CASE REPORT

Acute ST elevation myocardial infarction in fulminant systemic p-ANCA vasculitis: a rare catastrophic complication

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SUMMARY

A 45-year-old Caucasian man presented to the hospital with a 3-month history of fatigue, bilateral upper and lower limb paresthesias and gradually worsening ascending paralysis. A few weeks later, he developed acute renal failure requiring haemodialysis. Investigations revealed presence of myeloperoxidase (MPO) perinuclear antineutrophil cytoplasmic antibodies (ANCA). Renal biopsy was conclusive for rapidly progressive glomerulonephritis with crescents. Treatment for ANCA positive vasculitis was initiated with pulsed steroids, cyclophosphamide and plasmapheresis. The hospital course took an unexpected turn when the patient developed acute chest pain with an EKG consistent with inferior ST elevation myocardial infarction (STEMI). Urgent left heart catheterisation revealed distal occlusions in multivessel coronary distribution. Coronary involvement is rare in ANCA vasculitis and STEMI has not been reported in MPO-ANCA positive vasculitis, to the best of our knowledge.

BACKGROUND

Antineutrophil cytoplasmic antibodies (ANCA) vasculitis is predominantly a small vessel disease and coronary involvement is rare. ST elevation myocardial infarction (STEMI) is a medical emergency and, to the best of our knowledge, has not been reported in myeloperoxidase (MPO) antibody-ANCA vasculitis. Most myocardial infarctions in patients with small vessel vasculitis reported in the literature have been silent, with coronary involvement being confirmed on imaging (cardiac MRI) or on postmortem examination.^{1–3}

CASE PRESENTATION

A 45-year-old Caucasian man was admitted to a tertiary care university teaching hospital, with a chief symptom of worsening fatigue and weakness in upper and lower extremities. He had been discharged from the neurology service 2 weeks prior with diagnosis of diffuse axonal demyelinating polyneuropathy in a glove and stocking distribution. This diagnosis was based on electromyogram and nerve conduction studies. These neurological symptoms had actually started 4 months earlier and had been progressively worsening. The aetiology of neuropathy in this patient remained unclear on initial work up.

On current admission, he also reported decreased appetite, subjective fevers, chills, night sweats, cough and brown coloured sputum

production intermittently for 2 weeks. He had tingling and numbness in his hands and feet, and left foot drop, symptoms that were unchanged since his earlier discharge. He denied headaches, blurry vision, chest pain, shortness of breath, palpitations, dizziness or abdominal symptoms. Vitals on admission were within normal limits.

On his prior admission, he had been noted to be positive for hepatitis C (viral load 248, 969 IU/mL). He had not received treatment for it. Liver ultrasound had revealed cirrhotic changes. He had undergone a splenectomy secondary to trauma some years earlier. He was an active smoker and had been smoking one pack per day for 30 years. He also reported heavy alcohol and illicit drug use (cocaine and marijuana) in the past. His family history was unknown to the patient.

On examination, he was lethargic but conversant. He appeared cachectic. There was significant proximal and distal muscle atrophy and profound weakness in upper and lower extremities: 3.5/5 power in bilateral upper extremity muscle groups and 1–2/5 in the lower extremities. The patient's deep tendon reflexes were diminished throughout. His lungs were clear on auscultation. He had an unremarkable cardiac examination. No peripheral oedema was noted, pulses were intact and no rash was seen.

Initial work up revealed leucocytosis of 21 000/mm³ with a left shift, normocytic anaemia and a creatine level of 3.3 (baseline 1.1). Urinalysis revealed leucocytes and numerous red blood cells but no casts. Renal ultrasound showed diffusely increased cortical echogenicity. Urine culture was negative and blood cultures drawn at weekly intervals twice showed no growth. The patient had a transoesophageal echocardiogram for persistent leucocytosis considering his history of drug abuse, which was unremarkable with no valvular vegetations.

Renal biopsy showed pauci-immune necrotising and crescentic glomerulonephritis (figure 1A, B). A rheumatological panel, ordered at the time of admission, came back positive for MPO antibodies with levels >8 Antibody Index using micro-bead technology (AI): >1.0 is considered positive and 8 is the highest reported value. Antinuclear antibodies (ANA) was negative. C3 and C4 titres were in normal range. Anti-Glomerular basement membrane antibody was negative. ANCA proteinase was <0.2 AI and rheumatoid factor <10 IU/mL.

Serum cryoglobulins were negative. A diagnosis of MPO-ANCA positive vasculitis was made.

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Figure 1 Renal biopsy (methenamine silver stain) showing necrosis (red arrow), crescent (white arrow).

Treatment was initiated with high-dose pulse steroid and plasmapheresis. Dialysis was initiated for worsening renal failure, oliguria and uraemic symptoms. Cyclophosphamide was added to the regimen.

On the fifth day of treatment, the patient awoke with sharp midsternal chest pain associated with shortness of breath. He became hypoxic with oxygen saturation of 84 percent on room air, and required high-flow oxygen therapy. Lung examination revealed diffuse bibasilar crackles. EKG had inferolateral ST elevations with new Q waves (figure 2). The patient was emergently

Figure 2 Twelve-lead ECG during episode of chest pain, showing ST segment elevations in leads II, III, aVF (inferior leads) and V5, V6 (lateral leads) with reciprocal changes in anterior leads.

