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CASE REPORT

BEGINNER

CLINICAL CASE

A 31-Year-Old Man With Angina Pectoris Resulting From Large Vessel Vasculitis

Giselle M. Statz, MD,[a] Noah N. Williford, MD,[a,b] Vivian R. Sardone, MD,[c,d] Zuhair K. Ballas, MD,[e,f] Svjetlana Dolovcak, MD,[e,f] James Rossen, MD,[g] Milena A. Gebska, MD, PhD, MME[g]

ABSTRACT

We present the case of an apparently healthy 31-year-old man with malignant progression of coronary artery disease and recurrent angina resulting from suspected large vessel vasculitis. This case highlights the importance of considering vasculitis in patients without atherosclerotic risk factors, using a multidisciplinary team approach, and suppressing inflammation *before* coronary revascularization to improve outcomes. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2021;3:1191–3) © 2021 The Authors. 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/).

A healthy 31-year-old man presented with angina, a normal resting electrocardiogram, and effort ischemia. His vital signs and physical examination were unremarkable. Coronary angiography revealed subtotal occlusion of the left main coronary artery (LMCA) and the left circumflex (LCx) artery, with collateral vessels from the right coronary artery (RCA) (**Figures 1A and 1B,** Supplemental Figures 1A and 1B, Video 1).

LEARNING OBJECTIVES

- To suspect vasculitis, rather than atherosclerosis, in young patients with myocardial ischemia and no cardiovascular risk factors.
- To prioritize the importance of suppression of inflammation in patients with coronary vasculitis prior to referral for revascularization in order to prevent premature bypass graft failure or malignant in stent restenosis.

Results of all laboratory tests were acceptable, except the following: C-reactive protein (CRP), 1.8 mg/dl (reference: ≤0.5 mg/dl); and erythrocyte sedimentation rate, 19 mm/h (reference: 0 to 15 mm/h). He underwent coronary artery bypass grafting (CABG) using bilateral internal mammary arteries. Intraoperatively, no suitable LCx artery target was identified; the aortic wall appeared thickened.

Two months after CABG, he developed fevers, night sweats, severe temporal headaches, and myalgias. Comprehensive infectious disease and rheumatologic evaluations were unrevealing despite elevated CRP (12.2 mg/dl), cardiac CRP (169 mg/dl; highest risk: 3.9 to 15 mg/dl), and erythrocyte sedimentation rate (100 mm/h). Large vessel vasculitis or aortitis was suspected (negative antineutrophil

From the [a]Department of Internal Medicine, Carver College of Medicine, University of Iowa, Iowa City, Iowa, USA; [b]Division of Cardiovascular Medicine, Washington University in St. Louis, Saint Louis, Missouri, USA; [c]Department of Undergraduate Medical Education, Carver College of Medicine, University of Iowa, Iowa City, Iowa, USA; [d]Department of Internal Medicine, Scripps Clinic and Scripps Green Hospital, San Diego, California, USA; [e]Division of Immunology, Iowa City VA Health Care System, Iowa City, Iowa, USA; [f]Division of Immunology, Carver College of Medicine, University of Iowa, Iowa City, Iowa, USA; and the [g]Division of Cardiovascular Medicine, Carver College of Medicine, University of Iowa, Iowa City, Iowa, USA.

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ABBREVIATIONS AND ACRONYMS

CABG = coronary artery bypass grafting

CRP = C-reactive protein

IL = interleukin

LCx = left circumflex

LMCA = left main coronary artery

PCI = percutaneous coronary intervention

RCA = right coronary artery

cytoplasmic antibodies test result). Results of a temporal artery biopsy were normal. Fluorine-18 fluorodeoxyglucose positron emission tomography confirmed hypermetabolic uptake along the ascending aorta (**Figure 1C**), so prednisone and, later, methotrexate were initiated for suspected giant cell arteritis.

