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MINI-FOCUS ISSUE ON CORONARY, PERIPHERAL, AND STRUCTURAL INTERVENTIONS

CLINICAL CASE

Left Main Coronary Artery Ostial Stenosis Caused by Syphilitic Aortitis Presenting With Syncope

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ABSTRACT

A 51-year-old man with a history of antibiotic therapy for syphilis 1 month ago presented with syncope. Computed tomography revealed circumferential aortic wall thickening complicating severe stenosis of left main coronary ostium. Abnormalities in serologic and cerebrospinal fluid tests led to the diagnosis of syphilitic aortitis and neurosyphilis. Coronary angiography demonstrated the severe stenosis of left main coronary artery ostium, and cardiac magnetic resonance imaging showed subendocardial late gadolinium enhancement involving basal to mid anteroseptal wall of the left ventricle. He was successfully treated with coronary artery bypass grafting with bilateral internal thoracic artery grafts. The postoperative course was uneventful, with the computed tomography scan showing that all grafts were patent. He was discharged without any symptoms on the 10th postoperative day. (JACC Case Rep. 2025;30:102720)

HISTORY OF PRESENTATION

A 51-year-old male patient was referred to the emergency department for syncope. The electrocardiogram showed sinus rhythm and ST-segment depression in leads I, aVL, and V_4 to V_6 (**Figure 1A**). Chest x-ray showed no cardiomegaly or pulmonary congestion (**Figure 1B**). The troponin I level was 0.054 ng/mL, N-terminal pro-brain natriuretic peptide level was 415 pg/mL. Transthoracic echocardiography showed no regional wall motion abnormalities with left ventricular ejection fraction of 58% and mild hypertrophy with wall thickness of 10 mm (**Figures 1C and 1D**). After the initial evaluation, we considered that cardiac computed tomography (CT) to rule out the coronary artery disease and magnetic resonance imaging (MRI) to evaluate cardiomyopathy were necessary in addition to the continuous electrocardiographic monitoring. The cardiac CT demonstrated circumferential

TAKE-HOME MESSAGES

- In case of coronary ostial stenosis without significant lesions in the distal vascular bed, especially in young patients without coronary risk factors, aortitis should be suspected.
- Although tertiary syphilis is rare in the antibiotic era, syphilitic aortitis should still be considered as a potential cause of coronary ostial stenosis.

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ABBREVIATIONS AND ACRONYMS

CABG = coronary artery bypass grafting

CT = computed tomography

MRI = magnetic resonance imaging

RPR = rapid plasma reagin

ascending aortic wall thickening with intramural contrast enhancement involving left main coronary ostium and severe stenosis of left main coronary ostium (Figure 2), which raised the need for further investigation for aortitis.

PAST MEDICAL HISTORY

One month before the visit, a screening test before the fertility treatment revealed the positive results of nontreponemal (rapid plasma reagin [RPR]) and treponemal (Treponema pallidum hemagglutination assay) tests. The antibiotic therapy with amoxicillin 1,500 mg/day for 4 weeks was initiated as a primary syphilis; however, serum RPR remained reactive in high titer.

