

An unusual precipitant of acute heart failure—ANCA-associated vasculitis in a patient with ischaemic cardiomyopathy: a case report

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Background

Antineutrophil cytoplasmic antibody (ANCA)-associated pulmonary renal vasculitis is an uncommon disease entity. Its presentation as acute heart failure for the first time in a patient with established coronary artery disease (CAD) is even rarer. We present here a case of such an association and an approach to managing this clinical situation.

Case summary

A 60-year-old male patient presented to the emergency room with recent-onset dyspnoea New York Heart Association Class IV. He was having hypertension, uncontrolled diabetes mellitus, chronic kidney disease (CKD), and CAD. He also underwent a percutaneous coronary intervention to left anterior descending in the past for acute coronary syndrome and had moderate left ventricular dysfunction. He was being managed as a case of acute decompensated heart failure (ADHF) and was mechanically ventilated. Suddenly his ventilator requirement increased and endotracheal aspirate contained blood. The chest radiograph showed bilateral hilar infiltrates. Simultaneously he also had recurrent episodes of ventricular tachycardia (VT) requiring direct current (DC) cardioversion. Blood investigations showed deranged renal function and severe hyperkalaemia, but no evidence of coagulopathy. High-resolution computed tomography chest showed features of diffuse alveolar haemorrhage. Further investigations revealed high titres of c-ANCA and raised inflammatory biomarkers. A diagnosis of ANCA-associated vasculitis presenting as acute on CKD with dyselectrolytaemia (hyperkalaemia) leading to VT was made. Apart from standard management for associated illness, he was treated with plasma exchange, steroids, and cyclophosphamide to which he responded and was later on discharged.

Discussion

Antineutrophil cytoplasmic antibody-related pulmonary renal vasculitis can lead to rapidly progressing renal failure and may present as ADHF in a patient with existent CAD. The associated VT storm in our patient can be attributed to hyperkalaemia secondary to acute renal failure. A multidisciplinary approach is required for the successful management of such a complex clinical scenario.

Keywords

Antineutrophil cytoplasmic antibodies-associated vasculitis • Coronary artery disease • Renal failure • Diffuse alveolar haemorrhage • Ventricular tachycardia storm • Case report

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Learning points

- Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis is a rare diagnosis which requires a high index of suspicion and can precipitate heart failure in patient with underlying heart disease.
- Hyperkalaemia in combination with hypoxaemia and acidosis can precipitate ventricular tachycardia in a previously scarred myocardium.
- A multidisciplinary team approach is essential for successful management of patients with ANCA vasculitis and heart failure.

Introduction

Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis is a rare disease entity¹ and its occurrence for the first time in a patient with established coronary artery disease (CAD) and presenting as acute heart failure is even rarer. A high index of suspicion is needed for making such a diagnosis and every other possibility needs to be ruled out before embarking upon such an association.² This is because the treatment of such aggressive vasculitis involves aggressive immunosuppression and may have a serious implication on existent CAD status.³ Here we report a case of ANCA-associated pulmonary renal vasculitis presenting for the first time as acute decompensated heart failure (ADHF) complicated by ventricular tachycardia (VT) storm in a patient with CAD and an approach to management in such cases.

Timeline

2000	Anterior wall myocardial infarction Percutaneous angioplasty to left anterior descending coronary artery done Left ventricular ejection fraction: 40% Creatinine: 1.2 mg/dL
2018	Hospital admission for heart failure with acute kidney injury
Day 1	Hyperkalaemia and multiple episodes of haemodynamically unstable ventricular tachycardia. Direct current cardioversion, amiodarone infusion, and sustained low-efficiency dialysis given
Day 2	Blood in endotracheal tube and haemoglobin drop; diffuse alveolar haemorrhage confirmed (DAH)
Day 3	Antineutrophil cytoplasmic antibody (ANCA) by indirect immunofluorescence and ELISA—positive and diagnosed as ANCA-associated vasculitis, high-resolution computed tomography chest suggestive of DAH
Day 4	Plasma exchange therapy and immunosuppression under cover of antibiotics started
Day 10	Patient improved and discharged home. Counselling for automated implantable cardioverter-defibrillator insertion, refused by attendants
Day 11	Cyclophosphamide pulse therapy continued for 6 months. Patient remained dialysis dependent
9 months	Patient had a sudden cardiac arrest during renal replacement therapy

