

Recurrent myocardial infarction due to coronary artery aneurysm in Behçet's syndrome: a case report

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Background

Behçet's syndrome (BS) is a multisystemic vasculitis of unknown aetiology. Coronary involvement is extremely uncommon in BS, with a reported prevalence of 0.5%. The mortality of cardiovascular manifestation in BS is 20%, while there are still considerable challenges in the management of these patients.

Case summary

We report the case of a 30-year-old young man with a 12-year history of BS complicated by left anterior descending coronary (LAD) aneurysms, stenosis, and recurrent myocardial infarction. Percutaneous coronary intervention treated the stenosis and aneurysms of the LAD, but the coronary angiography clearly revealed an acute stent thrombosis after 7 months. This unusual coronary complication of BS treated successfully is presented.

Discussion

Our case demonstrated the feasibility of covered stent, immunosuppressive therapy, and escalated antithrombotic regimen in the treatment of BS patients with coronary artery aneurysm (CAA). We also demonstrated that drug-eluting stents may aggravate aneurysmal dilatation of the CAA under inefficient immunosuppressive therapy.

Keywords

Behçet's disease • Coronary arteritis • Coronary artery aneurysm • Case report

Learning points

- Behçet's syndrome (BS) is a systemic vasculitis, with coronary artery disease extremely uncommon, and there are enormous challenges in the management of BS patients with coronary artery aneurysm (CAA).
- Behçet's syndrome patients with CAA might benefit from covered stent implantation, anticoagulation, and immunosuppressive therapy.
- Drug-eluting stents (DESs) may aggravate vascular lesions in these patients, especially in the absence of immunosuppressive therapy. Accordingly, traditional DESs may be avoided in BS patients.

Introduction

First described in 1937, Behçet's syndrome (BS) is a systemic vasculitis with unknown aetiology and characterized by recurrent oral ulcers (OUs), genital ulcers (GUs), and ocular lesions as well as papulopustular, gastrointestinal, nervous system, and vascular involvement.¹ Among them, vascular and nervous system involvement are the most common causes of mortality in BS.¹ It is likely that both genetic and environmental factors influence the development of BS. Also, male gender and younger age at the onset of disease are associated with a more severe disease course.^{2,3} Herein, we describe the case of acute myocardial infarction (MI) in a 30-year-old BS patient complicated by coronary artery aneurysm (CAA) and acute stent

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thrombosis observed after percutaneous coronary intervention treatment.

Timeline

Day/month/year	Events
August 2015	The patient arrived at the emergency department complaining of chest pain for the previous 8 days. Coronary arteriography showed two aneurysms in the left anterior descending artery with a 90% stenosis lesion. A drug-eluting stent was implanted in the middle left anterior descending artery.
March 2016	The patient was admitted because of recurrent chest pain. Electrocardiogram showed elevated ST-segment in precordial leads and an emergency coronary angiography (CAG) showed a total left anterior descending artery occlusion. After thrombus aspiration, two large left anterior descending artery aneurysms were revealed.
April 2016	Intravascular ultrasound and computed tomography CAG were performed, and two covered stents were implanted in the middle and proximal left anterior descending artery, respectively.
A week after discharge	Immunosuppressive drugs (prednisone, cyclophosphamide, and thalidomide) were prescribed to the patient.
June 2018	Due to the poor management of oral ulcer and fertility needs, the prescription was changed into methylprednisolone and cyclosporine. Rheumatic antibody tests and pathergy test were negative; uneventful follow-up.

