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ISCHEMIC HEART DISEASE

CLINICAL CASE

Managing Complex Coronary Revascularization of Acute Coronary Syndrome During Cogan Syndrome

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ABSTRACT

Cogan syndrome, a rare multisystemic autoimmune vasculitis, can result in coronaritis and precipitate acute coronary syndrome. This paper reports a case of a young woman affected by Cogan syndrome with acute coronary syndrome due to a left main lesion and the complexities in selecting the revascularization strategy for concomitant active vasculitis. (JACC Case Rep. 2025;30:103080) © 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 33-year-old woman presented to the emergency department due to fatigue, dyspnea, and worsening oppressive chest pain during the days before. On admission, she was in pulmonary edema requiring intravenous diuretics and noninvasive mechanical ventilation. Electrocardiography showed aVR ST-T-segment elevation with diffuse ST-T-segment depression (Figure 1).

TAKE-HOME MESSAGES

- Cogan syndrome may cause coronaritis, which may precipitate acute coronary syndromes.
- Shared decision-making and multidisciplinary approach are essential for customizing coronary revascularization during acute coronary syndromes with active vasculitis. In particular, the intensity and extent of the vasculitis process must be considered.

PAST MEDICAL HISTORY

The patient had no known cardiovascular risk factors but a history of Cogan syndrome (CS) with neurosensorial deafness requiring a cochlear implant and previous eye scleritis. She was in chronic treatment with prednisone and methotrexate, after a failed attempt at treatment with adalimumab (ineffective), and the withdrawal of infliximab after acute infective salpingitis.

Two weeks before the presentation, she accessed the emergency department due to an episode of less intense chest pain. Clinical examination,

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ABBREVIATIONS AND ACRONYMS

ACS = acute coronary syndrome

CS = Cogan syndrome

CTA = computed tomography angiography

SCA = selective coronary angiography

electrocardiogram, 2-point high-sensitivity cardiac troponin T, and echocardiogram were negative. A computed tomography angiography (CTA) of the aorta and a contrast-enhanced computed tomography of the chest and abdomen showed a normal aorta with regular walls and signs compatible with past flares of vasculitis (right renal and superior mesenteric artery stenosis with celiac trunk occlusion, filled by collaterals from the inferior mesenteric artery); therefore, the patient was discharged.

DIFFERENTIAL DIAGNOSIS

Cardiac chest pain in young women mandates the exclusion of unusual causes of acute coronary syndrome (ACS) such as spontaneous coronary dissection and the differential diagnosis with acute pulmonary embolism, Takotsubo syndrome, myocarditis, and acute aortic dissection. Her history of CS facilitated diagnosis.

INVESTIGATIONS

A noncardiosynchronized CTA of the aorta showed de novo remarkable thickening of the distal aortic arch wall and thoracic descending aorta with ostial stenosis of the left subclavian artery. Venous and late contrast enhancement phases suggested active vasculitis (**Figures 2 and 3**). Both coronary ostia could not be visualized.

A single bolus of 1 g methylprednisolone was administered. An urgent selective coronary angiography (SCA) was carried out (right radial approach), demonstrating a subocclusive (99%) ostial stenosis of the left main trunk with collaterals from the right coronary artery supplying the anterior descending artery (Rentrop grade III) (**Figure 4**, Videos 1 to 3).

MANAGEMENT

After a collegial discussion between the cardiac surgeon, interventional cardiologist, and cardiac intensivist, percutaneous angioplasty was performed with a single drug-eluting stent implanted.

Subsequently, the patient was transferred to the intensive cardiac care unit. She remained hemodynamically stable and asymptomatic during the hospitalization, with a peak high-sensitivity troponin T of 1447 pg/mL. On admission, her left ventricular ejection fraction was reduced to 38% with apical akinesia (Videos 4 and 5); both recovered on the ninth day echocardiogram (Videos 6 and 7). Blood test revealed leukocytosis (14.30 $\times$ 10^9/L) and absolute neutrophilia (13.68 $\times$ 10^9/L), a C reactive protein of 10 mg/L (reference limit: <5), and an erythrocyte sedimentation rate of 53 mm/h (reference limit: 2-30 mm/h). Serum low-density lipoprotein cholesterol was 92 mg/dL and lipoprotein-a was 20 mg/L. Viral screening and autoimmune serology yielded negative results. Intravenous methylprednisolone 1 g was administered once daily for 3 days and then

