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CASE REPORT
Coronary heart disease

Coil embolization to giant left anterior descending artery and left circumflex artery coronary artery aneurysm after failed coronary aneurysmal repair in IgG4-related disease: a case report

Panupong Pota [1], Pannipa Suwannasom [1]*, Surin Woragidpoonpol[2], and Tanop Srisuwan [3]

[1]Division of Cardiology, Department of Internal Medicine, Faculty of Internal Medicine, Chiang Mai University, 110 Intavaroros Road, Sripoom District, Muang, Chiang Mai 50200, Thailand; [2]Division of Cardiothoracic Surgery, Department of Surgery, Faculty of Internal Medicine, Chiang Mai University, Chiang Mai, Thailand; and [3]Division of Interventional Radiology, Department of Radiology, Faculty of Internal Medicine, Chiang Mai University, Chiang Mai, Thailand

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Background IgG4-related disease (IgG4-RD) is a chronic fibroinflammatory condition with multiple-organ involvement. Rupture of coronary artery aneurysms (CAAs) in IgG4-RD is rare.

Case summary A 65-year-old man with IgG4-RD has suffered from recurrent episodes of arterial aneurysms since 2003. He presented with chest pain and hypotension caused by localized cardiac tamponade at right ventricle free wall due to the rupture of coronary artery aneurysm (CAA) of left anterior descending artery (LAD). An urgent LAD aneurysm repaired with bovine pericardium and obliterated aneurysmal sac with cryo-acrylate glue was done together with coronary artery bypass grafting (CABG) using saphenous vein graft (SVG) to LAD and SVG to posterior descending artery. Three-month after surgery, the follow-up coronary computed tomography angiography (CCTA) revealed a growing in size of LAD and the second obtuse marginal (OM) branch aneurysm. Heart team discussion agreed to schedule the patient for double coil embolization to LAD and second OM aneurysm under intravascular ultrasound guidance. Both aneurysms were successfully obliterated with vascular coils. Two-week follow-up coronary angiogram showed complete occlusion of LAD aneurysm and near occlusion of the second OM branch aneurysm.

Discussion Coronary artery aneurysm rupture is a life-threatening condition that required prompt detection and treatments. In IgG4-RD patients, acute cardiac tamponade suggesting the rupture of CAA. Coil embolization is an alternative treatment in patients who suffered from recurrent CAA after surgical repair. Serial CCTA is important for early detection of aneurysm in IgG4-RD patients who had vascular involvement.

Keywords Case report • Cardiac tamponade • Coronary artery aneurysm • Rupture • IgG4-related disease • Coil embolization

ESC Curriculum 2.4 Cardiac computed tomography • 3.1 Coronary artery disease • 3.4 Coronary angiography • 7.5 Cardiac surgery

* Corresponding author. Tel: +66-53-934453, Email: pannipa.s@cmu.ac.th
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Learning points

- The most common IgG4-related aneurysm was found in aorta and primary aortic branches. Coronary artery aneurysms (CAAs) in IgG4-related disease are uncommon.
- Coronary artery aneurysms are mostly asymptomatic. Aneurysm rupture is rare but life-threatening condition since it leads to acute cardiac tamponade
- Non-invasive interval coronary computed tomography angiography study is important to assess the progression of CAAs due to the lack of biomarkers data that related to IgG4-related disease prognosis.