Case presentation

A 30-year-old Chinese man with 8-day intermittent chest pain was admitted to our hospital on 17 August 2015. His cardiovascular examination revealed an initial blood pressure of 115/72 mmHg, heart rate of 68 b.p.m., and his electrocardiogram showed Q-wave formation in the precordial and inferior leads and T-wave inversion in precordial leads (Figure 1E and F) with a raised initial high-sensitivity troponin level which was 304.7 ng/L (0–15 ng/L). On auscultation, his chest was clear and heart sounds were normal. He had experienced oral and GUs for the previous 12 years and complained about episodes of fever for the last 2 months. He had neither a history of smoking nor a family history of coronary heart disease. No medication was taken before admission. Coronary angiography (CAG) revealed two aneurysms in the left anterior descending artery (LAD) with a 90% stenosis following the distal aneurysm (Figure 1A and B;

Supplementary material online, Video S1). Consequently, a drug-eluting stent (DES, 3.5 × 24 mm) was implanted successfully in the middle LAD (Figure 1C and D). Additionally, aspirin, clopidogrel, atorvastatin, metoprolol, benazepril, trimetazidine, and pantoprazole were prescribed to the patient. A transthoracic echocardiogram showed apical hypokinesia, left ventricular ejection fraction of 50%, and an apical ventricular aneurysm (35 × 18 mm).

Four months after discharge, the patient was admitted because of recurrent chest pain, associated with nausea and vomiting. The electrocardiogram showed elevated ST-segment in precordial leads (Figure 2I) with a raised initial high-sensitivity troponin level which was 932.1 ng/L. The patient received double antiplatelet therapy and an emergency CAG revealed a total occlusion of the LAD (Supplementary material online, Video S2). A guidewire was successfully advanced across the occlusive lesion (Supplementary material online, Video 3) and a large fresh red thrombus was removed by aspiration catheter. The intervention was ended due to good blood flow of distal LAD after an intracoronary tirofiban bolus. The C-reactive protein (CRP) level was 75.5 mg/L (0–8 mg/L) and erythrocyte sedimentation rate (ESR) 84 mm/h (0–15 mm/h). Four days later, a GRAFTMASTER covered stent was implanted successfully in the middle LAD. In order to better characterize coronary anatomy, intravascular ultrasound, and computed tomography coronary angiography were performed, confirming the presence of two CAA at the level of the proximal and middle LAD (8 × 9 mm and 16 × 5 mm, respectively). Accordingly, a second covered stent was implanted in the proximal LAD on 11 April 2016 (Supplementary material online, Video S4).

The patient was treated appropriately with warfarin for 9 months. Immunosuppressive drugs were prescribed to the patient after his second discharge with a combination of prednisone, cyclophosphamide, and thalidomide to maintain normal CRP and ESR levels. Due to the poor management of the OU and fertility needs, the above prescription was then changed into cyclosporine and methylprednisolone. Additional testing for antibody and pathergy test were negative. The CRP and ESR level were normal. During the follow-up in 2019, the patient remains asymptomatic.

Discussion

In this case report, we detail the case of a young man with BS complicated by CAA. Behçet's syndrome is a systemic vasculitis and the diagnosis of BS is made purely based on clinical criteria because no specific laboratory examination exists.^{4–6} Additionally, a history of recurrent fever is present in 22% of BS patients which is associated mostly with vascular involvement.⁷

Coagulation disorders due to anti-cardiolipin antibodies or vasculitis are believed to be involved in the cardiovascular manifestations of BS. However, some evidence supports the use of an escalated antithrombotic regimen in these patients.⁵ The latest guideline-recommended intensive anticoagulation only for patients with large or rapidly expanding CAA. Due to the higher associated thrombus burden, thrombectomy and anticoagulation were performed in this case.

As the mortality of cardiovascular manifestation in BS is 20%, immunosuppressive therapy is crucial for BS.⁵ Notably, the ideal time