FIGURE 1 Admission Electrocardiogram: Sinus Tachycardia, aVR ST-T-Segment Elevation, and Diffuse ST-T-Segment Depression

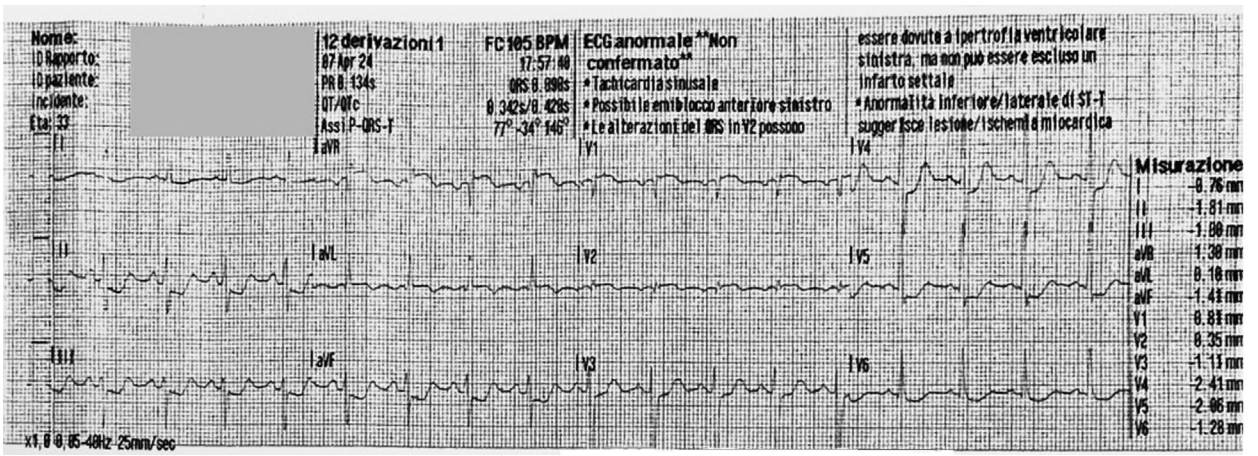
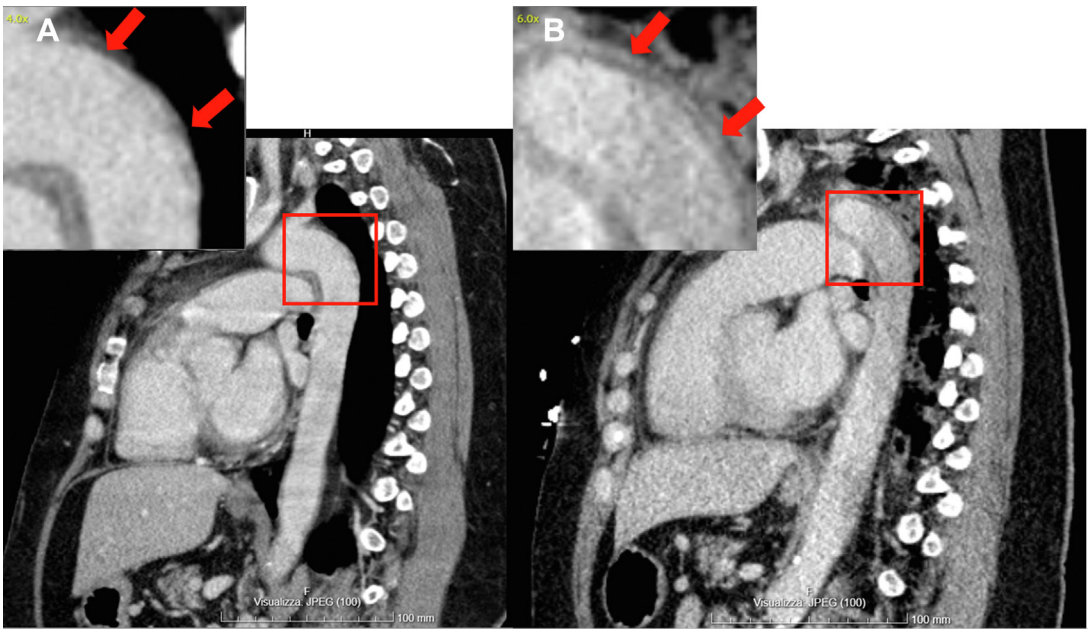