Case presentation

A 60-year-old male presented to the emergency room (ER) of our hospital with recent-onset New York Heart Association (NYHA) Class IV dyspnoea. He had hypertension (HT) and uncontrolled diabetes mellitus (DM) for the past 20 years. He also had an acute coronary event 18 years ago for which he underwent coronary angiography and stenting was performed to left anterior descending (LAD) artery. His last documented left ventricular ejection fraction (LVEF) before this event was 35–40% with severe hypokinesia involving the LAD territory. He was on aspirin, statin, and beta-blocker for CAD; oral hypoglycaemic drugs, and insulin for DM and angiotensin-converting enzyme inhibitor for HT. His clinical status deteriorated rapidly from NYHA II to IV in the last 4 days before presentation at ER. On examination, he had a heart rate of 130/min, blood pressure of 170/90 mmHg, respiratory rate of 30/min, systemic oxygen saturation of 70% on room air, and raised jugular venous pressure. Auscultation revealed fine basal crepitation and left ventricular (LV) third heart sound. Echocardiogram showed an LVEF of 35% with regional wall motion abnormality in LAD territory. N-terminal pro-hormone of brain natriuretic peptide levels were 8923 pg/mL. With the above history, he was initially managed as ADHF with non-invasive ventilation, diuretics, and vasodilators.

Electrocardiogram (ECG) showed sinus tachycardia, Q waves in anterior leads (*Figure 1*). However, no new ST-T changes were noticed in comparison with his previous ECG. Serial cardiac biomarkers (Troponin I and CK MB) were in the normal range. Blood investigations showed anaemia (haemoglobin of 10.4 g/dL), hyperkalaemia (6.9 mEq/L), deranged renal function (serum creatinine—6.7 mg/dL and blood urea nitrogen—46 mg/dL), and poor glycaemic control (HbA1c of 8.9%). Arterial blood gases analysis showed hypoxaemia (pO₂ of 60%) and metabolic acidosis (pH of 7.22). Urine analysis showed active sediments (3–5 red blood cells/high power field) and proteinuria (24-h urine protein of 1.4 g/day). A chest radiograph showed pulmonary oedema with bilateral pleural effusion (*Figure 2*).

He was managed with anti-hyperkalaemic measures (insulin-dextrose infusion, salbutamol nebulization, oral potassium binders). Despite the above measures, his hyperkalaemia was persistent. Later in the course, he had multiple episodes of haemodynamically unstable monomorphic VT (*Figure 3*) in <3 h duration requiring electrical cardioversion supplemented with intravenous amiodarone. He was mechanically ventilated due to worsening respiratory symptoms and hypotension. Sustained low-efficiency dialysis was also performed for 8 h on Day 1.

On Day 2 of admission, he had increasing requirements of oxygen (FiO₂) and endotracheal tube suction showed bloodstained secretions. There was also a haemoglobin drop of ~5 g/dL (10.4–5.4 g/dL).

