Cardiology in the Young

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A case of myocardial infarction in a teenager with new diagnosis of granulomatosis with polyangiitis

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Brief Report

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Abstract

A 16-year-old male with newly diagnosed granulomatosis with polyangiitis presented to the emergency room with chest pain. He was found to have a myocardial infarction involving the right coronary artery and the left circumflex artery. He underwent mechanical thrombectomy and stent placement without significant sequelae. This is a rare complication associated with granulomatosis with polyangiitis.

Case presentation

A 16-year-old male with a recent diagnosis of granulomatosis with polyangiitis presented to the emergency department with severe acute chest pain that began prior to arrival. The pain radiated to his back with associated dyspnoea and diaphoresis. In the emergency room, an electrocardiogram showed ST changes (Figure 1b) as compared to baseline (Figure 1a) and elevated troponin (8287 ng/L; reference <31 ng/L). A focused echocardiogram showed hypokinetic inferior left ventricular wall and dyskinetic left ventricular septal wall motion with low normal left ventricular systolic function (ejection fraction 49%). He was given a dose of aspirin (325 mg) and acetaminophen. His chest X-ray was normal. A repeat troponin level roughly 2 hours later was 27,500 ng/L. Given concern for myocardial infarction, he was transferred to the catheterisation lab for intervention.

In the catheterisation lab, he had thrombotic occlusions in the right coronary artery and distal left circumflex artery. He underwent mechanical thrombectomy of both arteries, and a Resolute Onyx 4.5 × 22 mm drug-eluting stent was placed in the right coronary artery. He tolerated the procedure well and was prescribed ticagrelor (oral loading dose 180 mg and 2-hour intravenous infusion). He was then transferred to the cardiothoracic ICU.

In the cardiothoracic ICU, he was well appearing without chest pain. He remained on ticagrelor. He was also prescribed daily aspirin (81 mg) for dual antiplatelet therapy, metoprolol (25 mg), and atorvastatin (20 mg). His follow-up electrocardiogram showed improvement (Figure 1c) and repeat echocardiogram showed improved left ventricular function (ejection fraction 57%) and normal left ventricular global longitudinal strain (–20.9%) without regional wall abnormalities. Cardiac MRI performed 3 days post procedure revealed mild biventricular dilation and normal biventricular function with left ventricular wall motion abnormalities consistent with his myocardial infarction. There was a filling defect in the inferior wall of the left ventricle at the mid-papillary muscle level on initial perfusion imaging. There was abnormal myocardial extracellular volume on T1-mapping (42% ± 16%, normal is 26% ± 3%) and enhancement in the inferior left ventricle consistent with oedema on T2-weighted imaging. Late gadolinium enhancement showed mid-wall patchy and sub-epicardial non-ischemic fibrosis.

Before this hospital admission, he was admitted for 1 week with fever, polyarticular joint pain, conjunctivitis, left lower extremity purpura, dark urine, haematuria, proteinuria, and elevated inflammatory markers. He also had non-specific, intermittent mild chest pain with normal troponin level and normal echocardiogram. He was diagnosed with granulomatosis with polyangiitis by positive anti-neutrophil cytoplasmic antibodies 1:160 and proteinase 3 serologies. His chest pain resolved prior to discharge. He completed three days of high-dose steroids and was discharged home on oral daily prednisone 60 mg. He subsequently presented to the emergency room with the aforementioned chest pain one day later.

His family history consisted of elevated cholesterol on both maternal and paternal sides and one family member with a myocardial infarction at age 60 requiring quadruple coronary bypass. The patient had lipid testing within the last year, which showed elevated cholesterol, triglycerides, and LDL with low HDL. Repeat levels on this admission showed improvement with persistently elevated triglycerides and low HDL. His lipoprotein A was normal (Table 1).

During both hospitalisations, the patient had evidence of pulmonary and renal manifestations of granulomatosis with polyangiitis. A chest CT showed evidence of pulmonary