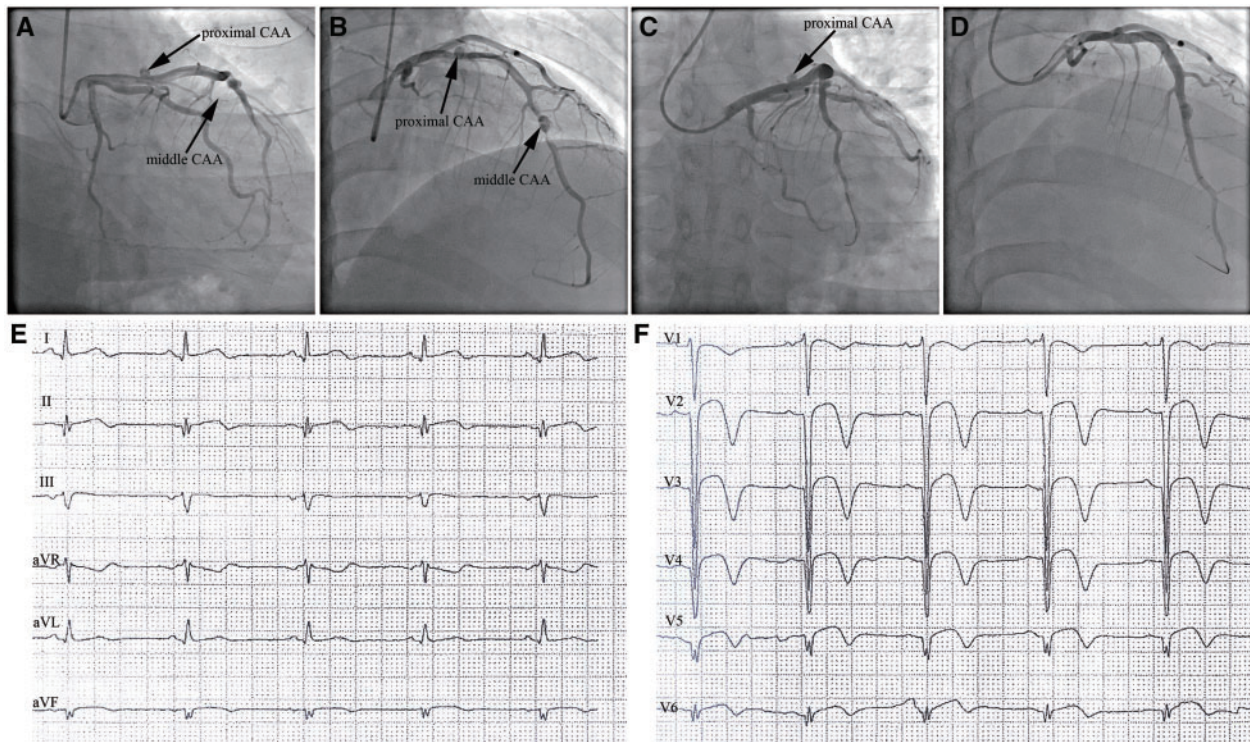


Figure 1 (A and B) Coronary angiography on the first admission. (C and D) Coronary angiography after drug-eluting stent implantation. (E and F) A 12-lead resting electrocardiogram on admission.

point for drug administration is still a controversial topic. It has been suggested that the administration of intensive immunosuppressive treatment at 2–3 days post-MI may be most beneficial and cause less adverse effects. However, immunosuppressive treatments may interfere with the ventricular healing process and increase the risk of scar thinning and myocardial rupture. In addition, intervention should not be delayed if the patient is symptomatic.⁸ Therefore, we initiated immunosuppressive therapy after discharge.

Opinions regarding the stent type for BS patients with CAA remain sparse. A recent recommendation suggested that the covered stent is most commonly used in CAA.⁹ In our case, CAAs became larger after DES implantation, possibly due to the vascular complication of BS or rapamycin damage to the vascular wall. Accordingly, we believe that traditional DESs should be avoided in BS patients. The ideal interventional method remains to be elucidated by further investigations.

Conclusions

This case highlights the use of the covered stent, immunosuppressive therapy, and intensive anticoagulation therapy in the treatment of BS patients with CAA. However, the management of these patients still is considerably disputed, and the ideal method of management needs to be investigated further.

