

A Case of Acute Thrombotic Myocardial Infarction in Polyarteritis Nodosa

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ABSTRACT

This is a case of a 33-year-old male with acute myocardial infarction from complete thrombotic occlusion of the distal left anterior descending and 1st obtuse marginal artery secondary to polyarteritis nodosa. This case highlights the treatment course and need for continued awareness of vasculitis as a cause for myocardial infarction.

KEYWORDS: acute myocardial infarction, polyarteritis nodosa, vasculitis

function with an ejection fraction (EF) of 64%. He was switched from ibuprofen to methylprednisolone due to concern for systemic vasculitis. Three days later, his chest pain recurred in association with ST segment elevations in leads I, II, V3-V6 with new atrial fibrillation (AF) [Figure 1].

Emergent transthoracic echocardiogram (TTE) demonstrated an EF of 35% with extensive regional dysfunction of the apical and anterior walls. Cardiac catheterization demonstrated diffuse ectasia of the coronary arteries with thrombotic occlusion of the distal left anterior descending (LAD) artery and the first obtuse marginal (OM) branch

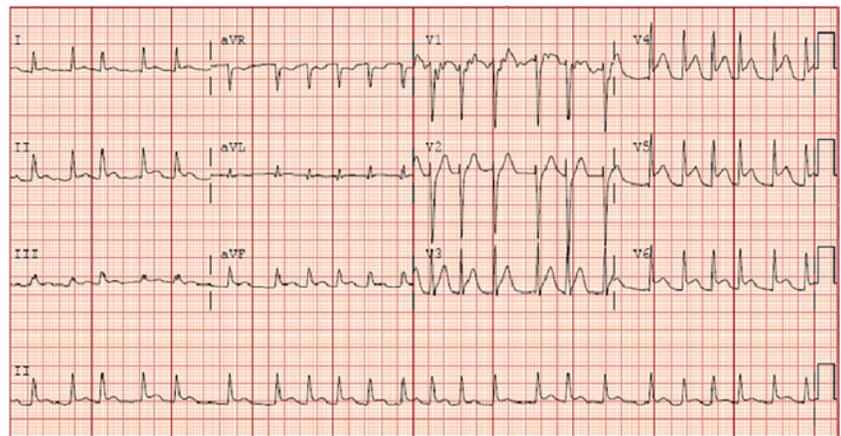
CASE REPORT

A 33-year-old man with a history of biopsy-proven testicular vasculitis and myopericarditis diagnosed 3 weeks prior presented with acute, left-sided, non-radiating, pleuritic chest pain worse with exertion. He also endorsed night sweats and unintentional weight loss of 20 lbs. in 4 months. Due to the recent inflammatory disorders, he had been treated with colchicine and a taper of ibuprofen, which had helped relieve his symptoms earlier, but were no longer effective for his chest pain.

On physical exam, the patient had a BP of 133/83 with a HR of 114 bpm. He was afebrile with an SpO₂ of 99% on room air. He was noted to have bilateral conjunctivitis. He otherwise had clear lungs, normal heart sounds with no murmurs or rubs, soft and non-tender abdomen, and no skin lesions or rashes. Labs noted CRP 91 ml/L, ESR 130 mm/hr, and troponin 0.174. The patient had followed-up with outpatient rheumatology prior to the current presentation; however, autoimmune work-up involving ANA, ANCA, IgA, Anti-dsDNA antibody, rheumatoid factor, and hepatitis B surface antigen was negative. Admission ECG showed sinus tachycardia.

During the hospitalization, the patient was initially treated for recurrent myopericarditis with ibuprofen and colchicine. However, his chest pain persisted with development of shortness of breath. On hospital day 2, troponin increased to 10.5 without ECG changes, but with cardiac MRI showing focal areas of myonecrosis and normal biventricular

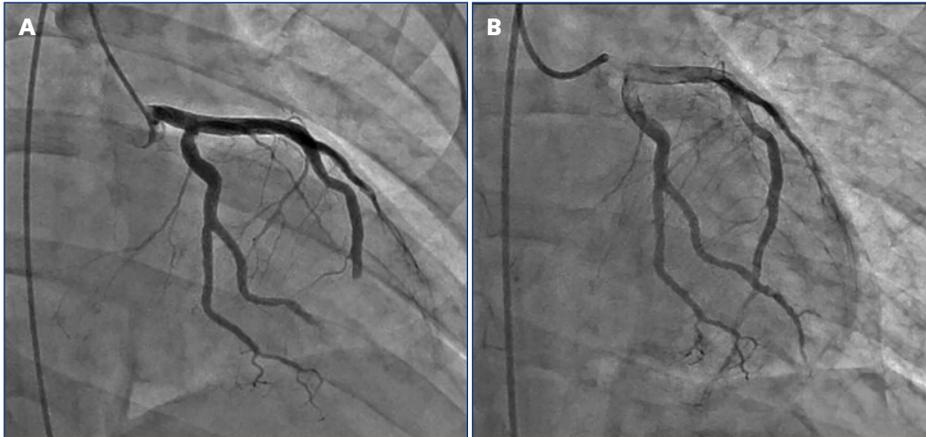
Figure 1. ECG showing new ST segment elevations in leads I, II, V3-V6 as well as new atrial fibrillation.



of the circumflex. (Figures 2A, 2B) He underwent primary percutaneous coronary intervention (PCI) to both culprit vessels with balloon angioplasty and thrombectomy with a bare metal stent (BMS) in the LAD. These cardiac findings were consistent with vasculitis and coronary thrombosis or embolism.