FIGURE 2 **Aortic Computed Tomography Angiography 1**

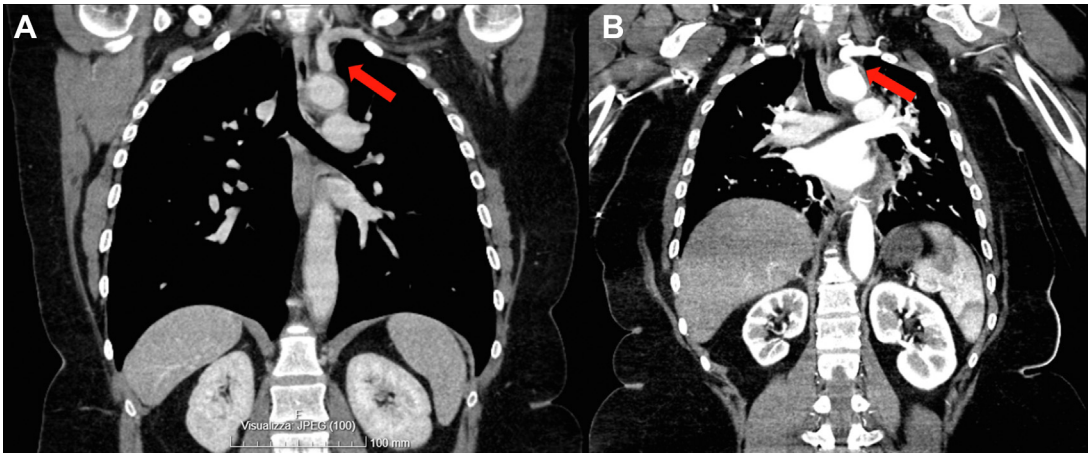


Irregular and thickened aortic wall with contrast enhancement compatible with active aortitis (B, red arrows), absent on the examination performed during the first emergency department access (A, red arrows).

de-escalated to 250 mg by mouth. On the fourth day of steroid therapy, C-reactive protein was <5 mg/L. Two days later, a total body fluorodeoxyglucose positron emission tomography scan revealed the absence of remarkable glucose uptake and metabolism within the aorta and its branches (Figure 5).

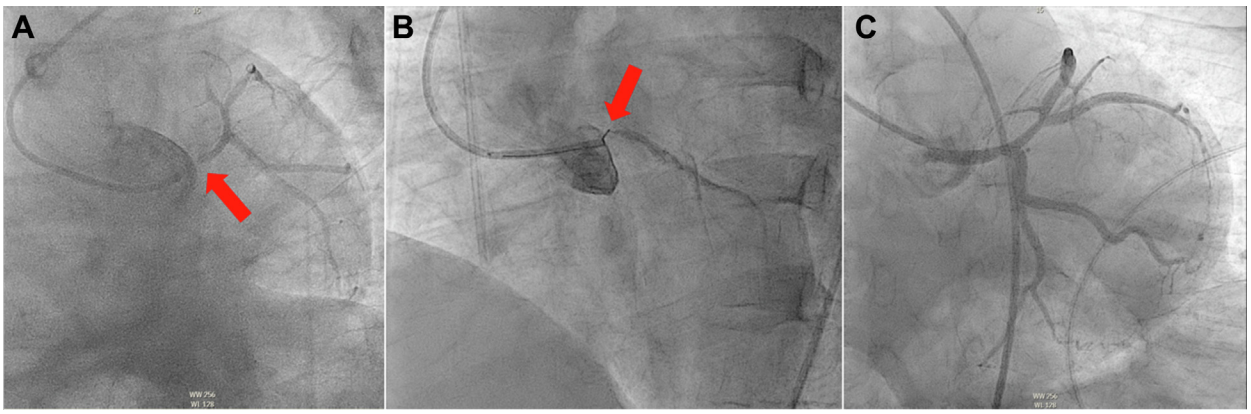
A nonconditional cochlear implant contraindicated cardiac magnetic resonance.

FIGURE 3 **Aortic Computed Tomography Angiography 2**



Left subclavian ostial stenosis (B), absent on the examination performed during the first emergency department access (A).