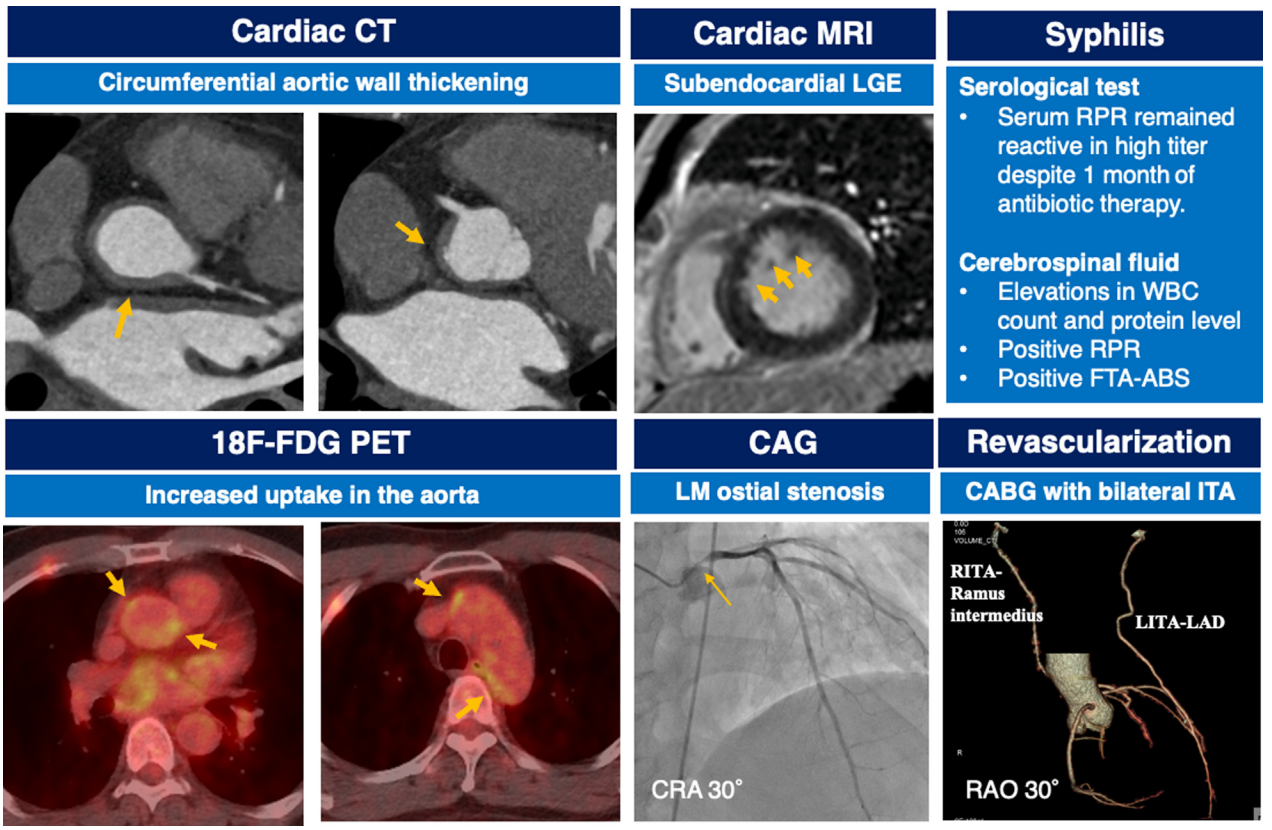
DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes large vessel vasculitis such as Takayasu arteritis, giant cell arteritis, immunoglobulin (Ig)G4-related aortitis, vascular Behcet's disease, or infectious aortitis. Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis, lupus vasculitis, or rheumatoid vasculitis also should be excluded.

INVESTIGATION

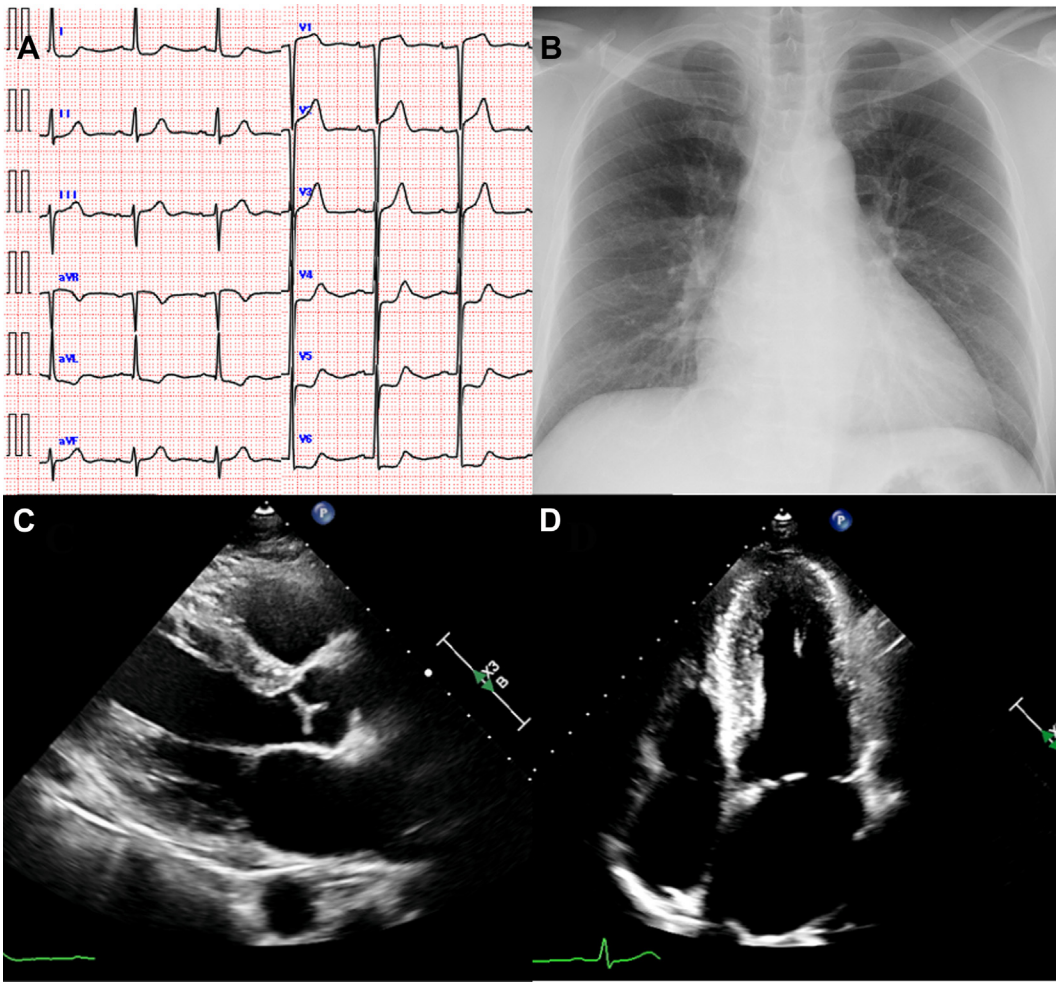
Thoracic MRI showed a contrast enhancement in the thickened aortic wall over the ascending aorta to the thoracic descending aorta, and fluorine-18 fluorodeoxyglucose positron emission tomography revealed an increased uptake in the ascending aorta and aortic arch (Figures 3A and 3B). There were no

