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VOL. 2, NO. 10, 2020

MINI-FOCUS ISSUE: HEART FAILURE

ADVANCED

CASE REPORT: CLINICAL CASE

Relapsing Polychondritis Requiring Orthotopic Heart Transplant Despite Coronary Artery Bypass and Surgical Aortic Valve Replacement

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ABSTRACT

A 32-year-old man with a history of relapsing polychondritis presented with acute coronary syndrome due to aortitis with ostial coronary artery involvement from his underlying autoimmune condition. Concomitant aortic insufficiency with ostial coronary lesions is a rare complication of relapsing polychondritis, requiring a multidisciplinary team approach for management. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2020;2:1527–31) Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 32-year-old man with relapsing polychondritis (RP) presented to our emergency department for worsening chest pain of 10 days' duration. The pain was substernal and pressure-like, radiating to the left shoulder and arm. Examination findings at the time were as follows: blood pressure, 159/94 mm Hg; heart rate, 87 beats/min; respiratory rate, 16 breaths/min; temperature, 36.3°C; 99% oxygen saturation on room air; and grade 4 of 6 decrescendo diastolic murmur in the right upper sternal edge. His troponin I level was 0.21 ng/ml.

LEARNING OBJECTIVES

- To broaden the differential for etiologies of ST-segment elevation myocardial infarction/non-ST-segment elevation myocardial infarction.
- To recognize aortic insufficiency and ostial coronary artery disease as a rare cardiac complication of RP.

MEDICAL HISTORY

The patient had an 11-year history of RP and coarctation of the descending thoracic aorta requiring percutaneous stenting 1 year before presentation. He initially presented with nasal and auricular chondritis, inflammatory arthritis, and aortitis. He was started on azathioprine, but it was discontinued due to pancreatitis. He was transitioned to prednisone and methotrexate, which was tapered 3 weeks before presentation.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Case Reports* author instructions page.

Manuscript received April 1, 2020; accepted May 29, 2020.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2020.05.100

ABBREVIATIONS AND ACRONYMS

ECG = electrocardiography

ECMO = extracorporeal membrane oxygenation

LAD = left anterior descending

MMF = mycophenolate mofetil

OHT = orthotopic heart transplant

RCA = right coronary artery

RP = relapsing polychondritis

DIFFERENTIAL DIAGNOSIS

Fulminant aortitis with involvement of the coronary arteries was our initial concern, as the patient's symptoms coincided with recent taper of immunosuppression, and diagnostic testing (electrocardiography [ECG] and troponin levels) was concerning for coronary ischemia. Aortic dissection, a severe complication of aortitis, was also considered. Vasculitis of the coronary arteries, plaque rupture, spontaneous coronary artery dissection, and coronary artery embolism are in the differential diagnosis for a patient with vasculitis and hypercoagulability.

INVESTIGATIONS

The first 12-lead ECG revealed no significant ST-segment changes (**Figure 1A**). One hour later, the patient's chest pain acutely worsened. A repeat ECG revealed diffuse ST-segment depressions (**Figure 1B**). Subsequent computed tomography angiography of the aorta showed thickening of the aortic root to the proximal descending thoracic aorta and left subclavian artery consistent with large vessel vasculitis, with no evidence of superimposed dissection (**Figure 2**). Another ECG immediately after aortic imaging showed ST-segment elevation in V_1 to V_3 (**Figure 1C**). The patient was emergently taken to the cardiac catheterization laboratory, and coronary angiography was performed (**Figure 3**). This showed 95% ostial stenosis of the left main coronary artery. The left anterior descending (LAD) and left circumflex arteries were patent (Video 1). The right coronary artery (RCA) was unable to be engaged, with no vessel noted on aortography. The aortogram demonstrated severe aortic regurgitation (Video 2).