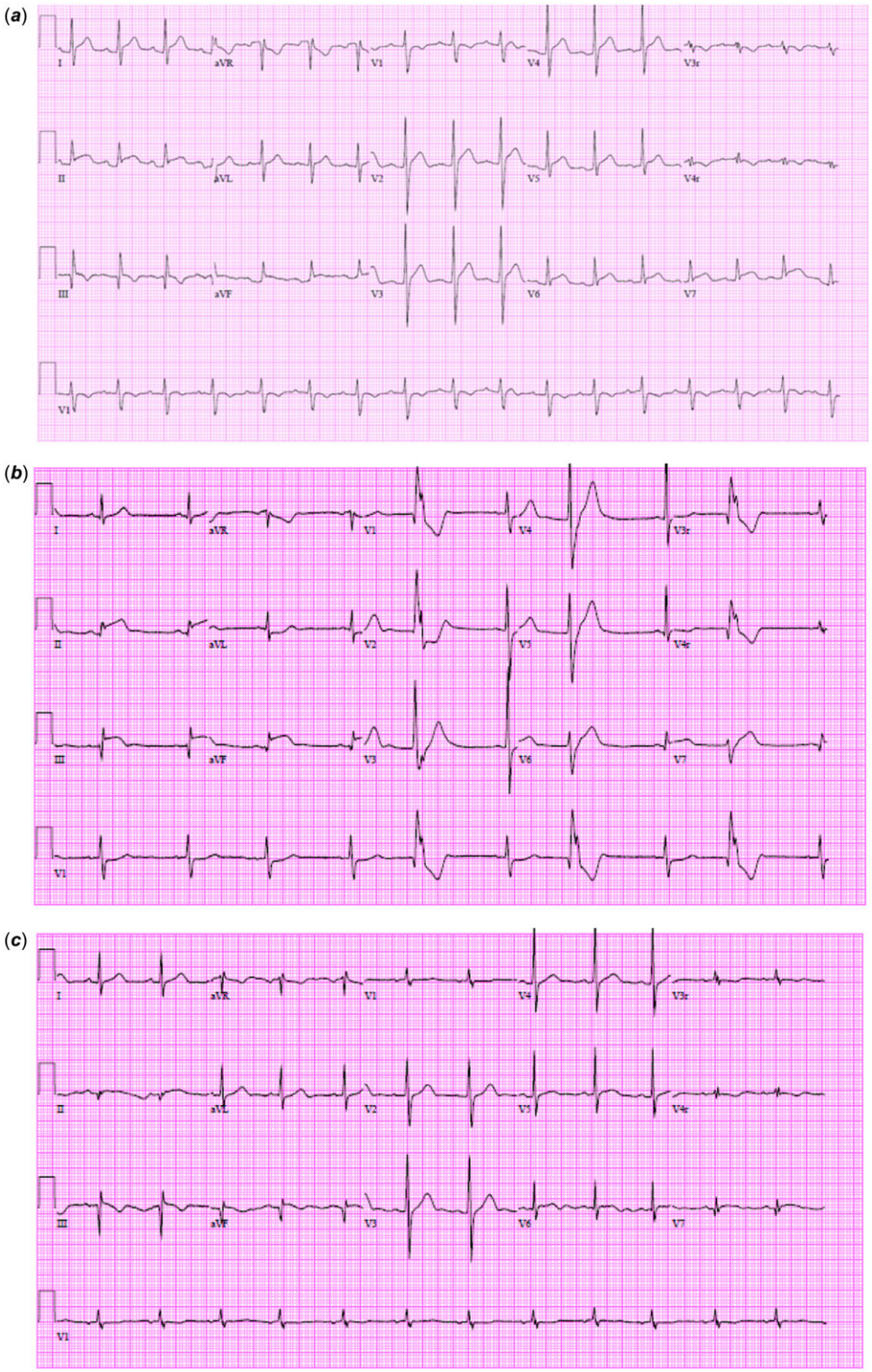

Figure 1. Electrocardiograms (ECGs) showing (***a***) Baseline ECG. (***b***) Emergency room ECG at time of acute chest pain showing new ST elevation in inferior leads II, III, and aVF and new ST depression in the right precordial leads V1 and V2. Toward the end of this strip there is also a pattern of premature ventricular contractions (PVCs) in a pattern of bigeminy. (***c***) ECG following cardiac catheterization with resolution of ST elevation, ST depression, and PVCs.

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Table 1. Patient's lipid testing results from 11 months prior to admission and during current admission with reference ranges

Lipids	11 months prior	This admission	Reference ranges
Total cholesterol	290 mg/dL	246 mg/dL	<200 mg/dL
HDL	19 mg/dL	20 mg/dL	>35 mg/dL
LDL	226 mg/dL	121 mg/dL	<130 mg/dL
Triglycerides	223 mg/dL	246 mg/dL	<200 mg/dL
Lipoprotein A	–	15 nmol/L	<75 nmol/L

ground-glass opacities and pulmonary haemorrhage, and the patient required dialysis for kidney failure during this latter hospital stay. Ultimately, he completed three additional days of high-dose steroids and single doses of rituximab and cyclophosphamide. He was discharged home with close subspecialty follow-up and without further cardiac complications.

Discussion

This case represents a rare finding of myocardial ischaemia in the setting of a new diagnosis of granulomatosis with polyangiitis. The mean age of presentation is roughly 40 years of age.[1] There are several studies that show a wide range of cardiac complications in patients with granulomatosis with polyangiitis. Historically, there are two studies that compare cardiac involvement in granulomatosis with polyangiitis. One study by Hoffman et al. from 1992 shows 6% of patients with cardiac involvement (pericarditis), while a study from 1977 by Pinching et al. shows 30% cardiac involvement (including coronary vasculitis with myocardial infarction and pericarditis).[1–3] A study reviewing literature from 2007 to 2017 investigated cardiac complications in patients with granulomatosis with polyangiitis and showed a rate of 0.3% of coronary artery disease, which was less common than congestive cardiac failure (6%), ischaemic cardiac pain (4%), pericarditis (2%), cardiomyopathy (0.6%), and only more common than valvular disease (0.03%).[4]

A review of the literature in 2018 shows coronary vasculitis was noted in up to 50% of patients with post-mortem autopsy evaluation, though angina and myocardial infarction are rare complications.[5] For myocardial infarction, six cases have previously been described in patients ranging from age 20 to 42, of which 4 patients were asymptomatic and 3 cases were fatal.[5] Finally, one recent study in 2016 by Aviña-Zubieta et al. investigated 504 patients with granulomatosis with polyangiitis, 23 patients developed myocardial infarctions, which is an incidence of 11.7/1000 person-years as compared to 5.2/1000 person-years in patients without granulomatosis with polyangiitis (hazard ratio 1.86, 95% confidence interval 1.05–3.31).[6] This risk was noted to be the highest during the first year after granulomatosis with polyangiitis diagnosis.[6] Myocardial infarction is an extremely rare complication in patients with granulomatosis with polyangiitis that can be fatal when it is the presenting symptom.

Conclusion

It is important to consider cardiac manifestations in patients with rheumatologic disorders such as granulomatosis with polyangiitis. Providers should maintain a high level of suspicion for cardiac involvement in this patient population, especially in patients with new diagnoses with ongoing inflammation. This patient made a full recovery following cardiac catheterisation and continues to receive care for other aspects of granulomatosis with polyangiitis.

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Competing interests. None.

Ethical standards. Obtained.

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