FIGURE 4 Admission Selective Coronary Angiography



(A and B) Subocclusion of left main trunk. (C) Final angiographic result after drug-eluting stent implant.

The patient was discharged after 10 days of hospitalization, with dual antiplatelet therapy (ie, acetylsalicylic acid and ticagrelor 90 mg twice daily) and prednisone 75 mg once daily.

FOLLOW-UP

One month later, cyclophosphamide 15 mg/kg was started after ovary preservation with enantone. Immunomodulating therapy was well tolerated (Video 8), with no vasculitis or chest pain recurrences and persistently negative inflammatory markers.

A repeated coronary angiogram 2 months after discharge demonstrated a persistently good outcome of coronary revascularization without in-stent restenosis (**Figure 6**, Videos 9 and 10).

DISCUSSION

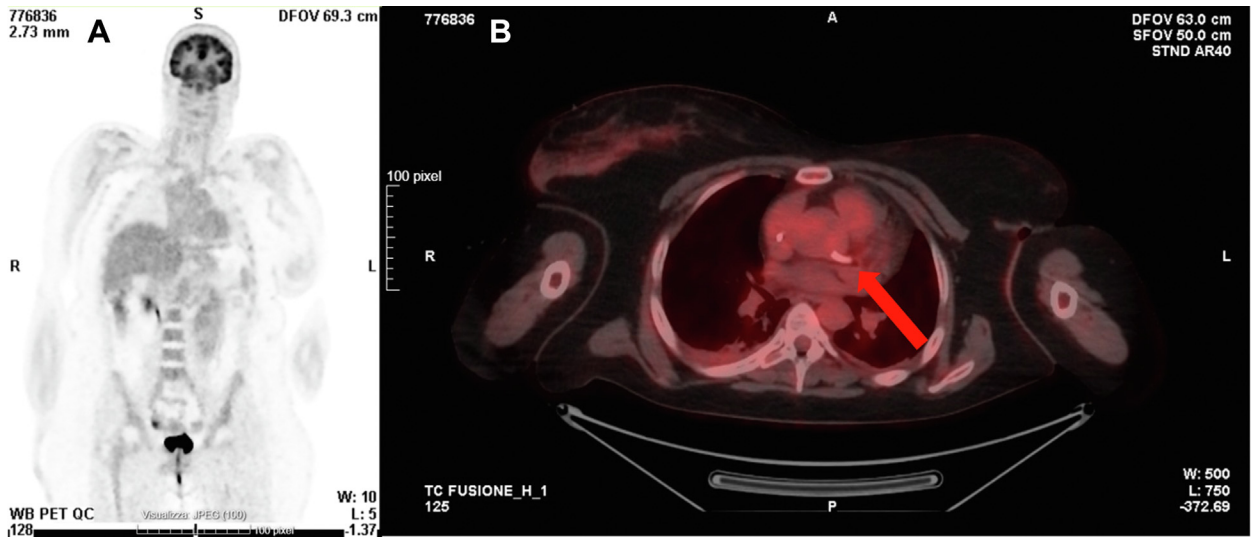
Ophthalmologic inflammation and audiovestibular dysfunction are the main manifestations of CS, a rare multisystemic autoimmune vasculitis typically affecting young adults.[1] Systemic disease is observed in 15% to 20% of patients; in particular, cardiovascular involvement may occur as aortitis (up to 10% of patients),[2] and rarely, as pericarditis, myocarditis, and coronaritis.[1] During coronaritis, autoantibodies and proinflammatory cytokines (interferon-γ, tumor necrosis factor-α, Th-1 interleukins) induce vessel wall inflammation, possibly leading to stenosis or occlusion of the ostia/proximal segments.[3,4] Therefore, ACS and myocardial infarction may occur in patients with CS; however, few instances are reported.

Chest pain in young women can be challenging to deal with. Although ACS with heart failure and electrocardiographic signs of left main lesion mandates urgent/emergent catheterization, aortic CTA was first performed to exclude aortic dissections due to the CS history. Subsequently, SCA confirmed a left main subocclusive stenosis. Chronic inflammation during systemic vasculitis is a known determinant of coronary injuries.[3] Indeed, the patient's young age and absence of cardiovascular risk factors made atherosclerosis an unlikely primary cause of the left main lesion. The fluorodeoxyglucose positron emission tomography showed no notable coronary inflammation, but this test was only performed after steroid-induced immunosuppression to confirm disease regression.