Figure 3 Coronary angiogram of the left coronary system showing multiple distal coronary occlusions involving left anterior descending and left circumflex territories as depicted by arrows.

taken to the cardiac catheterisation laboratory. Coronary angiogram (figures 3 and 4 and videos 1 and 2) revealed multiple distal coronary occlusions involving apical left anterior descending (LAD), diagonal branches of LAD, obtuse marginal and atrial branches of left circumflex system, distal segment of the ramus intermedius artery, posterior descending artery and right ventricle marginal branches of the right coronary artery. This angiographic anatomy was not amenable to intervention due to multiple distal vessel occlusions with no clear target.

Medical management was planned with dual antiplatelet therapy, and heparin drip was initiated and continued for 48 hours. Antianginal therapy was added for symptom control. Troponins peaked at 110 ng/mL before trending down. A two-dimensional transthoracic echocardiogram post infarct showed severely depressed left ventricle function with ejection fraction of 20–25% with severe hypokinesis of all the anterior and inferior walls, with akinesis of the apex.

OUTCOME AND FOLLOW-UP

Following initial transient improvement with steroids, plasmapheresis and cyclophosphamide, the further hospital course was

Figure 4 Coronary angiogram of the right coronary artery showing occlusion of right ventricle marginal branch and distal posterior descending artery.

Video 2 Coronary angiogram of the right coronary artery showing occlusion of right ventricle marginal branch and distal posterior descending artery.

complicated by acute respiratory failure with pulmonary oedema requiring invasive mechanical ventilation; hospital acquired pneumonia; sepsis treated with broad spectrum antibiotics; bowel ischaemia and pulmonary haemorrhage. Sural nerve biopsy, performed in the initial days of hospitalisation, revealed evidence of early vasculitis. The patient was started on rituximab, and continued on steroids and antibiotics, with minimal improvement in his symptoms. His clinical status continued to decline despite optimal therapy. He expired after 44 days of inpatient management.

DISCUSSION

Coronary involvement is very rare in ANCA vasculitis. Cardiac manifestations of ANCA vasculitis include cardiomyopathy,

Video 1 Coronary angiogram of the left coronary system showing multiple distal coronary occlusions involving left anterior descending and left circumflex territories as depicted by arrows.

pericarditis, coronary dissection, valvulitis, arrhythmias and aneurysm formation. Coronary arteritis is an uncommon phenomenon in ANCA vasculitis. In one of the largest case studies on Wegener's vasculitis,⁴ 158 patients were followed for 8 years; 10 patients (6%) had detectable heart involvement and all had specific pericarditis. No myocardial ischaemia was reported. This highlights the rarity of the condition. Coronary vessels are medium sized and their involvement is uncommon in small vessel vasculitis. To the best of our knowledge, this is the first reported case of STEMI in a patient with fulminant anti-MPO+ANCA vasculitis. One case of STEMI was reported by Lazarus *et al*⁵ in a patient with Wegener's (cytoplasmic-ANCA (c-ANCA) positive vasculitis) where coronary involvement was the initial presentation of the disease. Another case was reported in the literature in a woman who had recurrent STEMI, again, in the setting of Wegener's disease.⁶ Our patient had been diagnosed with rapidly progressive glomerulonephritis and perinuclear antineutrophil cytoplasmic antibodies vasculitis, and at the time of STEMI was on immunosuppressive therapy. Our case is different from the two reported cases as our patient did not have c-ANCA vasculitis/Wegener's disease. He was positive for MPO-ANCA, which favours the diagnosis of microscopic polyangiitis in the setting of negative glomerular basement membrane antibodies, proteinase-3 Ab, ANA and normal complement levels. Serum cryoglobulins were undetectable in this hepatitis C positive patient, ruling out another important cause of fulminant vasculitis. There was no evidence of sepsis prior to this episode of myocardial infarction, as evident from the physical examination, blood and urine culture results and transoesophageal echocardiogram.

Cardiac involvement in microscopic polyangiitis was described in a case report from Japan, where the autopsy findings revealed no coronary obstruction but, rather, scattered infarcts in the left ventricular wall and interventricular septum.⁷ A case of coronary angiitis and cardiac arrest but no ST elevations was recently published by Shah *et al*,⁸ again in c-ANCA-associated vasculitis. Coronary angiogram in that case revealed normal major epicardial vessels with no obstructive

atheroma. Pruning in the first, second marginal and first diagonal branches was noted on angiogram.

Ours is a unique case of fulminant anti-MPO positive ANCA vasculitis complicated by acute ST elevation myocardial infarction. Multiple distal coronary arteries were found to be occluded on angiogram and were unamenable to intervention. This pattern of scattered occlusions on coronary angiogram does not favour the classic setting of plaque rupture and atherosclerotic cardiovascular disease, but is highly indicative of a vasculitic phenomenon. Unfortunately, our patient succumbed to

this fulminant systemic vasculitis despite aggressive medical treatment. However, this case highlights the importance of prompt diagnosis and treatment of ANCA vasculitis, and recognising coronary ischaemia as a potential serious complication.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Small vessel antineutrophil cytoplasmic antibodies (ANCA) vasculitis can rarely involve the coronary arteries, which are medium-sized vessels.
- ▶ Acute ST elevation myocardial infarction is an extremely rare but potentially life-threatening complication of ANCA vasculitis; this is the first reported case in the setting of antimyeloperoxidase positive ANCA vasculitis.
- ▶ This case highlights the importance of prompt diagnosis and treatment of ANCA vasculitis, and recognising coronary ischaemia as a potential serious complication.

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