The diagnosis of systemic polyarteritis nodosa (PAN) was made based on his history of testicular vasculitis, weight loss, episcleritis, and acute thrombotic MI. The patient was started on pulse dose steroids with transition to oral prednisone and cyclophosphamide. For the myocardial infarction and subsequent PCI, he received aspirin, clopidogrel, atorvastatin, and metoprolol. For paroxysmal AF, he was anticoagulated with unfractionated heparin until therapeutic on coumadin and started on digoxin for better rate control.

Figure 2. [A] Cardiac Angiography during catheterization showing 100% thrombotic stenosis in the distal Left Anterior Descending (LAD) and 1st Obtuse Marginal (OM) branch artery. **[B]** Revascularization of the distal LAD and 1st OM branch artery.



By time of discharge, he had converted to a normal sinus rhythm on ECG.

At two months post-discharge, the patient had no recurrent chest pain and continued to attend cardiac rehabilitation. However, TTE from 10-month follow-up shows a severely reduced EF of 25% with severely dilated left ventricle.

DISCUSSION

Polyarteritis Nodosa

PAN is an ANCA-negative necrotizing vasculitis of small- and medium-sized arteries, not associated with glomerulonephritis. It can result in a multitude of disorders and can be either idiopathic or triggered by an agent, i.e. hepatitis B.¹ While it can affect many organ systems, PAN is a rare disease with a prevalence of about 31 cases/million affecting individuals of any gender, age, and race.¹

Diagnostic Criteria

Systemic PAN can be diagnosed based on the American College of Rheumatology criteria,² which requires 3 out of the following:

- weight loss ≥ 4 kg
- livedo reticularis
- testicular pain or tenderness
- myalgias
- mono- or polyneuropathy
- diastolic blood pressure >90 mmHg
- elevated blood urea nitrogen or serum creatinine levels
- presence of hepatitis B reactants in the serum
- arteriographic abnormality
- presence of granulocyte or mixed leukocyte infiltrate in an arterial wall biopsy

With weight loss, testicular pain with testicular mass status post orchiectomy, biopsy proven testicular vasculitis, and arteriographic evidence during cardiac catheterization, this patient fulfilled four criteria.

Rates of Complication

Cardiac complications are known sequelae of some vasculitides like Kawasaki's, Takayasu's, and eosinophilic granulomatosis with polyangiitis,³ but are not as well recognized with PAN vasculitis. A recent pathological study of 37 autopsy cases of PAN showed 81% had cardiac vascular involvement,

most commonly with panarteritis;⁴ however, clinically significant cardiac manifestations are rarely seen pre-mortem. Cardiac involvement has been reported to occur in 4–30% of PAN cases, usually manifesting as congestive heart failure.¹ The occurrence of acute MI is rare and has been described to stem from aneurysmal, thrombotic, or vasospastic events caused by widespread inflammation from PAN.⁵ As these manifestations can be silent in early stages of vasculitides, heart function should be evaluated by an ECG or TTE. If these reveal abnormalities or the patient develops symptoms, further investigations, such as cardiac MRI or more invasive procedures can be pursued.⁶

Treatment

Symptomatic cardiac involvement requires aggressive treatment given poor prognosis with 13% five-year survival.⁷ Those with evidence of cardiac, gastrointestinal, neurological, or renal involvement are categorized as having moderate to severe PAN and should be treated with both glucocorticoids and cyclophosphamide, as the combination has demonstrated better outcomes including rates of survival.^{8,9} In recalcitrant cases of PAN, biologic therapies such as tumor necrosis factor antagonists (i.e rituximab and infliximab) have been shown to be potential alternative options.¹⁰ Various case reports have also identified tocilizumab, a humanized monoclonal anti-interleukin-6 receptor antibody, as a treatment option for those who do not respond to the glucocorticoid-cyclophosphamide combination.¹⁰

Along with glucocorticoids and cyclophosphamide, our patient also required balloon angioplasty and thrombectomy with placement of a BMS. A stent had to be placed in this case because despite multiple passes with a thrombectomy catheter and balloon angioplasty, the flow in the LAD remained slow due to the thrombus. When vessel size is adequate, trapping the thrombus behind the stent scaffold can be beneficial for limiting the extent of the infarction.

As to why a BMS was used instead of a drug eluting stent (DES), the operator did not specify. However, one possible reason could be that while we do not have data on endothelial healing in patients with vasculitis, DES have delayed healing compared to BMS, which can increase the risk of late stent thrombosis, especially after discontinuation of dual antiplatelet therapy (DAPT). Furthermore, given that this patient had developed new atrial fibrillation necessitating oral anticoagulation, BMS may shorten the duration of triple therapy of DAPT and anticoagulation as that has a high risk of bleeding.

CONCLUSION

Patients with PAN occasionally present with cardiac manifestations, but rarely do they present with acute coronary syndrome (ACS). Multiple case studies have been reported of patients with PAN presenting with ACS, but there is still no consensus on how to treat cardiac stenotic lesions in patients with PAN. In a case of acute MI due to PAN in a young female with myocardial SPECT with minimal reversible ischemia and no recurrent chest pain, the patient had no additional events with standard cardiac medical management and immunosuppressive therapy.¹¹ In another case, a patient with a BMS followed by placement of DES and another BMS on a separate occasion, underwent a coronary artery bypass graft after a repeat coronary angiography found mildly aneurysmal left coronary vessels with multiple stenoses.¹² While clinically significant cardiac involvement in PAN is rare, it can be life threatening. It is not only important to recognize PAN early, but also to evaluate cardiac function soon after the diagnosis of PAN is made. We believe this patient was appropriately treated in a timely manner, though with the subacute presentation of his chest pain in setting of a recent diagnosis of myopericarditis, ACS superficially may have seemed less likely the cause of his symptoms. As such, thrombotic occlusion and possible acute coronary syndrome should always be maintained in the differential in any PAN patient with chest pain.

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Disclosures

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