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CORONARY, PERIPHERAL, AND STRUCTURAL INTERVENTIONS

CLINICAL CASE

Giant Coronary Pseudoaneurysm Due to Behçet's Disease

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ABSTRACT

Behçet's disease (BD) is a systemic vasculitis which manifests through diverse clinical manifestations, including oral and genital ulcers, and ocular lesions, and has potential for neurologic, gastrointestinal, and cardiovascular involvement. Coronary artery pseudoaneurysm is a very rare complication of BD. This paper presents the case of a giant coronary pseudoaneurysm presenting with acute coronary syndrome. Immunosuppressive therapy and coronary artery bypass grafting remain the most common treatment approach and are associated with excellent outcomes in surgically suitable candidates. This paper presents a case of a 27-year-old man with a history of BD presenting with acute coronary syndrome due to giant saccular coronary pseudoaneurysm. (JACC Case Rep. 2025;30:103076) © 2025 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/).

HISTORY OF PRESENTATION

A 27-year-old man presented to the emergency department with a chief complaint of left-sided chest pain that radiated to his left shoulder. The pain began a few days prior, progressively worsened, and was not relieved by over-the-counter pain medications. The pain worsened on deep breathing and coughing. Of note, he had similar episodes of less severe pain the month prior and had been diagnosed with both a small pulmonary embolism (PE) and pericarditis. He was on apixaban at time of representation along with colchicine. All other review of systems were negative on presentation.

On examination, his temperature was 37.1 °C, blood pressure was 122/73 mm Hg, heart rate was 72 beats/min, respiratory rate was 22 breaths/min, and oxygen saturation was 99% on room air. Cardiac examination demonstrated regular cardiac rate and rhythm with no rub or appreciable murmur. Lungs were clear. He had no rashes or other skin or mucosal lesions.

TAKE-HOME MESSAGES

- This case highlights the rare but important complication of coronary artery aneurysm and pseudoaneurysm among patients with Behçet's disease and the need to obtain prompt coronary artery imaging in patients with chest symptoms, even in the absence of traditional cardiovascular risk factors.
- Management of coronary pseudoaneurysm involves a combination of immunosuppression and coronary revascularization, generally surgical, with a treatment plan determined and coordinated by a multidisciplinary care team.

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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 Author Center.

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ABBREVIATIONS AND ACRONYMS

ACS = acute coronary syndrome

BD = Behçet's disease

CTA = computed tomography angiography

LAD = left anterior descending

PCI = percutaneous coronary intervention

PE = pulmonary embolism

PAST MEDICAL HISTORY

The patient was diagnosed with Behçet's disease (BD) at 16 years of age. He had been lost to rheumatology follow-up and had not taken prescribed methotrexate (10 mg once a week) for more than a year. Home medications were colchicine 0.6 mg oral twice daily, metoprolol tartrate 25 mg oral twice daily, naproxen 50 mg twice daily as needed, apixaban 5 mg twice daily, and fluoxetine 10 mg daily.

DIFFERENTIAL DIAGNOSIS

At this point in his evaluation, differential diagnosis included worsening pericarditis and/or pericardial effusion, myocarditis, pulmonary infarction, pneumonia or pneumonitis, BD-related costochondritis, acute coronary syndrome (ACS), aortic dissection, tension pneumothorax, esophageal rupture, esophagitis, cholecystitis, peptic ulcer, pancreatitis, and shingles.