VISUAL SUMMARY **Left Main Coronary Artery Ostial Stenosis Caused by Syphilitic Aortitis Presenting With Syncope**



18F-FDG PET = fluorine-18 fluorodeoxyglucose positron emission tomography; CABG = coronary artery bypass grafting; CAG = coronary angiography; CRA = cranial; CT = computed tomography; FTA-ABS = fluorescent Treponema antibody absorption; ITA = internal thoracic artery; LAD = left anterior descending artery; LGE = late gadolinium enhancement; LITA = left internal thoracic artery; LM = left main coronary; MRI = magnetic resonance imaging; RAO = right anterior oblique; RITA = right internal thoracic artery; RPR = rapid plasma reagin; WBC = white blood cell.

FIGURE 1 **Findings on Initial Evaluation**



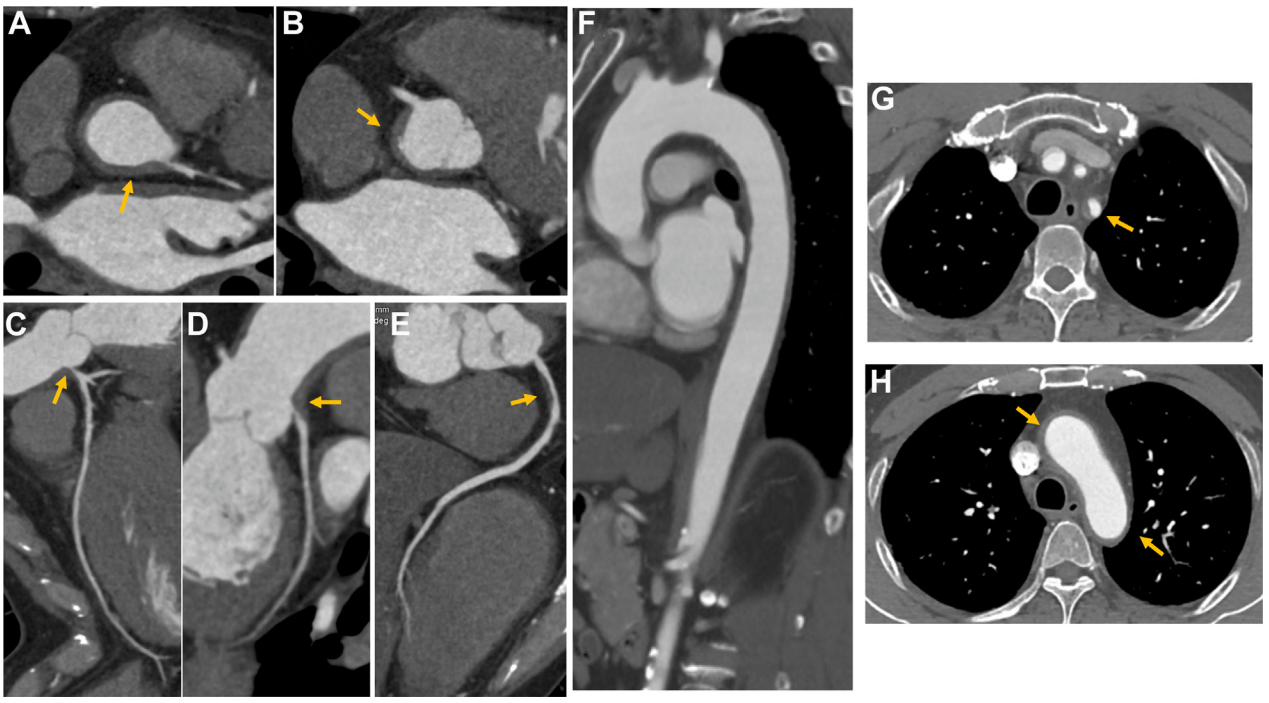
(A) ST-segment depression in leads I, aVL, V_4 to V_6. (B) No enlarged heart and no pulmonary congestion. (C) Echocardiography showed left ventricular ejection fraction of 58%, with mild hypertrophy with circumferential wall thickness of 10 mm.