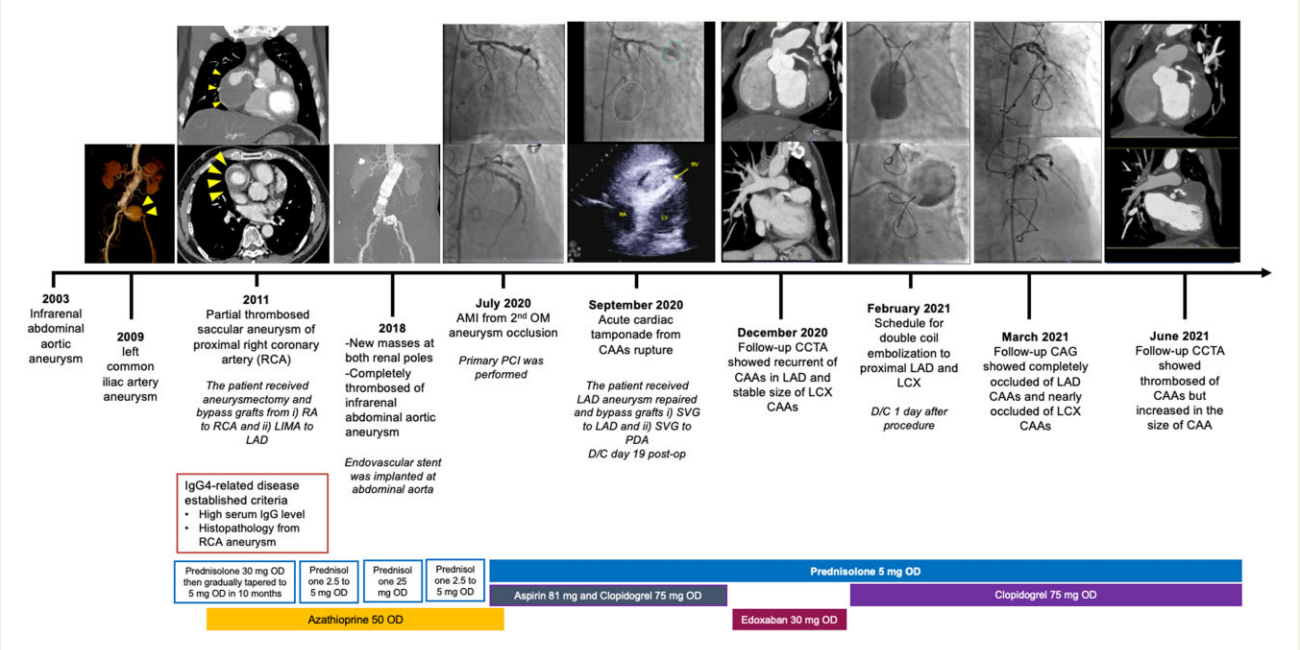
Introduction

Giant coronary artery aneurysm (CAA) is defined by the aneurysm occurring in a coronary artery greater than 20 mm in size.[1] Coronary artery aneurysm leads to various clinical manifestations including myocardial ischaemia or infarction, compression of adjacent structures, and acute cardiac tamponade as a result of aneurysm rupture.[1] IgG4-related disease (IgG4-RD) is an immune-mediated fibroinflammatory condition involving multiple organs.[2] The clinical manifestations when the cardiovascular system is involved could include aortitis/arteritis and inflammatory aneurysms.[3] However, ruptured CAA is an uncommon presentation in patients with IgG-4 RD.[4]

Timeline

Case presentation

A 65-year-old man was diagnosed with IgG4-RD nine years prior to the presentation based on multiple sites of arterial aneurysm (infrarenal abdominal aortic aneurysm and left common iliac artery aneurysm), retroperitoneal fibrosis and high serum IgG concentration (5470 mg/dL, normal value 800–1700 mg/dL). Since then, he has been treated with prednisolone and azathioprine. After being diagnosis of IgG4-RD for 4 months, a follow-up CT scan showed a partially thrombosed saccular aneurysm of the proximal right coronary artery (RCA), 5.3 cm × 5.4 cm × 6.4 cm in size and enhanced soft tissue in the left anterior descending artery (LAD) and left circumflex artery (LCX) suggesting lesions due to vasculitis (*Figure 1A,B*). The patient has never had angina. Coronary angiography (CAG) showed 70% diameter stenosis of the mid-LAD, total occlusion of the proximal RCA, and mild disease of LCX. Right coronary artery aneurysmectomy was performed together with coronary artery bypass graft (CABG) surgery using a right radial artery (RA) graft to the distal RCA and a left internal mammary artery (LIMA) graft to the distal LAD. Histopathology of the RCA aneurysm showed fibrosis with extensive lymphoplasmacytic inflammation in the tunica media and adventitia supporting the diagnosis of IgG4-RD. Six years prior to the presentation, a follow-up coronary CT scan showed patent RA to distal RCA but occlusion in the LIMA to LAD. Multiple small fusiform aneurysms with periarterial soft tissue lesions were noted proximal to the mid LAD, proximal LCX to 2nd obtuse marginal branch (*Figure 2A–D*). Two years prior to the presentation, endovascular stent was implanted at abdominal aorta. The follow-up CT scan showed complete thrombosed of infrarenal abdominal aortic aneurysm.