INVESTIGATIONS

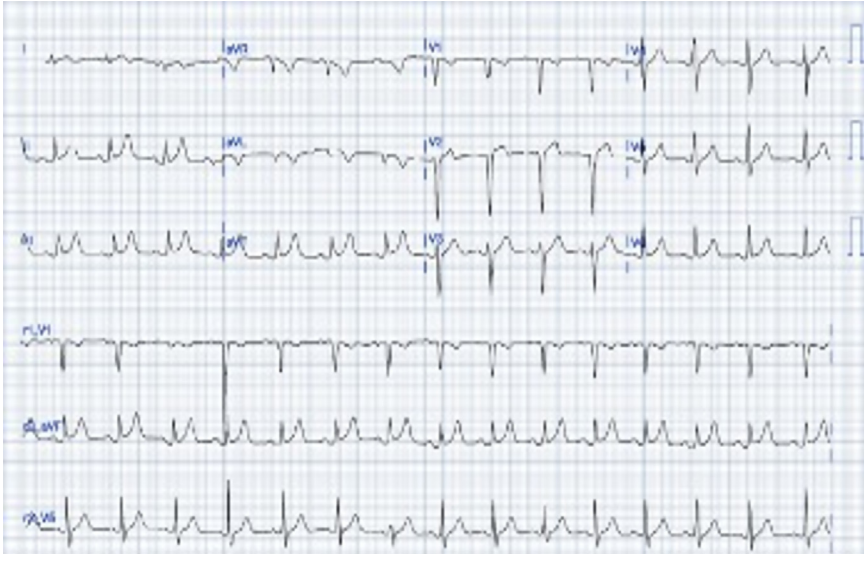
Laboratory results were significant for hemoglobin 12.3 g/dL, leukocytes 13.9 $\times$ 10^3 μL, creatinine 0.8 mg/dL, and potassium 4.5 mmol/L. C-reactive protein was 6.8 mg/dL and erythrocyte sedimentation rate was 42 mm/h; initial high-sensitivity troponin I level was 48 ng/L. Chest radiograph showed no acute cardiopulmonary process. PE protocol computed tomography angiography (CTA) chest revealed no PE, normal caliber thoracic aorta, and a small pericardial effusion.

Electrocardiogram demonstrated ST-segment elevations and hyperacute T waves in leads II, III, and aVF, suggestive of acute ischemia (Figure 1). Repeat high-sensitivity troponin I was 593 ng/L.

MANAGEMENT

The patient was loaded with aspirin 325 mg and was sent for left heart catheterization, which found a large aneurysm of the mid-left anterior descending (LAD) coronary artery measuring 4.3 $\times$ 3.5 cm, with tubular stenosis proximal to the aneurysm. There was poor flow beyond the aneurysm into the distal LAD (Figure 2). There was no evidence of extravasation of blood into the pericardial sac. The other coronary arteries were normal. The procedure was otherwise uncomplicated, and the patient remained hemodynamically stable. Subsequent coronary CTA confirmed a giant eccentric saccular coronary aneurysm arising from the mid segment of the LAD measuring 3.9 $\times$ 3.5 $\times$ 2.8 cm (Figure 3), which in retrospect, was seen in limited fashion on the PE protocol CTA. Transthoracic echocardiogram demonstrated normal left ventricular ejection fraction of 55% to 60% with no regional wall motion abnormality

FIGURE 1 Electrocardiogram With ST-Segment Elevations and Hyperacute T Waves in Leads II, III, and aVF, Suggestive of Acute Ischemia



detected. A small pericardial effusion was noted without evidence for cardiac tamponade.

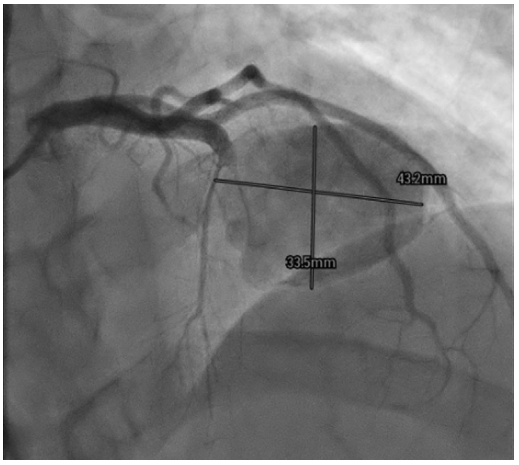
The patient was transferred to the cardiac intensive care unit of a quaternary center for monitoring and further care. Cardiothoracic surgery, vascular medicine, and rheumatology services were consulted. A therapeutic heparin infusion was administered along with aspirin 81 mg daily. Colchicine and metoprolol tartrate were continued. Laboratory testing for antinuclear and antineutrophilic antibodies were negative. After multidisciplinary discussion, the patient was deemed more appropriate for cardiac surgical management. Intraoperative findings of the LAD lesion were more consistent with pseudoaneurysm/contained rupture than a true aneurysm. The LAD was found to be fully communicating with the giant pseudoaneurysm sac, with the thrombus occluding the distal segment of the LAD. The pseudoaneurysm was opened with evacuation of contained thrombus. The proximal LAD was sutured, and left internal mammary artery to LAD bypass was performed (Figure 4). Surgical pathology of the coronary pseudoaneurysm demonstrated transmural acute inflammation, mural myxoid changes, and attached fibrin thrombus with the arterial wall. Findings were thought to be consistent with acute BD involvement (Figure 5).