MANAGEMENT

Given the severe aortic regurgitation with critical left main coronary artery stenosis, cardiothoracic surgery was consulted for emergent surgical aortic valve replacement and coronary artery bypass graft surgery. During anesthesia induction, the patient developed pulseless electrical activity. Cardiopulmonary resuscitation was initiated while the surgeon performed a median sternotomy. Internal cardiac massage was continued as the patient was centrally cannulated for extracorporeal membrane oxygenation (ECMO). The patient had return of spontaneous circulation in 21 min. Given the patient's history of vasculitis, the left internal mammary artery was not harvested. The patient underwent saphenous vein graft bypass of the LAD and RCA with mechanical aortic valve replacement (Regent 21 mm, Abbott, Abbott Park, Illinois). An intra-operative transesophageal echocardiogram revealed severely reduced biventricular function. Despite surgical intervention, the patient was unable to be weaned from the ECMO apparatus, with a cardiac index <1.2 l/min/m^2. Six days after cannulation for ECMO, the patient was transitioned to a biventricular CentriMag assist device via central cannulation (Abbott, Abbott Park, Illinois). The patient was started on methylprednisolone and mycophenolate mofetil (MMF) with subsequent positron emission tomography-computed tomography imaging showing resolution of active inflammation. Ultimately, he was accepted for listing for an orthotopic heart transplant (OHT) with the United Network for Organ Sharing as status 1. Six weeks after a biventricular assist device was placed, the patient successfully underwent transplant. The patient was maintained on prednisone, MMF, and tacrolimus.