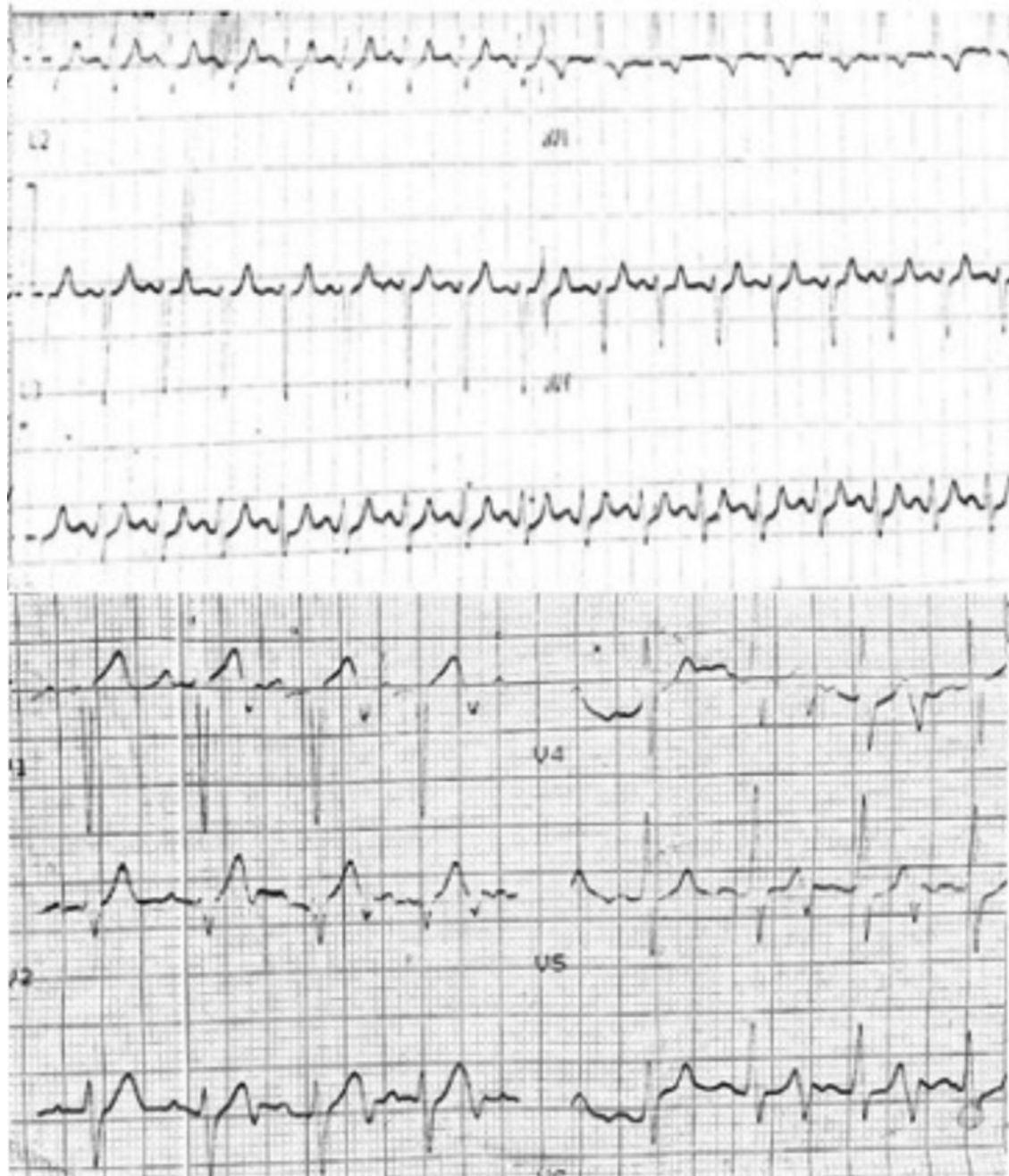


Figure 1 Baseline electrocardiogram showing normal sinus rhythm with qs pattern in leads V1 and V2 with left axis deviation.

High-resolution computed tomography was performed to find the cause of bleeding, which showed diffuse bilateral perihilar consolidation and areas of ground-glass opacities predominantly involving the dependent lobes suggestive of diffuse alveolar haemorrhage (DAH) (Figure 4). Taking into consideration a new onset of renal dysfunction and DAH, a possibility of active vasculitis was kept and an immunology consultation was taken.

Subsequent investigations showed strongly positive c-ANCA (ELISA and indirect immunofluorescence). Antinuclear antibodies and anti-glomerular basement membrane antibodies were negative.

Based on the above results, a diagnosis of ANCA-associated pulmonary renal vasculitis precipitating acute heart failure was made. Refractory hyperkalaemia and severe metabolic acidosis (secondary to acute kidney injury) combined with hypoxaemia and heart failure, precipitated the VT storm in an already scarred myocardium. In addition to the treatment of heart failure, he was started on immunosuppression (methylprednisolone pulse therapy—1 g daily for 3 days) and seven sessions of plasma exchange.