findings suggestive of Takayasu aortitis, Behcet's disease, ANCA-associated vasculitis, IgG-4-related aortitis, or giant cell arteritis. A cerebrospinal fluid test revealed elevations in white-cell count (15/μL), protein level (87 mg/dL), and reactive results in fluorescent Treponema antibody absorption and RPR tests, leading to the diagnosis of neurosyphilis. Based on these results, the patient was diagnosed with syphilitic aortitis. Invasive coronary angiography was performed, confirming the severe stenosis of left main coronary artery ostium that was wedged by 4-F catheter (**Figure 4**). Cardiac MRI showed subendocardial late gadolinium enhancement involving basal to mid anteroseptal wall of the left ventricle, which, taken together with the severe stenosis of left main coronary ostium and ST-segment depression, suggested cardiac ischemia-related syncope[1] (**Figure 3D**).

MANAGEMENT

Treatment with intravenous aqueous crystalline penicillin G at a dosage of 2.4 million units per day for 14 days was started. Jarisch-Herxheimer reaction was not observed. After confirming no ostial stenosis in both internal thoracic arteries on contrast-enhanced CT (Supplemental Figure 1), we chose coronary artery bypass grafting (CABG) with bilateral internal

FIGURE 2 Contrast-Enhanced Computed Tomography



(A and B) Circumferential ascending aortic wall thickening (arrows) with intramural contrast enhancement involving left main coronary ostium and severe stenosis of left main coronary ostium. (C) Severe stenosis in the left main coronary ostium (arrow). There is mild plaque in the proximal left anterior descending artery. (D) Severe stenosis in the left main coronary ostium (arrow) without any stenosis in the left circumflex artery. (E) Moderate stenosis in the proximal right coronary artery (arrow), consistent with atherosclerotic changes. (F) Circumferential wall thickening was also found in the descending thoracic aorta. (G) Mild stenosis with plaque in the ostium of the left subclavian artery (arrow). (H) Circumferential wall thickening in the aortic arch (arrows).