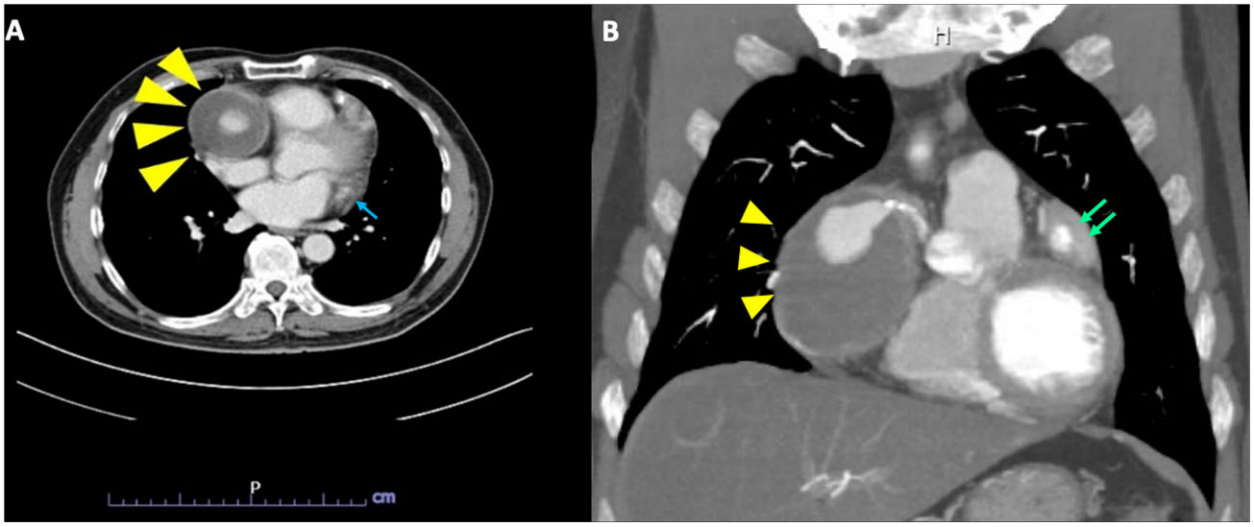


Figure 1 Coronary computed tomography angiography (CCTA) showed giant coronary artery aneurysm (CAA) of the proximal right coronary artery. The partially thrombosed saccular aneurysm of the proximal right coronary artery in axial view (*A*) and coronal view (*B*). Yellow arrowheads indicate the aneurysm. The enhancing of soft tissue was also noted at left circumflex artery (blue arrow, *A*) and anterior descending artery (green arrow, *B*) suggesting lesions due to vasculitis.