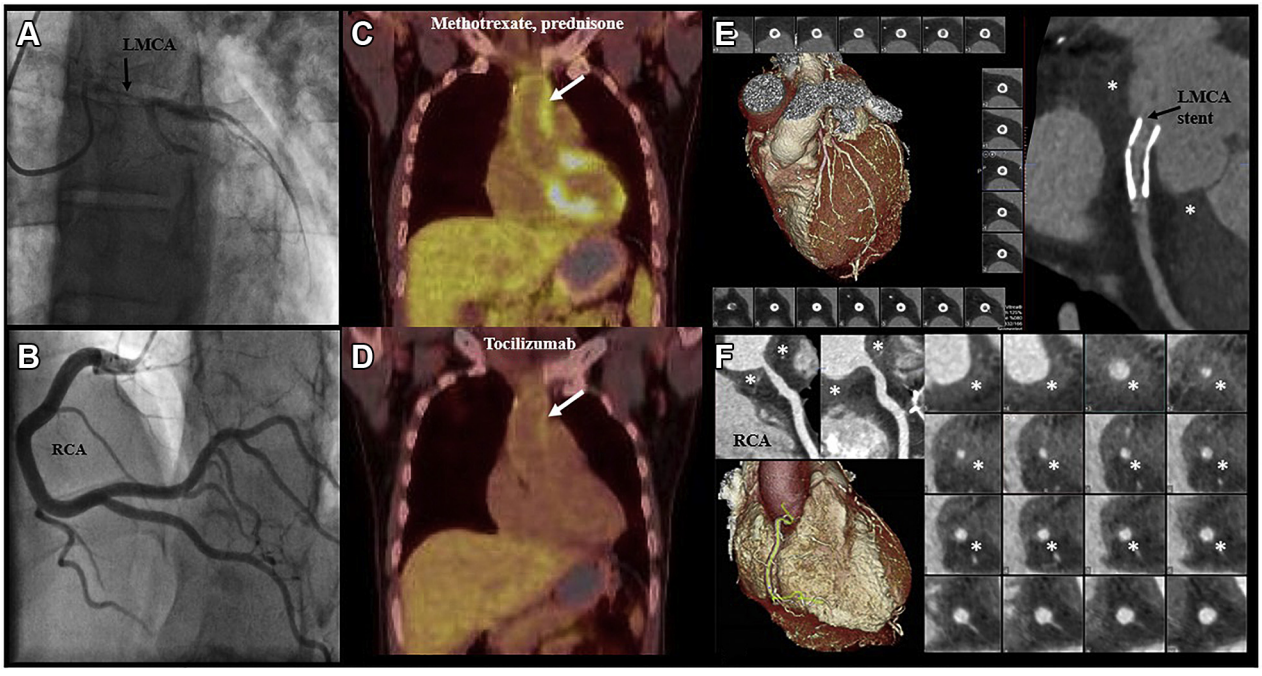
One year after CABG, angina recurred as a result of graft failure (Supplemental Figures 1C and 1D, Video 2). The patient required percutaneous coronary intervention (PCI) to the LMCA and ostial LCx artery, and this procedure was undertaken without intracoronary imaging guidance. Despite adherence to antiplatelet therapy, surveillance angiography 3 months later (17 months post-CABG) revealed in-stent restenosis and graft disease requiring redo PCI. The clinical team suspected that this condition was malignant (Supplemental Figures 1E and 1F, Video 3).

Three months later (20 months post-CABG), surveillance coronary angiography revealed LMCA in-stent complete occlusion with extensive RCA collateral vessels and new ostial RCA stenosis (Supplemental Figures 1G and 1H, Video 4) that required PCI. Surveillance coronary computed tomography (22 months post-CABG) showed a patent LMCA stent, circumferential thickening around the ascending aorta (**Figure 1E**), and new ostial RCA compression (**Figure 1F**). Angina recurred despite prednisone and methotrexate.

CLINICAL DECISION

There was a mutual agreement to pursue more effective targeted immunomodulation *before* any further coronary intervention. High-resolution human leukocyte antigen (HLA) tissue typing (DRB1 04:02, 07:01; HLA B 13:02, 35:01) suggested either Takayasu arteritis or giant cell arteritis. Serum cytokine testing revealed slightly elevated interleukin (IL)-2 (24 pg/ml; reference: ≤12 pg/ml), IL-6 (11 pg/ml; reference: ≤5 pg/ml), and IL-13 (14 pg/ml; reference: ≤5 pg/ml). On the basis of a multidisciplinary team approach, tocilizumab was added despite a lack of guideline recommendations at that time.

FIGURE 1 Imaging Before and After Treatment



Initial coronary angiography showing **(A)** a subtotally occluded left main coronary artery (LMCA) and left circumflex artery with **(B)** a patent dominant right coronary artery (RCA). **(C and D)** Positron emission tomography fluorine-18 fluorodeoxyglucose uptake **(arrows)** in response to therapy. Cardiac computed tomography showing **(E)** patent LMCA stent with **(F)** new ostial RCA compression (**asterisks** represent aortitis). LAD = left anterior descending (coronary artery).

At 7-year follow-up (5 years of tocilizumab treatment), the patient remained without angina, inflammatory markers normalized, and inflammation in the ascending aorta decreased (**Figure 1D**). Surveillance angiography showed patent stents (Video 5).

In conclusion, vasculitis should be suspected in patients without typical atherosclerotic risk factors who present with angina. This case highlights the importance of a multidisciplinary team approach and inflammatory modulation before coronary revascularization. We support the European League Against Rheumatism (EULAR) updates and the addition of tocilizumab to prevent relapse (1); however, large vessel vasculitis or aortitis limiting coronary flow remains a clinical challenge.

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ADDRESS FOR CORRESPONDENCE: Dr. Milena A. Gebska, Division of Cardiovascular Medicine, Carver College of Medicine, University of Iowa, 200 Hawkins Drive, Iowa City, Iowa 52242, USA. E-mail: milena-gebska@uiowa.edu.

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KEY WORDS coronary artery bypass, coronary revascularization, large vessel vasculitis, vasculitis

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