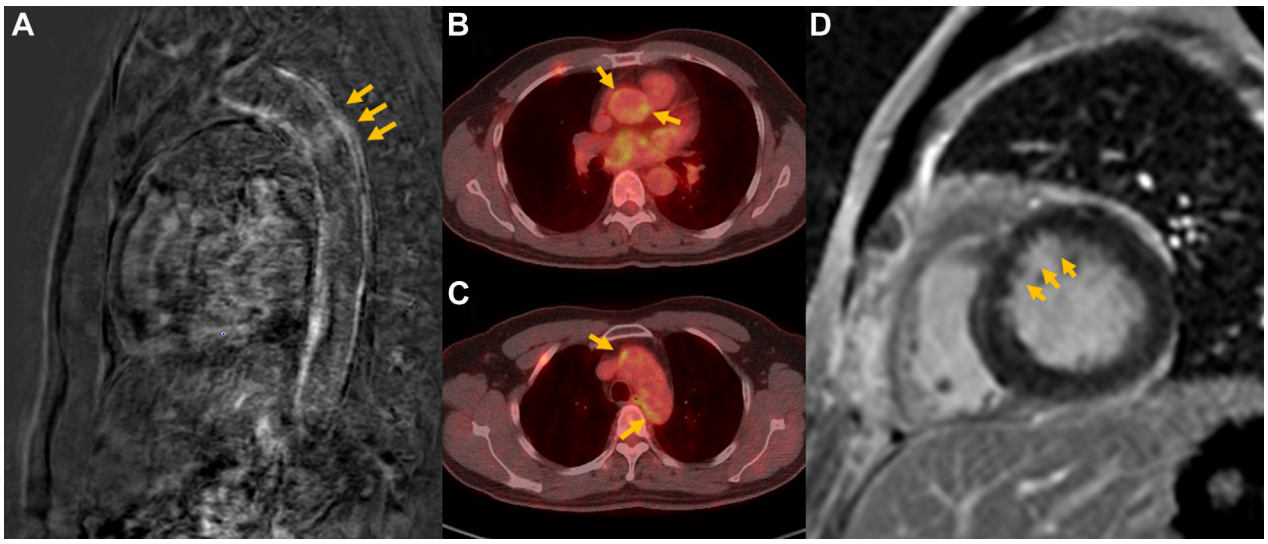
thoracic artery grafts over percutaneous coronary intervention (PCI) as a revascularization strategy. Off-pump CABG was successfully performed, with a left internal thoracic artery graft to the left anterior descending artery and a right internal thoracic artery graft to the ramus intermedius. A heart stabilizer was used to reduce the cardiac surface motion, and the coronary arteries were snared with a vessel loop without an intracoronary shunt during the anastomosis. Intraoperative graft flow measurements with ultrasonic transit-time flowmeter showed good results, with a mean flow of 16 mL/min, a pulsatility index of 1.9, and a diastolic filling index of 83% for the left internal thoracic artery graft to the left anterior descending artery, and a mean flow of 31 mL/min, a pulsatility index of 1.4, and a diastolic filling index of 76% for the right internal thoracic artery graft to the ramus intermedius. There were no surgical indications for procedures that would have allowed us to obtain a sample of the aortic wall. Considering the risk of pseudoaneurysm formation, we did not obtain a sample of the aortic wall for histopathological evaluation. Histopathological examination of distal parts of bilateral internal thoracic arteries showed no inflammatory changes or fibrous intimal thickening. Intraoperative transesophageal echocardiography further confirmed the circumferential ascending aortic wall thickening (**Figure 5**).

DISCUSSION

Syphilis is a systemic disease caused by *Treponema pallidum*. Tertiary syphilis can present with cardiovascular syphilis, gummatous lesions, tabes dorsalis, and general paresis. In all individuals who have tertiary syphilis or whose serum RPR remained reactive despite appropriate treatment, a concomitant neurosyphilis should be suspected.

Approximately one-third of patients with untreated syphilis develop tertiary syphilis.[2] Cardiovascular syphilis classically presents as aortitis resulting in aortic aneurysm, aortic valvulitis with

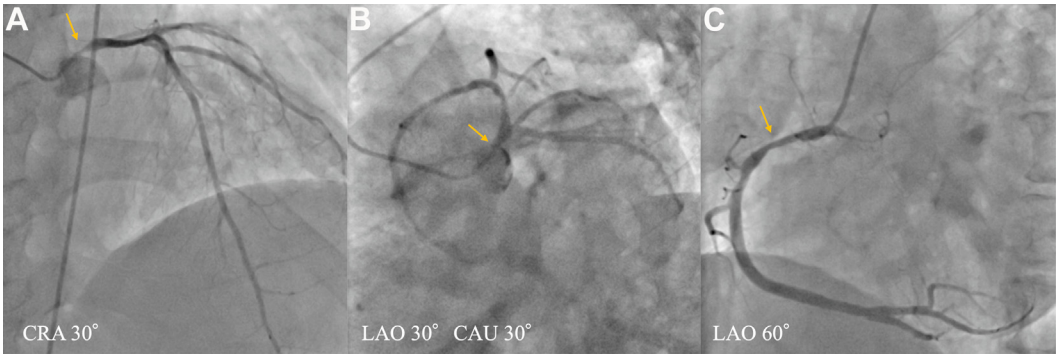
FIGURE 3 **Thoracic and Cardiac Magnetic Resonance Imaging and Fluorine-18 Fluorodeoxyglucose Positron Emission Tomography**



(A) Contrast enhancement in the thickened aortic wall over the ascending aorta to the thoracic descending aorta (arrows). (B) An increased uptake in the thickened aortic wall in the ascending aorta (arrows). (C) An increased uptake in the thickened aortic wall in the aortic arch (arrows). (D) Subendocardial late gadolinium enhancement in the basal to mid anteroseptal wall of the left ventricle (arrows).