With the above measures, the patient improved significantly and could be extubated 3 days later. Renal biopsy was initially deferred as

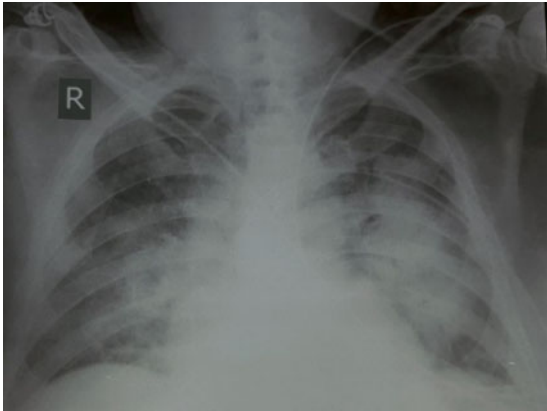


Figure 2 Chest X-ray showing bilateral diffuse infiltrates predominantly perihilar in location.

he was on dual antiplatelets (clopidogrel was added at admission) and later on because of anticipated technical difficulties (the patient being obese). He was also started on intravenous cyclophosphamide (10 mg/kg/dose), initially every 2 weeks for the first three doses followed by every 3 weeks for the next 6 months.

At 6 months follow-up, he was doing well from a cardiac point of view with LVEF of 40% and NYHA II functional status. However, his

renal function deteriorated further requiring renal replacement therapy (RRT). He was also counselled about the need for an intracardiac defibrillator for the prevention of sudden cardiac death which he refused because of financial constraints. A telephonic follow-up performed at 9 months revealed that he had a sudden cardiac arrest and died during a session of dialysis.

Discussion

The spectrum of ANCA-associated vasculitis includes granulomatosis with polyangiitis (GPA), microscopic polyangiitis, and eosinophilic GPA (Churg-Strauss syndrome).¹ It is an autoimmune disorder affecting small and medium-sized vessels. It usually presents as fatigue, fever, weight loss, arthralgias, rhinosinusitis, cough, dyspnoea, urinary abnormalities, purpura, and neurologic dysfunction.² Cardiac manifestations in ANCA-associated vasculitis are known to occur in ~3.3–90% in various cohorts depending on the method of evaluation and diagnostic tests applied.^{3–5} Pericarditis is the most common manifestation (35%), followed by cardiomyopathy (30%), CAD (12%), valvular disease (6%), concomitant CAD and valvular disease (6%), concomitant pericarditis and cardiomyopathy (1.6%), and severe conduction disorders (1.6%).⁴ Ventricular arrhythmias and particularly VT storm due to active vasculitis are rarely observed in ANCA-associated vasculitis. It has been reported in only two cases in the past and both patients had histopathological evidence of