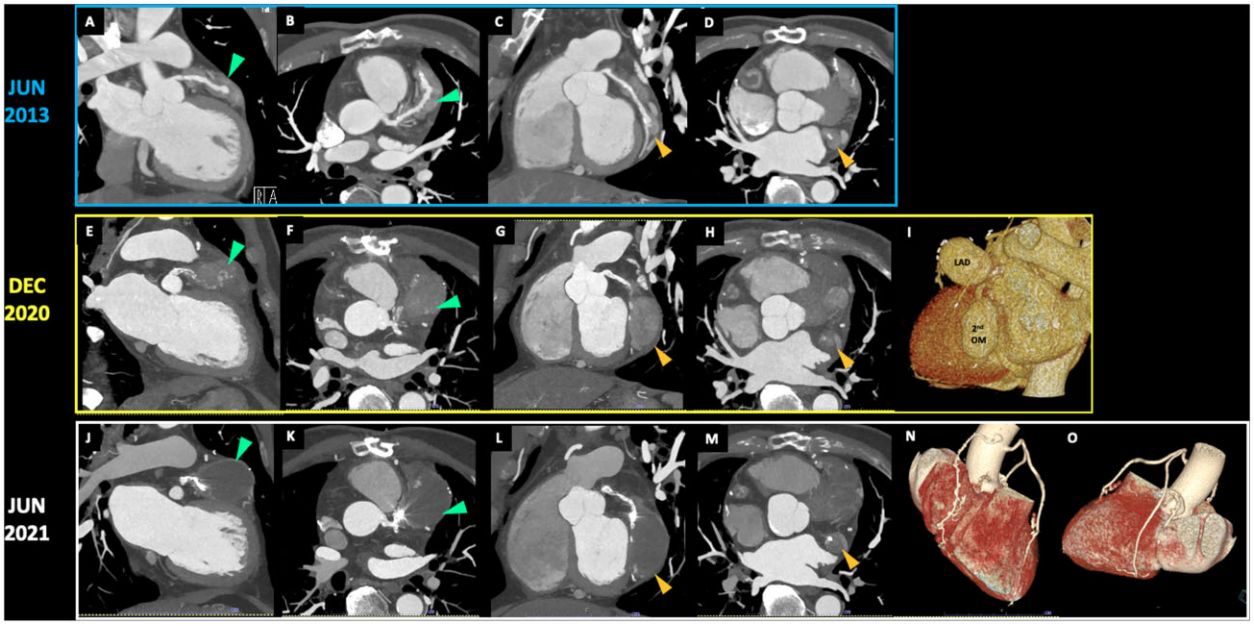


Figure 2 Serial coronary computed tomography angiography of the patient after right coronary artery aneurysmectomy in 2011. The series of coronary computed tomography angiography after coronary artery bypass grafting in 2011. The top row (*A–D*) shows the follow-up coronary computed tomography angiography in 2013. Multiple small fusiform aneurysms with periarterial soft tissue lesions were noted proximal to mid-left anterior descending artery (green arrowheads, *A,B*), proximal left circumflex artery to 2nd obtuse marginal branch (orange arrowheads, *C,D*). The follow-up coronary computed tomography angiography in year 2020 are shown in the middle row (*E–I*). The size of left anterior descending artery coronary artery aneurysm kept growing despite aneurysmectomy (green arrowhead, *E,F*). (*G,H*) illustrates 2nd obtuse marginal coronary artery aneurysm. (*I*) shows the giant coronary artery aneurysm of left anterior descending artery and the 2nd obtuse marginal branch from the volume rendering technique. The follow-up coronary CT scan in June 2021 shows the left anterior descending artery to be completely thrombosed (green arrowheads, *J,K*) and a 2nd obtuse marginal coronary artery aneurysm (orange arrowheads, *L,M*) but an overall increase in size. (*N*) The patency of the RA-distal right coronary artery and SVG-distal left anterior descending artery. (*O*) shows the patency of the SVG-2nd obtuse marginal branch.