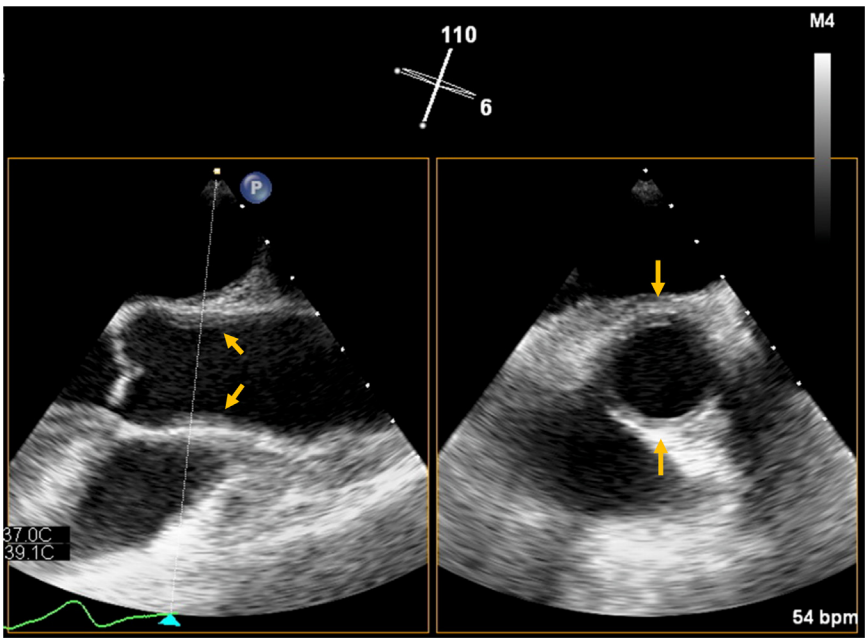
insufficiency, and coronary ostial stenosis. An autopsy study of 100 patients with sysphilic aortitis reported that coronary ostial stenosis was found in 26 patients.[3] This narrowing was the result of extension of the syphilitic process in the ascending aorta to the orifices of the coronary arteries. A more recent autopsy study revealed more frequent involvement of right coronary artery in 15% of patients, and the left coronary artery in 2%.[4] Aortic regurgitation can develop due to either aortic root dilatation or syphilitic involvement of aortic valve, accounting for 20% to 30% of patients with syphilitic aortitis. Aortic aneurysm occurs in 5% to 10% of patients with syphilitic aortitis, most often in the ascending aorta, and predominantly in a saccular rather than fusiform pattern.[5,6] Severe aortic regurgitation and aortic aneurysm require surgical intervention for definitive therapy.

FIGURE 4 **Coronary Angiography**



(A and B) Severe stenosis of the left main coronary artery ostium (arrows). A 4-F catheter was placed near the ostium because the ostium was wedged with catheter engagement. (C) Right coronary artery with 50% stenosis in the proximal portion (arrow).

FIGURE 5 Intraoperative Transesophageal Echocardiography



The circumferential ascending aortic wall thickening (arrows) was observed.

The optimal revascularization strategy for syphilitic coronary ostial stenosis has not been clearly elucidated.[7] An observational study including 60 patients with syphilitic coronary ostial stenosis demonstrated that patients treated with CABG had lower incidence of adverse cardiac events. In addition, the use of the left internal thoracic artery graft was associated with a lower restenosis rate compared with the saphenous vein graft, because the persistent infection of the ascending aorta can lead to the proximal anastomotic restenosis.[8] The higher rate of restenosis after PCI may be related to inflammatory etiology and inadequate expansion due to the smooth muscle of the aorta. In patients with Takayasu arteritis, a lower restenosis rate in CABG compared with PCI was reported,[9] despite the reduced availability of internal thoracic artery grafts due to the involvement of the ostial lesions of innominate and subclavian arteries, whereas syphilitic aortitis does not typically involve the main branch of the aortic arch.[3,4,10] On the other hand, PCI may be a viable option in patients with hemodynamic instability or high surgical risk.[7]

In the case of coronary ostial stenosis without stenosis in the distal vascular bed, especially when patients are young and lack coronary risk factors, aortitis should be suspected. Although tertiary syphilis is rare in the antibiotic era, syphilitic aortitis still needs to be considered as a cause of coronary ostial stenosis.