FOLLOW-UP

The patient had no surgical complications. His chest pain resolved and electrocardiogram normalized. Vascular medicine and rheumatology services followed the patient during the hospital stay. He was treated with prednisone 1 mg/kg/d with instructions for outpatient taper and further follow-up with rheumatology for initiation of steroid-sparing immunosuppression. On discharge on postoperative day 5, he was also continued on aspirin 81 mg and metoprolol tartrate 25 mg twice daily given his ACS and cardiac surgery. Colchicine was also continued due to prior likely pericarditis and small effusion. Apixaban was not resumed postoperatively because rereview of his prior presentation and imaging were ultimately not diagnostic of PE, rather symptoms of the undiagnosed giant coronary pseudoaneurysm.

At 1-month follow-up with cardiology/vascular medicine, the patient was recovering well with some incisional chest pain. He was started on methotrexate therapy 10 mg/wk for 1 month. At 2-month follow-up with rheumatology, prednisone was further tapered, and the patient was started on the tumor necrosis factor-blocking agent adalimumab for active organ involvement of BD including recurrent genital ulcers.

FIGURE 2 Coronary Angiography of LAD Pseudoaneurysm

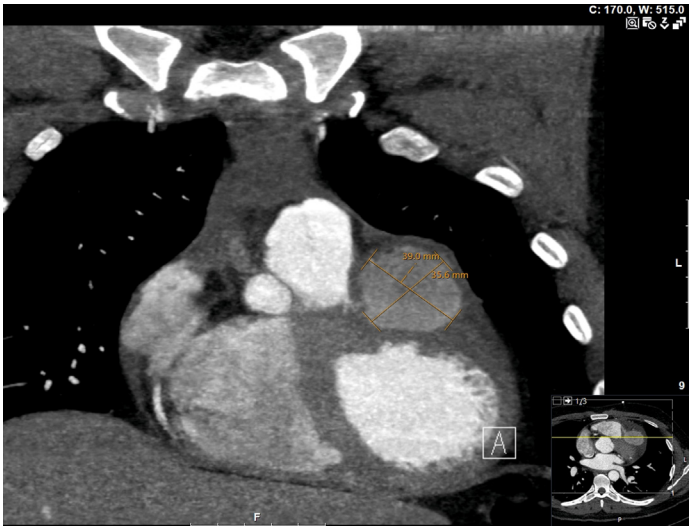


Coronary angiography demonstrating a giant pseudoaneurysm of the mid-left anterior descending (LAD) measuring 4.3 × 3.5 cm, with area of tapered LAD stenosis proximal to the aneurysm and poor flow into the distal LAD.