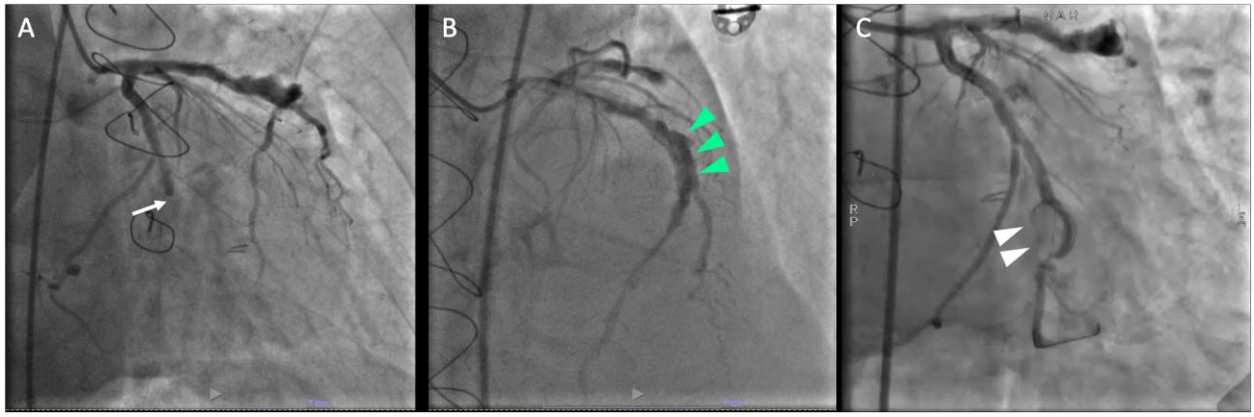


Figure 3 Coronary angiography in July 2020. RAO caudal view showed near occlusion of the 2nd obtuse marginal (OM) branch (white arrows) (*A*). PA cranial view showing fusiform coronary artery aneurysm (green arrowheads) at mid-left anterior descending artery (*B*). After percutaneous balloon angioplasty to the 2nd obtuse marginal branch, TIMI 3 flow was noted but a large coronary artery aneurysm (white arrowheads) distal to the occlusion site with thrombus was found (*C*).

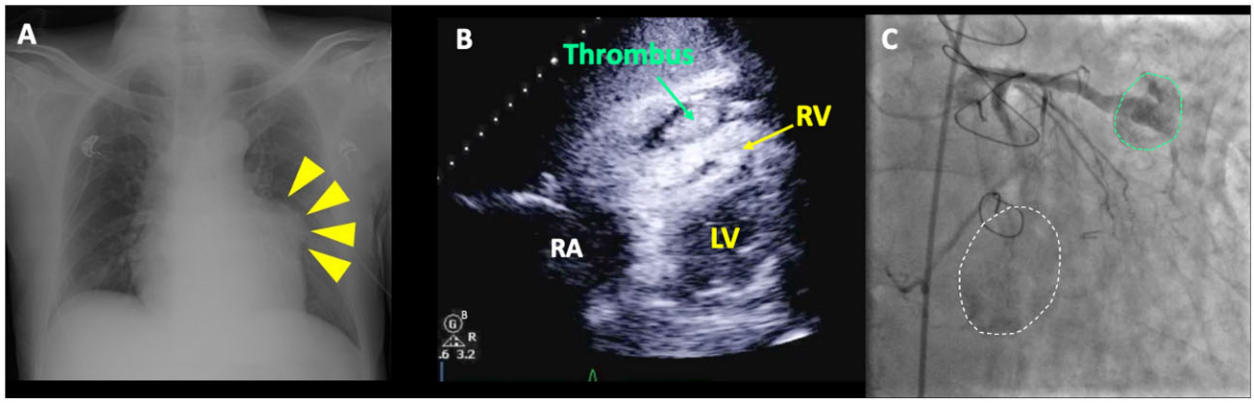


Figure 4 Diagnostic imaging in September 2020. Chest radiograph demonstrating ill-defined soft tissue density (yellow arrowheads) at the left heart border (*A*). Still images from echocardiography showing mixed echogenic large loculated pericardial effusion above right ventricle free wall causing cardiac tamponade (*B*). Giant coronary artery aneurysm at mid-left anterior descending artery and the 2nd obtuse marginal branch are outlined in green and white dotted lines (*C*).