Lead author biography



Shenghua Zhou, MD, graduated from Hunan Medical College, China. Currently, he is the chairman of the Hunan provincial Society of Cardiology and the director of the Department of Cardiology of the Second Xiangya Hospital, Central South University, China. His research is focused on coronary heart disease and arrhythmia.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

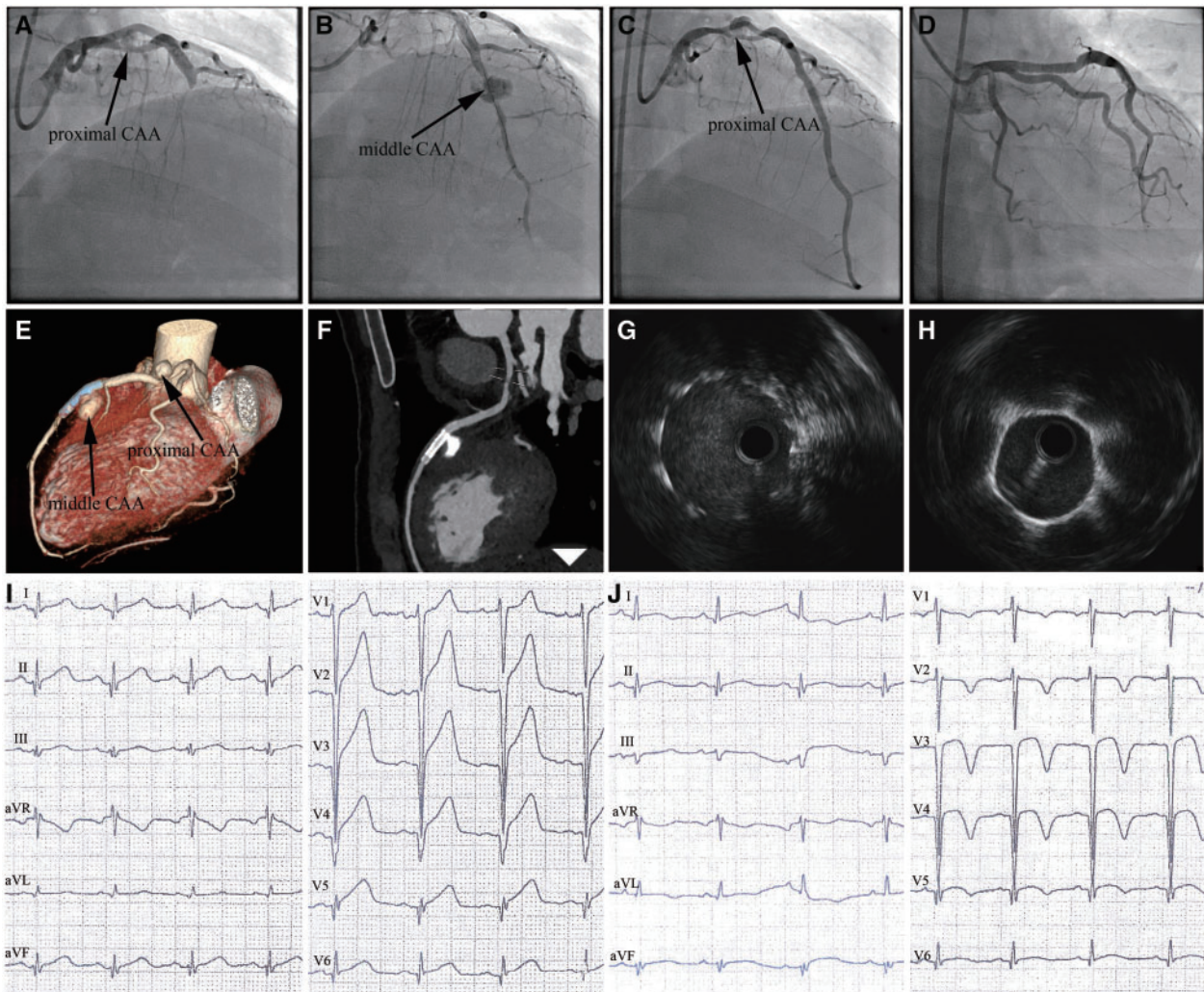


Figure 2 (A) Coronary angiography on the second admission. (B) Coronary angiography after thrombus aspiration. (C) Coronary angiography after the first covered stent implantation. (D) Coronary angiography after the second covered stent implantation. (E and F) Computed tomography coronary angiography imaging of the left anterior descending coronary aneurysm. (G and H) Intravascular ultrasound imaging of the middle coronary artery aneurysm before and after stent implantation. (I and J) Electrocardiogram on admission and discharge.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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