CASE REPORT

ST-elevation myocardial infarction in a young adult secondary to giant coronary aneurysm thrombosis: an important sequela of Kawasaki disease and a management challenge

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SUMMARY

Thrombosis of a coronary artery aneurysm (CAA) is a rare trigger for ST-elevation myocardial infarction (STEMI) and an important cause of STEMI in young adults previously affected by Kawasaki disease. Initial management should proceed in line with standard STEMI-management guidelines advocating antiplatelet medication and emergency coronary angiography. Acute CAA thrombosis presents the interventional cardiologist with unique challenges during attempted percutaneous revascularisation. In the absence of consensus guidelines, experiential reporting can therefore be of great value. We report on a 36-year-old Vietnamese woman presenting with an inferior STEMI secondary to two giant thrombosed aneurysms of the right coronary artery. Coronary wiring and thrombus aspiration temporarily improved coronary flow but recurrent thrombus with distal embolisation resulted in ventricular fibrillation and cardiogenic shock. Emergency surgical revascularisation subsequently provided a definitive and successful outcome. We discuss the challenges of percutaneous coronary intervention in this scenario and review previous reports to give an overview of principles of decision-making and management.

BACKGROUND

Coronary artery aneurysm (CAA) with acute thrombosis is an uncommon but recognised cause of acute coronary syndromes. CAAs are observed in 4.9% of elective coronary angiograms when defined as an arterial segment with a luminal diameter of 1.5 times that of the non-diseased vessel.¹ Much larger aneurysms can occur and are termed giant when the diameter exceeds 8 mm. While CAA formation is most commonly secondary to destruction of the vessel wall media due to atherosclerosis, it is also seen as a complication of Kawasaki disease (KD), among other disease processes.¹ KD is a childhood vasculitis of unknown aetiology with the highest incidence in Asian, particularly Japanese, children.² The diagnosis is clinical and if the acute phase of KD is untreated the risk of coronary sequelae rises from 5% to 25%.³

CAA with or without accompanying focal stenosis is the most prevalent coronary aberration following KD, and there appears to be a predilection for the right coronary artery (RCA) and the proximal portions of the major epicardial vessels.^{4 5}

'Silent' aneurysms carry a 23% annual risk of thrombosis⁶ and this represents the commonest cause of acute coronary syndromes in KD,⁷ the timing of which may be delayed beyond the third decade.³ Thrombosis of a CAA secondary to clinically unrecognised KD is therefore an important cause of acute myocardial infarction (AMI) in young adults.⁸ In the setting of acute CAA thrombosis causing ST-elevation myocardial infarction (STEMI), there is a lack of consensus opinion regarding optimal acute management, especially with respect to percutaneous coronary intervention (PCI). We report the case of a young woman in whom percutaneous reperfusion was attempted but ultimately unsuccessful, necessitating emergency coronary surgery to definitively treat acutely thrombosed giant aneurysms in the proximal and mid-segments of her RCA.

CASE PRESENTATION

A 36-year-old woman of Vietnamese heritage presented to the emergency department with a 4 h history of new onset central chest pain. At the time of presentation, her cardiovascular risk profile was only notable for a family history of coronary artery disease. She had previously been well and was the mother of a 4-year-old child.

INVESTIGATIONS

Initial 12-lead ECG demonstrated sinus rhythm, rate 65 bpm, with 3 mm ST-segment elevation involving all three inferior leads, accompanied by ST-segment depression anteriorly, consistent with an acute inferoposterior STEMI (figure 1). Troponin I was already elevated at 0.11 µg/L (normal range 0–0.08), and the patient's blood pressure was initially 130/90 mm Hg.

She was given aspirin 300 mg and prasugrel 60 mg orally, and expediently underwent coronary angiography via a 6 Fr right femoral arterial sheath. This revealed a large aneurysm (estimated angiographically to be 7–8 mm) in the proximal segment of her left anterior descending artery (LAD), but otherwise no significant left coronary system epicardial lesions. Grade 2 retrograde collaterals from the LAD septal branches were seen to fill the distal posterior descending artery (PDA) and posterolateral ventricular (PLV) branches of the RCA.