DISCUSSION

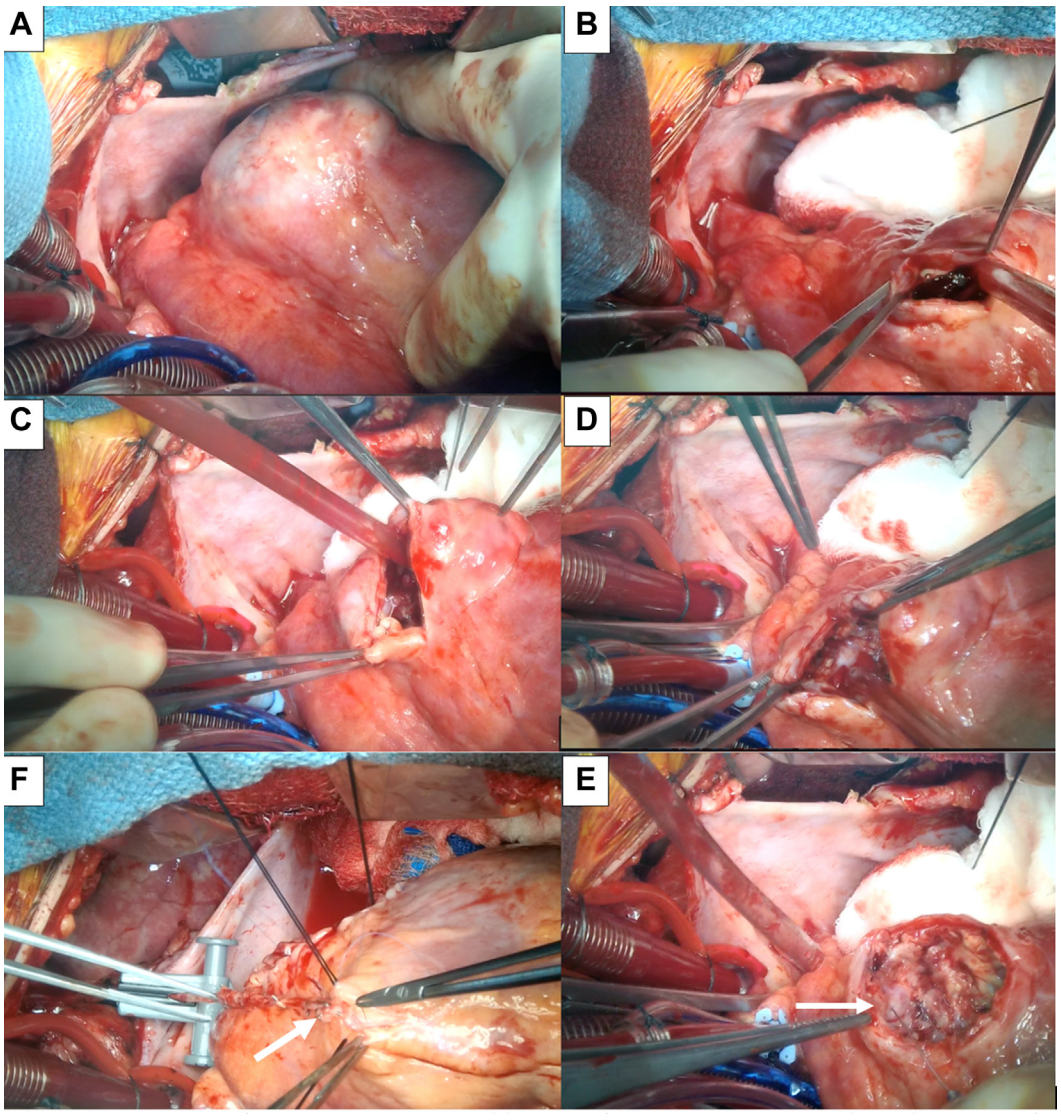
BD is a multisystem vasculitis disease that has variable clinical manifestations, including mucocutaneous, ocular, neurologic, gastrointestinal, and

FIGURE 3 Coronary CTA of LAD Pseudoaneurysm



Coronary computed tomography angiography (CTA) demonstrating large eccentric saccular pseudoaneurysm arising from the mid-segment of the left anterior descending (LAD) measuring 3.9 × 3.5 × 2.8 cm.

FIGURE 4 **Intraoperative Pseudoaneurysm Evacuation Steps**



(A) Left anterior descending (LAD) pseudoaneurysm before incision. (B to D) Resection and thrombus extraction from the pseudoaneurysm. (E) Probing of the proximal LAD (white arrow). (F) left internal mammary artery to LAD anastomosis (white arrow).

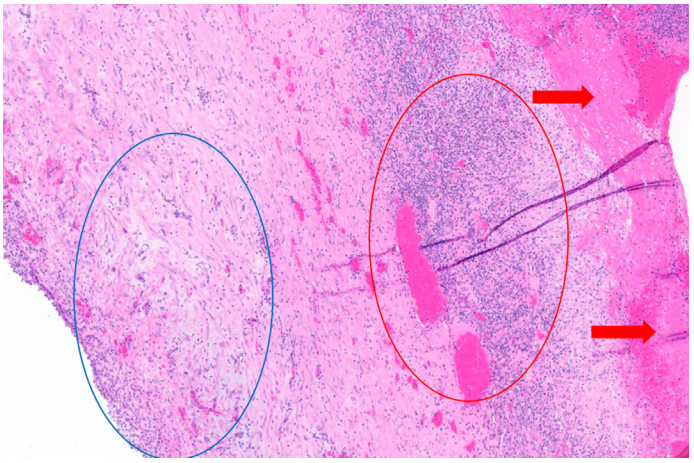
vascular lesions.[1] BD follows a relapsing and remitting nature, and treatment goals are currently focused on suppressing recurrent inflammatory recurrences to mitigate the risk of irreversible organ damage.[1] Vascular involvement in BD is associated with high mortality and is responsible for up to 43% of deaths in BD.[2] Coronary artery aneurysm is a rare complication of BD with a prevalence of 0.3% to 5%.[3,4] Even more uncommon are large coronary aneurysms (>8 mm) which have a reported prevalence of 0.02% among all patients undergoing coronary angiography.[5] Regarding coronary pseudoaneurysm in BD, the prevalence is unknown due to the rarity of this presentation and limitation of diagnostic certainty; however, it is thought that most coronary aneurysms in BD are in fact pseudoaneurysms.[4] In one systematic review of 62 cases of patients with BD presenting with ACS between 1980 and