At the index event, the patient presented with acute infero-lateral wall ST-elevation myocardial infarction. Emergency CAG showed total occlusion of the 2nd obtuse marginal (OM) branch (*Figure 3A*) and mid-LAD CAA (*Figure 3B*). The RA graft to the distal RCA was patent. After balloon angioplasty was performed in the 2nd OM branch, the CAA was found just distal to the culprit lesion with a thrombus in the aneurysmal sac (*Figure 3C*). Dual antiplatelet therapy was planned for 12 months.

Two months after the index event, the patient had recurrent chest pain 2 h prior to admission. The patient was taking a prescribed beta-blocker, his blood pressure at presentation was 77/46 mmHg, and pulse rate was 64 beats per minute. Cardiovascular examination showed regular rhythm, normal cardiac auscultation and negative pulsus paradoxus. The chest radiograph demonstrated ill-defined soft tissue density at the left heart border (*Figure 4A*). Echocardiography showed mixed echogenic large loculated pericardial effusion above the right ventricle free wall causing cardiac tamponade (*Figure 4B*). Emergency CAG showed giant CAAs at the mid LAD and the 2nd OM branch (*Figure 4C*, Supplementary material online, *Video S1*). No active extravasation of contrast media to pericardial space due to slow blood flow in CAAs was seen. However, there was a suspicion of rupture of CAAs from the acute cardiac tamponade. The patient was immediately transferred to the operating theatre. Intra-operative notes reported a rupture of the LAD CAA but no evidence of leakage from the 2nd OM branch CAA. The LAD CAA was repaired using bovine pericardium and the sac being filled with cryo-acrylate glue. Coronary artery bypass graft surgery was performed using a saphenous vein Y-graft to distal LAD and 2nd OM branch. The patient was extubated on post-operative Day 3 and discharged from hospital on Day 19.