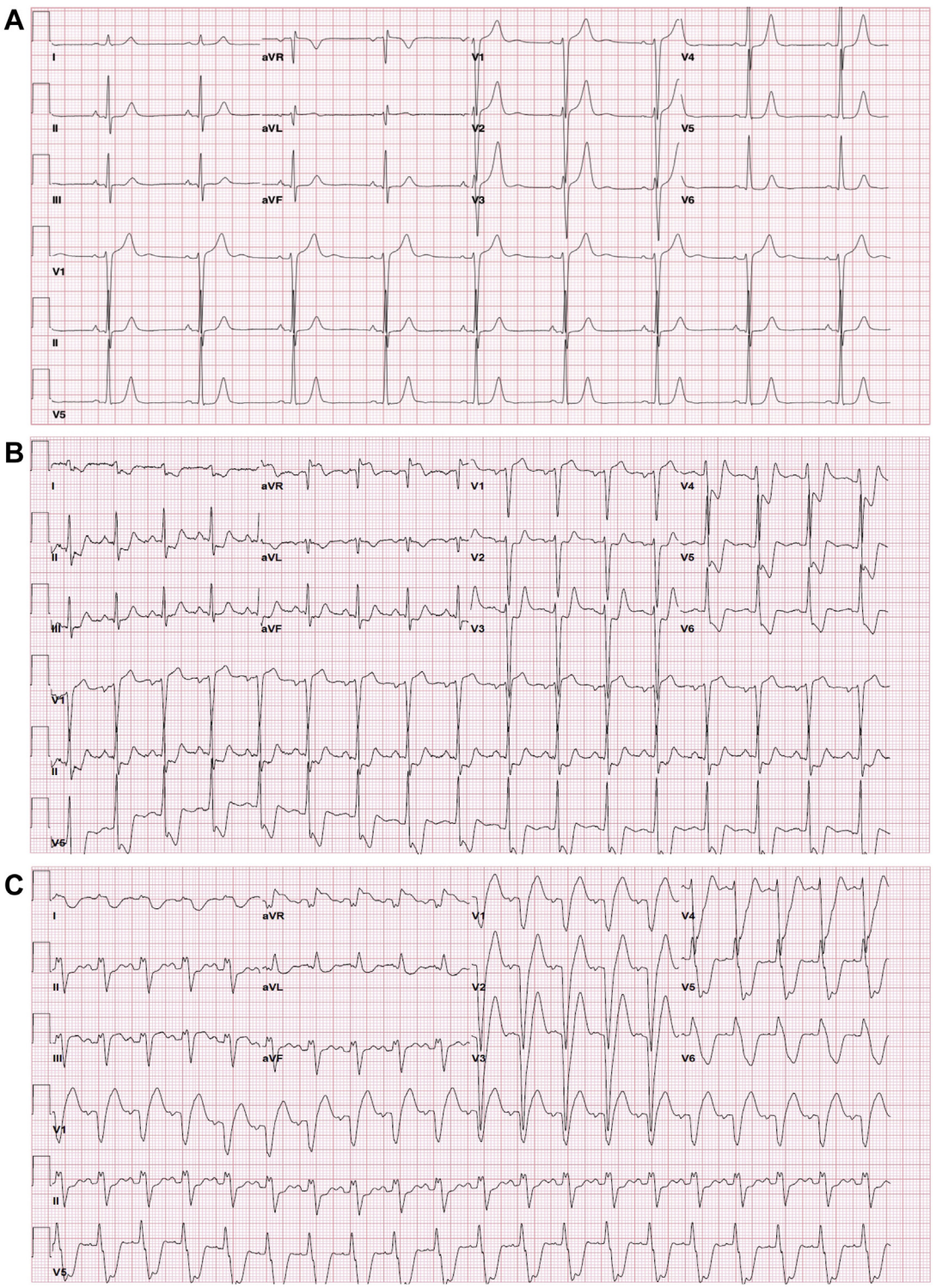
DISCUSSION

RP is a rare autoimmune condition that involves cartilaginous tissues, with an incidence of 3.5 cases per million (1). Cardiovascular manifestations have been reported in 11% to 56% of these patients and include vasculitis of both large- and medium-sized arteries, aortic regurgitation, and pericarditis (2). Although aortic insufficiency is the most common cardiovascular complication observed in 4% to 9% of these patients (3), the combination of aortic insufficiency from aortitis with ostial coronary lesion is exceptionally rare: 7 cases have previously been described in the literature. Of those cases, only 1 patient successfully underwent coronary artery bypass grafting with aortic valve replacement.

A combination of coronary artery bypass graft with aortic valve replacement for RP has been reported by Kisamori et al. (4), in which the RCA was involved. However, in the current case, both the left and right coronary arteries had critical ostial lesions. Given the left subclavian artery involvement with vasculitis in this case, there was concern that the left internal mammary artery was compromised. As such, saphenous venous grafts were opted to bypass the LAD and RCA. Despite revascularization and resolution of aortic insufficiency, the patient remained in cardiogenic shock requiring biventricular mechanical support. OHT was subsequently offered as rescue therapy.

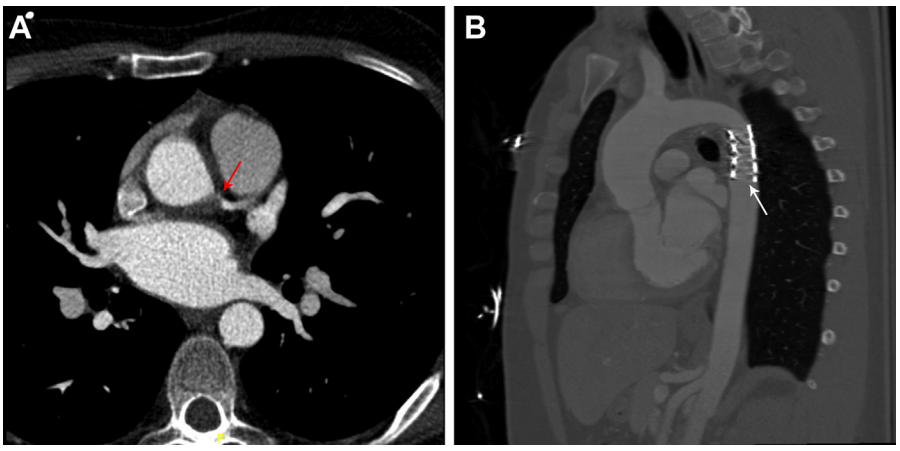
The mortality rate for RP with cardiovascular involvement is abysmal, with estimates as high as

FIGURE 1 Progression of Ischemia on 12-Lead ECGs During Hospitalization Course



(A) 12-lead electrocardiography (ECG) showing sinus bradycardia with left ventricular hypertrophy. **(B)** 12-lead ECG showing ST-segment elevation in aVR and ST-segment depressions in V_2 to V_6, III, aVF, and I. **(C)** 12-lead ECG showing ST-segment elevation in the precordial leads, V_1 to V_3.

FIGURE 2 Chest CT Angiography Demonstrating Significant Ostial Stenosis of the Left Main Coronary Artery



(A) Computed tomography angiography image obtained in the axial view, showing mural thickening of the aortic root with no evidence of ascending aortic dissection or aortic root aneurysm. Left main coronary artery with significant ostial stenosis **(red arrow)**. **(B)** Computed tomography angiography image obtained in the sagittal view, showing patent endovascular stent located in the descending aorta **(white arrow)** and eccentric thickening of the aortic wall proximal to the endovascular graft.

52.6% (5); this is a departure from the overall prognosis of RP, which has been reported as 91% in 10 years (6). Although many of these patients with RP will require surgical intervention for cardiac manifestations, outcomes have been poor in this cohort. Post-operative complications from valve and aortic root replacements include paravalvular leakage and prosthesis dehiscence related to the friable tissue from the inflammatory process, with up to 24% of patients requiring re-operation (5). Some authors have suggested a prophylactic Bentall operation for RP involving the ascending aorta (2).