2018, ST-segment elevation myocardial infarction was identified in 48%, and 6% (n = 3) were found to have a coronary pseudoaneurysm.[6]

Medical treatment with immunosuppressive therapy is the cornerstone of management of BD with vascular involvement.[1] For arterial involvement in BD, current immunosuppressive agent recommendations include high-dose glucocorticoids, cyclophosphamide, and methotrexate. However, in the setting of myocardial infarction or cardiac surgery, timing of immunosuppressive therapy is controversial due to the therapy's interference with myocardial healing and an increased the risk of wall rupture. Current recommendations are to administer immunosuppressive medications 2 to 3 days post-ACS presentation to balance the risk-benefit ratio associated with cardiovascular manifestations of BD.[7,8] In this patient, oral prednisone therapy was started with plans to taper and ultimately start steroid-sparing immunosuppression in the outpatient setting with frequent monitoring. At the 1-month follow-up appointment, methotrexate was added to the patient's regimen. At 2-month follow-up, the patient was having recurrent genital ulcers despite colchicine therapy, and thus was started on adalimumab therapy with further prednisone tapering. This was in accordance with current guidelines for patients with recurrent ulcers refractory to colchicine, that immunomodulatory agents such as azathioprine or tumor necrosis alpha inhibitors (tumor necrosis factor inhibitors) such as adalimumab, can be attempted.[9,10]

The current guidelines for BD are adopted from the European League Against Rheumatism (EULAR), which suggests against delaying surgery or stenting for symptomatic patients with aortic involvement.[1] Given that this recommendation is not specific for coronary artery involvement, and the scarcity of data on the optimal management of giant coronary aneurysms/pseudoaneurysms in patients with BD, the decision regarding modality and timing of intervention is an individualized one. Cases in the literature have pursued immunotherapy and immunosuppression first, followed by surgery after 1 month, whereas others did not pursue immunosuppression after surgery due to concerns of poor wound healing.[11-13] The present case highlights the use of corticosteroids in the postoperative period because this practice remains a topic of debate. Small studies on arterial lesions in patients with BD have shown decreased arterial relapse in patients with aneurysms treated with high-dose corticosteroids postoperatively.[14] The present case of urgent surgical revascularization followed by high-dose corticosteroids during the same admission demonstrated favorable outcomes and no recurrence at 6-month follow-up.

FIGURE 5 Histopathology



H&E 4× magnification. Full thickness section of the left anterior descending arterial wall showing the attached thrombus (red arrows), inflammation in the wall (red circle), and myxoid changes (blue circle).

The paucity of data, including clinical trials, comparing surgical repair and revascularization vs percutaneous coronary interventions (PCIs) for coronary artery aneurysms is a challenge. Most of what is currently known regarding PCI in such patients is obtained from limited case reports and small case series of patients presenting with ACS, primarily among patients with atherosclerotic-related coronary artery aneurysms or Kawasaki disease.[7,15,16] However, both interventional and surgical strategies are technically challenging, and multiple factors play into the decision-making of optimal management, including the severity of symptoms and the presence of high-risk anatomic features (eg, size of aneurysm, involvement of major side branches of the coronary arteries).[7] The current recommendation for giant coronary aneurysms and pseudoaneurysms (>20 mm) is cardiac surgical intervention as a first-line management strategy.[7] In addition, multiple studies have suggested inferior PCI outcomes, including higher incidence of no-reflow, distal embolization of thrombus, and higher mortality.[17-19] In the present case, multidisciplinary discussion was undertaken regarding management of the LAD aneurysm. Given the patient's young age, giant aneurysm size, and favorable anatomy, a surgical approach was pursued.

CONCLUSIONS

Coronary pseudoaneurysm is a very rare complication of BD. This case emphasizes the importance of early suspicion of coronary artery aneurysm/pseudoaneurysm and early incorporation of coronary imaging into the diagnostic process. Coronary pseudoaneurysms have significant risk of thromboembolism and rupture and are associated with devastating outcomes, including acute myocardial infarction. Although there are currently no clear guidelines for management of patients with coronary pseudoaneurysms associated with BD, coronary artery bypass grafting along with immunosuppressive therapy remains the most common approach. Multidisciplinary involvement to determine an individualized treatment plan is critical in these cases.

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