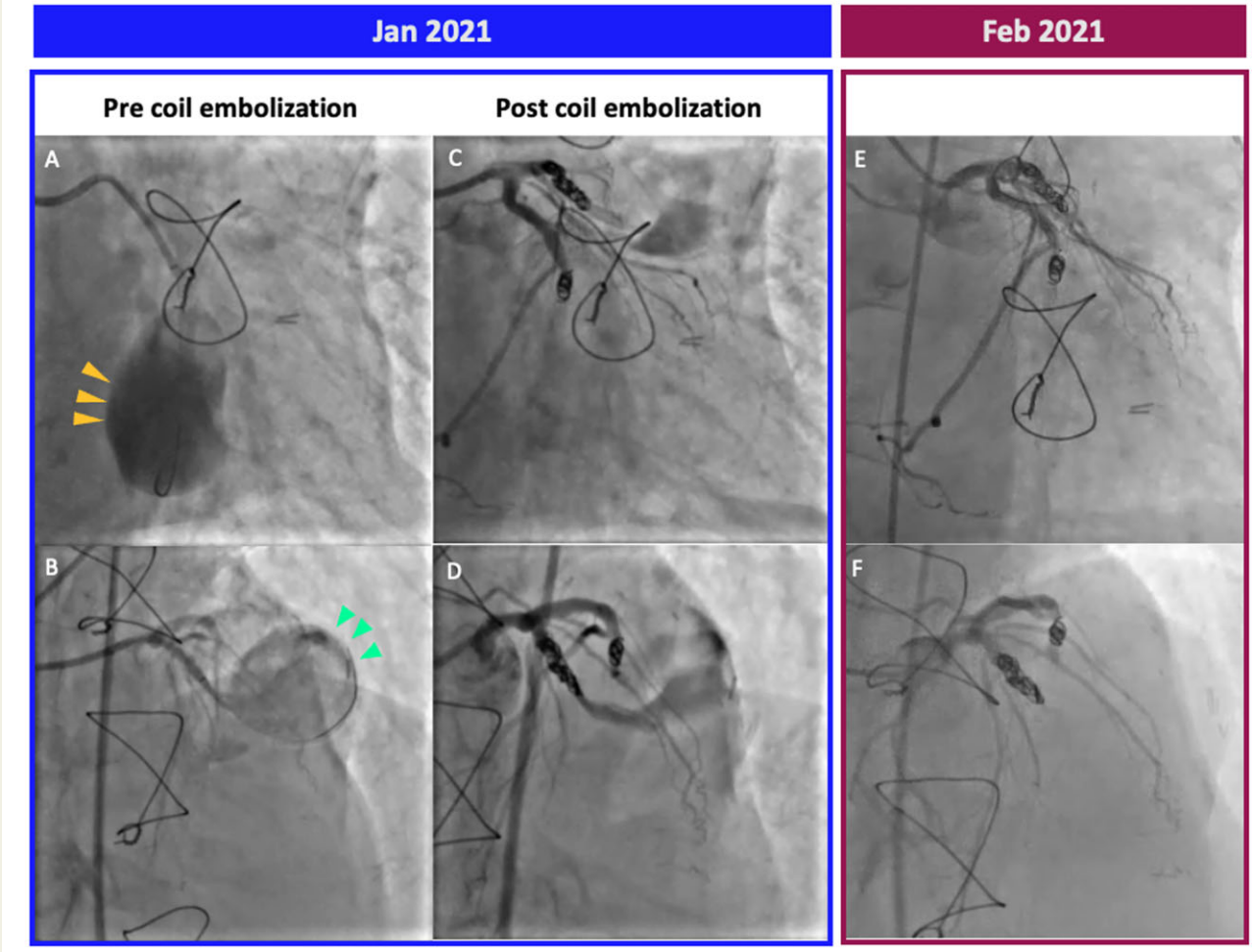


Figure 5 Comparison of the coronary angiograms pre-, immediately after coil embolization, and after 2 weeks. (*A* and *B*) Selective images of the angiography via extension catheter of the 2nd obtuse marginal branch (orange arrowheads) and left anterior descending artery (green arrowheads) coronary artery aneurysm before coil embolization. Immediately after coil embolization, antegrade flow of the 2nd obtuse marginal branch had almost vanished (*C*) whereas slow antegrade flow of left anterior descending artery was noted (*D*). At 2 weeks, a very faint antegrade flow was noted in both 2nd obtuse marginal branch and left anterior descending artery (*E* and *F*).

Three months after surgery, the follow-up coronary CT scan showed an increase in size of the CAAs in both the mid LAD and the 2nd OM branch (*Figure 2E–I*). Discussion among the heart team resulted in an agreement to treat both CAAs with coil embolization. Reasons for the choice of technique were: (i) avoidance of open-heart surgery due to the possibility of injury to SVG and (ii) it was not feasible to repair the CAA in the 2nd OM branch CAA using surgical techniques due to its anatomical position. The procedure was performed via the right femoral approach using the 6 Fr delivery system. Selective angiography of the CAAs was performed using an extension catheter to evaluate the extent of the aneurysm and confirm the size of the neck of the CAA (Supplementary material online, *Video S2*). The pre-operative CT angiographic image showed the proximal LAD aneurysmal neck size, measuring 4 mm in diameter and 2nd OM aneurysm neck size 2 mm in diameter (Supplementary material online, *Figure S1A,B*). The use of the detachable microcoil was planned due to its adjustability and facility for precise placement. Measurements from intravascular ultrasound confirmed the neck of LAD and LCX CAA were 3.5 mm and 3.0 mm, respectively. The size of the first coil for framing was determined by 10–20% oversizing of the aneurysm neck. Three vascular coils (size 5 mm × 8 cm, 4 mm × 8 cm, and 3 mm × 6 cm) were implanted via Renegade STC 18 microcatheter to the proximal LAD. Two vascular coils (size 4 mm × 8 cm, 3 mm × 6 cm) were implanted into the 2nd OM branch. Details of the procedure are shown in Supplementary material online, *Figure S1A–Y*. The final angiogram showed fade antegrade flow of both aneurysms (*Figure 5A–D*). The follow-up angiogram at 2 weeks showed total occlusion of the mid LAD CAA and near occlusion of the 2nd OM branch CAA (*Figure 5E,F*, Supplementary material online, *Video S2*). The follow-up coronary CT scan at 6 months showed completely thrombosed mid LAD and 2nd OM CAA but increase in overall size (LAD from 2.5 cm × 2.5 cm to 2.9 cm × 3.2 cm; LCX from 3.3 cm × 3.2 cm to 3.7 cm × 4.8 cm, *Figure 2J–M*). All grafts from the