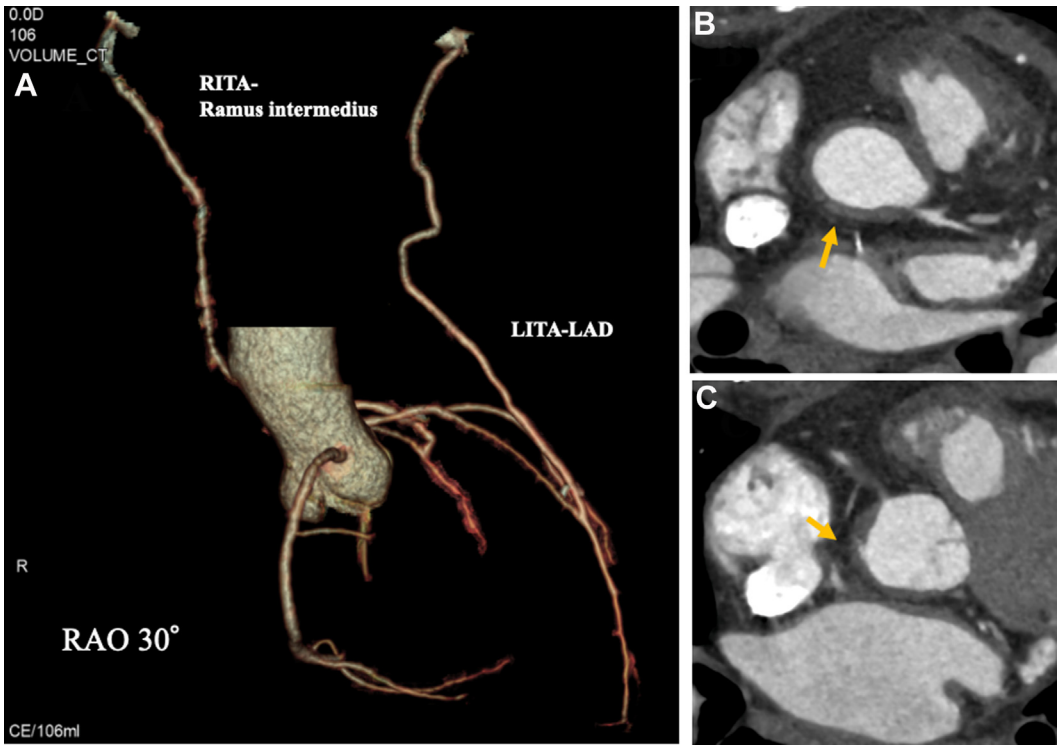
FOLLOW-UP

The postoperative course after CABG was uneventful except for the postoperative atrial fibrillation, which was successfully treated with intravenous amiodarone. The CT scan performed on the seventh postoperative day showed that all grafts were patent, with circumferential ascending aortic wall thickening still present (**Figure 6**). After completing a 14-day course of intravenous penicillin G, a single intramuscular injection of 2.4 million units of benzathine penicillin G was administered. He was discharged without any symptoms on the 10th postoperative day. A decrease in serum RPR titer was observed 3 months after penicillin G therapy.

CONCLUSIONS

A 51-year-old man was successfully treated with CABG for left coronary ostial stenosis caused by syphilic aortitis and definitive antibiotic therapy for neurosyphilis. Our case highlights the importance for the awareness regarding neurosyphilis as a cause of

FIGURE 6 Cardiac Computed Tomography After Successful Coronary Artery Bypass Grafting



(A) A patent left internal thoracic artery graft to the LAD and a patent right internal thoracic artery graft to the ramus intermedius. (B and C) Circumferential ascending aortic wall thickening (arrows) involving left main coronary ostium was still present. LAD = left anterior descending artery; LITA = left internal thoracic artery; RITA = right internal thoracic artery; RAO = right anterior oblique.

treatment failure, and syphilitic aortitis as a cause of coronary artery ostial lesions.

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KEY WORDS coronary artery bypass grafting, coronary ostial stenosis, computed tomography, syphilitic aortitis

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