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Figure 1 Twelve-lead ECG on presentation showing changes consistent with acute inferoposterior ST-elevation myocardial infarction (STEMI).

TREATMENT

Immediately after angiographic images were obtained of the left coronary system, the patient went into ventricular fibrillation (VF) requiring two defibrillations to restore sinus rhythm and cardiac output. A 6 Fr JR4 coronary guide catheter was then immediately used, and contrast injection of the RCA revealed a 100% occlusion involving a dilated segment of the proximal vessel, with thrombolysis in myocardial infarction (TIMI) 0 flow beyond the occlusion site (figure 2A).

After heparin was given, the occlusion was rapidly crossed with a balance middle weight (BMW) coronary guide wire

(Abbott Vascular), immediately restoring TIMI 2/3 flow to the RCA and revealing two giant aneurysms (figure 2B). The proximal aneurysm at the occlusion site appeared to be at least 10 mm in size, while there was a second aneurysm of similar diameter in the mid-segment of the RCA, both containing multiple filling defects consistent with thrombus. Three separate attempts were initially made to perform aspiration thrombectomy with a 6 Fr Export catheter (Medtronic), beginning at the site of the second aneurysm. However, relatively little material could be retrieved. Following this, coronary flow had diminished again (TIMI 1), and intravenous tirofiban was

Figure 2 Angiographic images of the right coronary artery (RCA) before and after attempted percutaneous intervention. Occluded proximal RCA with thrombolysis in myocardial infarction (TIMI) 0 flow, and partially visualised aneurysm before wiring (white arrow) (A). Proximal and mid-vessel giant aneurysms with considerable clot burden immediately after wiring (B). Subsequent appearance after attempted aspiration thrombectomy, start of the of intravenous glycoprotein IIb/IIIa and gentle balloon inflation of the mid-RCA aneurysm outlet, demonstrating loss of a right ventricular branch (yellow arrow) and downstream embolisation of clot, occluding the distal RCA and branches (blue arrow) (C). This was the final appearance before the patient was taken to coronary artery bypass surgery after resuscitative measures and haemodynamic stabilisation.

started. The cardiothoracic surgical team was contacted with a view to early definitive treatment by surgical ligation of the aneurysms and bypass revascularisation to the distal RCA.

While waiting for surgical review, we considered exchanging to an 8 Fr sheath and 8 Fr multipurpose guide catheter to enable more effective thrombus aspiration through a larger calibre system. In order to do this, we first exchanged the BMW wire for a balance heavy weight (BHW) wire through the lumen of a 1.5×8 mm over-the-wire balloon. We then tried unsuccessfully to position a second BHW wire into the distal RCA to provide support for upsizing to an 8 Fr system, but could not pass this wire beyond the outlet of the second (mid-segment) RCA aneurysm. By this stage, RCA flow had diminished to TIMI 0–1 beyond this site, and so we performed a gentle balloon inflation to 6 atm with a 2.5×15 mm Emerge balloon at the outlet of the mid-segment aneurysm. This improved flow to TIMI 2 grade, with the appearance of thromboembolic propagation distally. Further aspirations were performed in the mid-segment and distal segment of the RCA with the 6 Fr Export system, retrieving white thrombus but only marginally improving coronary flow appearance.

At this point, the patient's clinical status deteriorated markedly due to multiple recurrent VF episodes responsive to defibrillation (total of 9 shocks), and cardiogenic shock (blood pressure (BP) 50/20 mm Hg despite a total of 2 L intravenous fluid previously administered). She was given boluses of metaraminol and epinephrine, started on a low-dose epinephrine infusion for haemodynamic support, and intubated and ventilated mechanically. Lignocaine and amiodarone were also given to achieve electrical stability. After one more attempt at aspiration thrombectomy, she had TIMI 2 flow in the distal RCA, with TIMI 1 flow in the PDA but TIMI 0 flow in the PLV branch. She was transferred for urgent coronary artery surgery in sinus rhythm, 62 bpm, with BP 115/60 mm Hg, and with the 6 Fr JR4 guide catheter and BHW coronary guide wire left in situ.