last CABG operation were patent (*Figure 2N–O*). The patient currently takes prednisolone 5 mg once daily.

Discussion

IgG4-related disease (IgG4-RD) is an immune-mediated condition that mimics inflammatory disorder, malignancy, and infection in multiple organs. The classic IgG4-RD patient is a middle-aged to elderly male.[5] Arterial involvement in IgG4-RD is usually in association with large to medium-sized arteries[3] and mostly asymptomatic. In our case, the RCA aneurysm was incidentally detected from a follow-up CT scan for aortic aneurysm. The coronary artery lesions in IgG4-RD can present as wall thickening, stenosis, soft tissue thickening, aneurysm, and acute coronary syndrome.[6] Pseudo-tumour development around the coronary artery can lead to aneurysmal change at the disease segment.[3,7] Corticosteroids are acknowledged as being the first-line therapy for IgG4-RD; however, they do not attenuate the size of aneurysm following occurrence. Tajima *et al.*[8] reported that active inflammation was detected only in the early inflammatory phase whereas well-developed aneurysms showed mild or no inflammation. In addition, corticosteroids may lead to aneurysmal rupture as a result of the reduction of intimal thickening.[8] Therefore, the ideal dosage of the steroid will suppress inflammation but not compromise the strength of the arterial wall. In this case, the patient received low-dose prednisolone and azathioprine for several years, but new aneurysms still developed. The role of conventional steroid-sparing agents or B-cell depletion remains controversial[9] and may not have prevented aneurysmal formation in this case. The plausible cause of recurrent CAA in the mid LAD despite aneurysm repair could be a newly developed aneurysm proximal to the ligation site. As shown in *Figure 2B*, the inflow of the mid-LAD CAA was located 35 mm distal to the 1st septal branch, but the recurrent CAA was located 20 mm distal to the 1st septal branch. Both CAAs could not be treated with a covered stent since we could not locate their distal outflow from selective angiography. The challenge in performing coil embolization in this case was the short working length above the aneurysmal sac. Precise coil size selection is the key of the procedural success. Despite both aneurysms being completely thrombosed, the size of CAA was still growing. This increase in the size of the CAA may be explained by the antegrade and retrograde flow that filled the sac before it became completely thrombosed.

Conclusion

This case report describes cardiac tamponade from a spontaneous CAA rupture in an IgG4-RD patient. Despite the patient having undergone open heart surgery for CAA repair, recurrent CAAs still occurred. Coil embolization to huge CAA was successfully performed without complication in this case. Therefore, coil embolization of CAA could be an alternative treatment for patients in whom surgical repair has been unsuccessful or have unsuitable anatomical justification for surgery.

Lead author biography

Panupong Pota is currently cardiology fellow at Maharaj Nakorn Chiang Mai Hospital, Chiang Mai, Thailand. He earned medical degree from Ching Mai University in 2016. He completed Internal Medicine training at the same institute in 2020.

Supplementary material

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

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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