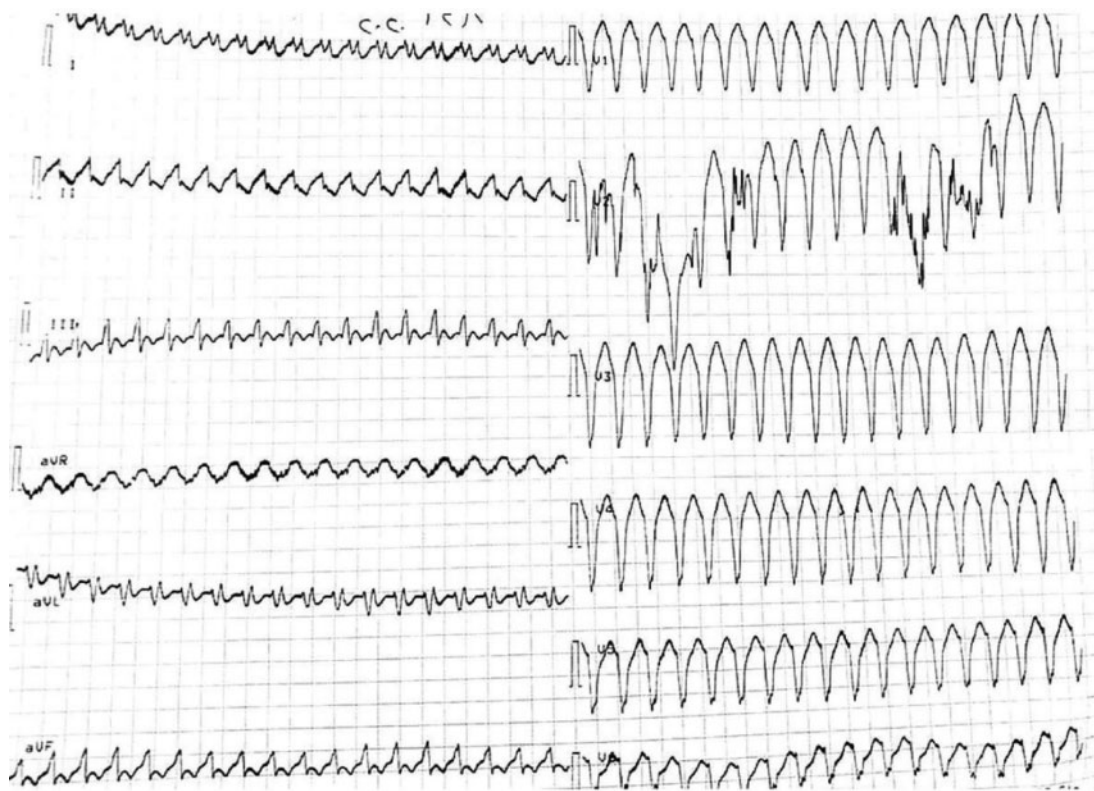


Figure 3 Electrocardiogram showing monomorphic ventricular tachycardia.

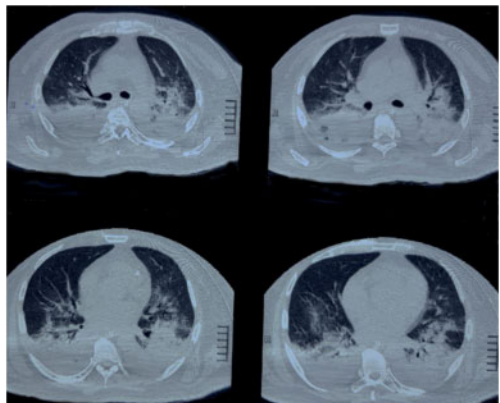


Figure 4 Computed tomography images showing large areas of consolidation with air bronchograms seen in posterior lung segments bilaterally in the dependent position. It also shows areas of ground-glass opacities in the surrounding areas.

myocarditis. In the report by Santos *et al.*,⁶ a 44-year-old female with GPA developed sustained VT preceded by bradycardia which improved after temporary pacing followed by a permanent pacemaker insertion. In another report by Hanna *et al.*,⁷ a patient with a history of recurrent otitis media presented with myocarditis, renal dysfunction, hyperkalaemia, and VT. She was treated with corticosteroids, cyclophosphamide, and plasma exchange. She showed significant improvement with treatment and her renal biopsy was suggestive of pauci-immune glomerulonephritis. Our patient differs entirely from the previous reports by the presence of pre-existent CAD. The initial presentation as ADHF made us treat him in those lines until his clinical status deteriorated because of VT storm and DAH. We also postulate that the substrate for VT storm in our patient was likely due to previously scarred myocardium as the VT morphology was monomorphic. Hyperkalaemia in the background of acidosis and hypoxaemia precipitated the VT storm in our patient. Hypoxia and acidosis may lead to global ventricular ischaemia and when combined with hyperkalaemia can promote phase 2 re-entry by further increasing repolarization reserve in epicardial ventricular tissue and thus predisposing to VT.⁸ Myocarditis being an aetiology of VT is unlikely as cardiac biomarkers were negative and there was no new LV dysfunction. However, this cannot be conclusively ruled out as cardiac magnetic resonance imaging was not performed. Also, the clinical features, ECG, and cardiac enzymes did not suggest acute coronary syndrome and thus, we did not perform coronary angiography. Though we could not get a histopathological diagnosis by renal biopsy; presence of active sediments in urine, rapidly progressive renal failure, evidence of DAH, and presence of ANCA positivity essentially established the diagnosis of ANCA-associated vasculitis.⁹ The dramatic response to steroids and plasma exchange further substantiated our diagnosis. Overall our case describes acute heart failure and VT as an uncommon first presentation of ANCA-related pulmonary renal vasculitis in a patient with pre-existing heart disease.

Conclusion

Antineutrophil cytoplasmic antibody-associated vasculitis rarely may present as ADHF in a patient with pre-existent LV dysfunction. A multidisciplinary approach is vital for diagnosis and successful treatment.

Lead author biography



Krishna Prasad is pursuing DM Cardiology (Fellowship) at PGIMER, Chandigarh, India, an institute of National importance. He has completed his MBBS from Osmania Medical College, Hyderabad, India and his MD from PGIMER, Chandigarh, India. His field of interests are complex coronary interventions and structural heart disease. He has 15 articles under his name published in national and international journals. After the fellowship

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Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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