The known post-operative complications were applicable in our patient undergoing OHT, requiring surgical anastomosis to an inflamed aortic root. As such, the patient's RP was first stabilized by using a combination of immunosuppressive therapies (glucocorticoids and MMF). Sharma et al. (7) describe a RP case series with successful use of second-line agents, including azathioprine, methotrexate, cyclosporine, and MMF. Considering our patient's tenuous renal function and pancreatitis with azathioprine use, MMF was selected as an adjunctive therapy, leading to resolution of the RP flare. At the time of OHT, the prophylactic Bentall procedure was deferred given that the sutures held well on the thickened aorta, the aortic root was of normal size (2.2 cm × 2.8 cm), and an aortic root replacement would significantly add to the complexity of the surgery.

This case still represents the second case of OHT for RP. Immunosuppressive treatment, the same for OHT and RP, led to successful resolution of the RP.

FOLLOW-UP

The patient is 1 year out of his OHT, with no relapse of RP. Subsequent coronary angiography revealed patent transplant coronary arteries, most recently 3 months from OHT. The patient is continued on prednisone indefinitely in addition to tacrolimus and MMF.

CONCLUSIONS

Concomitant aortic insufficiency from aortitis with ostial coronary lesions is a rare complication of RP. This case highlights the critical importance of a multidisciplinary team approach to managing a patient with this rare condition.

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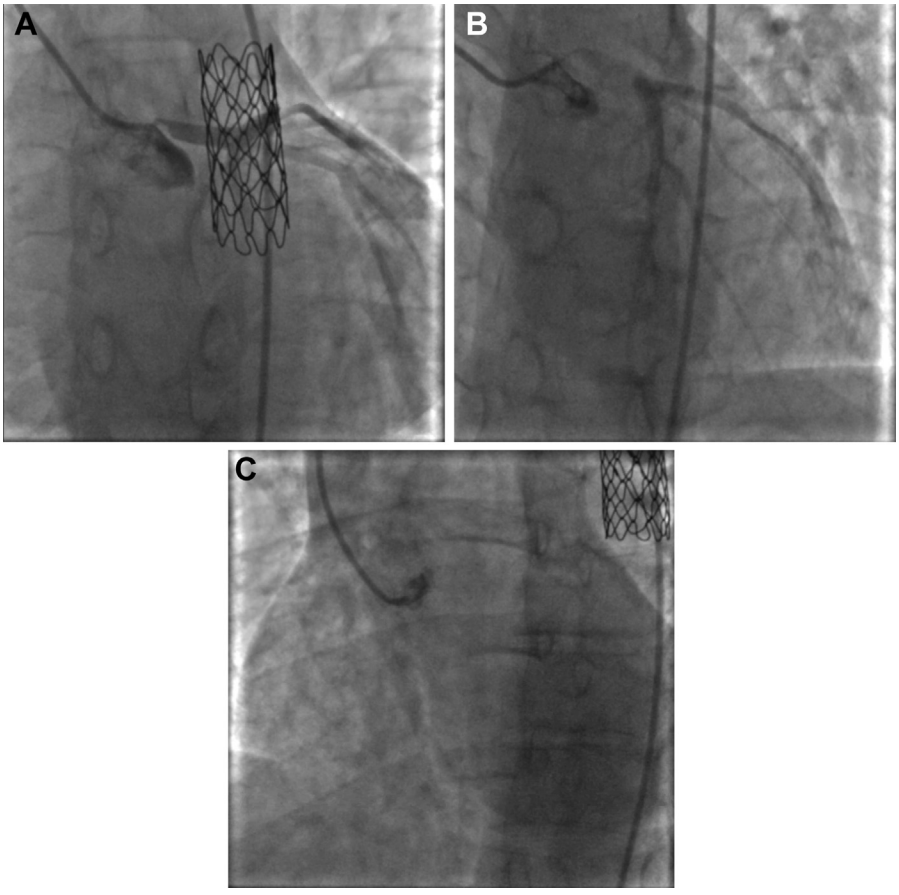
FIGURE 3 Coronary Angiogram Demonstrating Ostial Left Main Coronary Artery Stenosis and Right Coronary Artery Stenosis



(A) Right anterior oblique caudal angiographic image showing 95% lesion in the ostial left main coronary artery. **(B)** Left anterior oblique cranial angiographic image showing significant ostial left main coronary artery stenosis. The left circumflex and left anterior descending arteries are patent. **(C)** The right coronary artery is unable to be engaged.

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KEY WORDS aortic insufficiency, cardiogenic shock, heart transplant, myocardial infarction, ostial coronary lesion

APPENDIX For supplemental videos, please see the online version of this paper.