Cardiac complications affect the prognosis of patients with CS,[4] and their young age at presentation may pose significant treatment challenges. Percutaneous revascularization is controversial in vasculitis due to the higher risk of in-stent restenosis[5]; however, severe aortic and arch vessel inflammation may complicate the aortic anastomosis during coronary artery bypass graft,[6] and flow in the mammary arteries can be jeopardized, as in this case, by the development of ostial disease of the neck vessels.

Recently, the case of a 25-year-old woman with a CS-precipitated acute anterior myocardial infarction was published reporting effective treatment with a triple-vessel coronary artery bypass graft due to a large aneurysm of the left main and right coronary artery and proximal occlusion of the left anterior descending artery.[7] However, concerns regarding venous graft occlusion in patients with CS have been raised.[8] Conversely, percutaneous revascularization in CS-precipitated ACS has never been described,

FIGURE 5 Total Body Fluorodeoxyglucose Positron Emission Tomography Scan



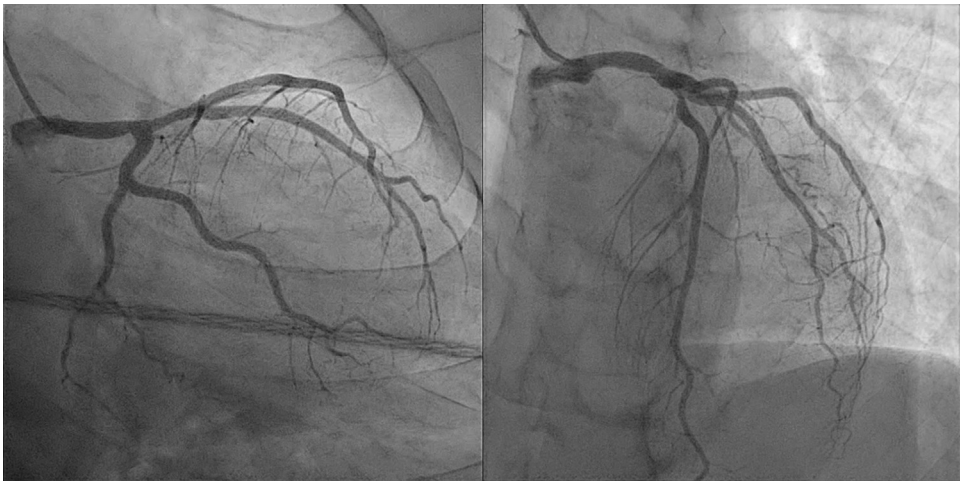
(A) No remarkable aortic glucose uptake. (B) Fluorodeoxyglucose positron emission tomography/low-dose computed tomography fused image: the stent implanted (arrow) not surrounded by enhanced coronary glucose metabolism.

with 1 case only of percutaneous intervention in a Korean patient with CS and stable angina.[9]

This patient exhibited rapid and intense aortic inflammation over 2 weeks and during anti-inflammatory maintenance therapy. Appropriate escalation of the anti-inflammatory treatment on top of the cardiovascular therapy is mandatory in this setting to prevent re-exacerbation and disease progression; indeed, this patient's therapy was potentiated by introducing cyclophosphamide after high-dose steroids with no subsequent recurrences.

One limitation of this case is the lack of intracoronary imaging during both the primary percutaneous coronary intervention and control SCA.

FIGURE 6 Follow-Up Selective Coronary Angiography



Absence of in-stent restenosis.

However, the left main lesion did not involve a bifurcation and its use would have prolonged the procedure, increasing the risk of complications in a patient on the brink of hemodynamic instability.

CONCLUSIONS

ACS in CS coronaritis is rare, and percutaneous revascularization in this setting has never been described. This case highlights the treatment challenges of a patient with CS with active inflammation of the aorta and its branches, underlying the pivotal role of shared decision-making and a multidisciplinary approach to ascertain the diagnosis, decide an optimal revascularization strategy, and perform an aggressive individualized anti-inflammatory and immunomodulatory treatment according to the patient's clinical presentation and response.

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KEY WORDS acute coronary syndrome, Cogan syndrome, coronaritis, percutaneous coronary intervention

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