were not detected. Antibodies to extractable nuclear antigens were negative. Immunoglobulin levels and serum electrophoresis were within normal limits.

Laboratory investigations and clinical features confirmed the diagnosis of granulomatosis with polyangiitis (GPA;

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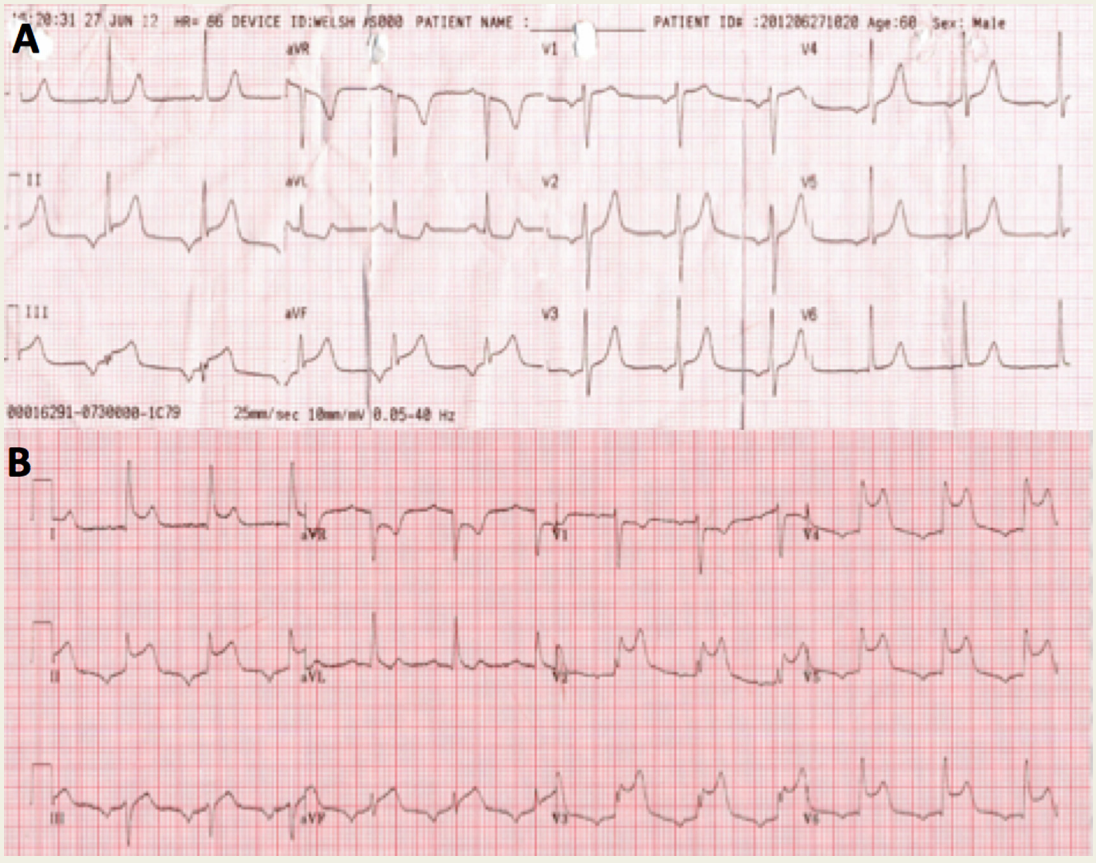

Figure 1 **Electrocardiogram**.
ECG at the time of initial presentation with inferior STEMI (A), and 2 hours after transfer to CCU demonstrating global ST elevation with ST depression in aVR (B). Note inverted 'P' wave in inferior lead, suggestive of lower atrial paced rhythm.

Wegener's granulomatosis). Granulomotosis with polyangiitis results in ANCA-associated vasculitis (AAV), affecting small and medium-sized vessels in the respiratory tract, kidney, skin and peripheral nerves. Although cardiac involvement is uncommon, cardiovascular mortality is more than three-fold higher in patients with GPA, than in the general population [1]. Coronary, myocardium, pericardium, valves and conduction system involvement have all been described [2]. The majority of published data is suggestive of silent myocardial infarction, found during routine investigations or post-mortem examination [3,4], whereas there are reports of patients presenting with MI and chest pain, within weeks of diagnosis of GPA [5]. Post-mortem examination in such cases has demonstrated diffuse coronary arteritis with thrombotic occlusion [6,7].

Our case is unique in that the patient presented with thrombotic occlusion of different coronary segments, and the diagnosis of GPA was made after presentation with recurrent STEMI. Additionally, electrocardiography demonstrated low-atrial-rhythm (inverted P-waves in inferior leads), suggestive of underlying sinus node dysfunction that had previously been reported in patients with GPA [8]. Dual anti-platelet therapy may not be adequately protective, as the underlying pathophysiological process was immune-related and non-atherosclerotic.

Pathophysiologically, primed neutrophils express myeloperoxidase, proteinase-3 and other antigens either on or near the cell surface. Anti-neutrophil cytoplasmic antibodies stimulate neutrophils by interacting with these antigens. Monocytes also express similar ANCA-related-antigens. Activated neutrophils, along with monocytes, interact with vascular endothelial cells and penetrate into the vessel-wall, where they release granule contents along with reactive-oxygen-radicals, form neutrophil extracellular traps containing pro-inflammatory proteins, and eventually lead to apoptosis and necrosis of cells. Simultaneous complement pathway activation amplifies the inflammatory milieu in the vessel wall by attracting more neutrophils. Neutrophil-rich segmental vascular necrosis and associated perivascular infiltration sets-in within hours. Subsequently, the presence of haemorrhage and coagulant proteins in the vessel wall results in fibrinoid degeneration [9].

Our case is very rare and unique, as it demonstrates thrombotic occlusion of multiple, angiographically normal coronary segments, over a short period of time, very likely due to ANCA-associated-vasculitis (Image-2;C,D,I). Patients