At surgery, both right coronary aneurysms were found to be approximately 2 cm in diameter, with thrombus present in both. The inlet and outlet of both aneurysms were suture ligated and each of their branch vessels oversewn. As there were no large right ventricular branches suitable for grafting, one saphenous vein graft was anastomosed to the segment between the two aneurysms, and another vein graft anastomosed to the distal RCA. A decision was made not to intervene on the LAD aneurysm.

OUTCOME AND FOLLOW-UP

The early postoperative course was complicated by hypotension and anaemia. Inpatient transthoracic echocardiogram demonstrated mild left ventricular dysfunction with akinesis of the basal inferior wall, and severe hypokinesis of the basal infero-septal and mid-posterior segments, along with a dilated right ventricle that had moderate systolic dysfunction but no significant tricuspid regurgitation. The patient was discharged after a total of 10 days, and has since been maintained on aspirin, bisoprolol, irbesartan, atorvastatin and, initially, frusemide. Six months later she has remained stable with New York Heart Association class I shortness of breath and residual mild biventricular dysfunction (left ventricular ejection fraction 45%). Retrospective history revealed that, at 4 years of age, she had been admitted to hospital with fever and a generalised exanthema, supporting our suspicion that the underlying aetiology of her CAAs was KD.

DISCUSSION

This is a case of spontaneous thrombosis of previously unrecognised CAAs in a woman of childbearing age. Emergency PCI was unsuccessful due to large thrombus burden that persisted, despite wiring of the occlusion site, thrombus aspiration, intravenous glycoprotein IIb/IIIa inhibitor use and balloon inflations. This was complicated by distal embolisation, recurrent ventricular arrhythmias and cardiogenic shock. Definitive surgical revascularisation of the infarct-related artery was considered early in this patient's management, and was ultimately effective with satisfactory clinical outcome.

Various therapeutic strategies at the time of emergency angiography for acute KD-related CAA thrombosis are described in the literature.^{7–14} Our case is consistent with previous reports of infarct-related CAAs (not KD specific) suffering large thrombus burden resulting in persistently poor coronary reflow (TIMI ≤2) and distal embolisation in 62.5% and 70.8% of cases, respectively.¹⁵ With this in mind, intravenous or intracoronary administration of glycoprotein IIb/IIIa inhibitors or antithrombotics should be considered early, along with aspiration thrombectomy, which may benefit from the use of larger bore (7 or 8 Fr) devices. Some have also reported success in children and adults, following the use of intracoronary or intravenous thrombolysis.^{7 12 13 16} Depending on extent of thrombus burden, aneurysm size and clinical stability, further percutaneous intervention with balloon angioplasty and stent deployment may not be indicated or appropriate, and focus instead should be turned to stabilisation prior to surgical revascularisation. This approach was favoured in a case series of 49 patients with AMI secondary to KD-related CAA thrombosis, in whom 53% received medical management following coronary angiography, with 46% then proceeding to elective coronary artery bypass grafting.⁷

While there are a number of published case reports and case series of successful sealing of CAA with stents, relatively few have been in the emergency setting of acute STEMI with large clot burden,^{9 17} making it difficult to strongly advocate for this approach. Polytetrafluoroethylene (PTFE)-covered stents, first used to treat coronary perforations, have been used in the percutaneous treatment of stable CAAs and AMI secondary to aneurysm thrombosis.¹⁸ Covered stents isolate the body of the aneurysm from luminal flow, resulting in thrombosis within the aneurysm. These tubes are composed of two stainless-steel stents separated by a layer of PTFE. This three-layered construct is relatively bulky and limits treatment of tortuous lesions.^{19 20} Other disadvantages are high re-stenosis rates (approximately 20% in one study¹⁸), comparable with rates for bare metal stents (BMS), and poor endothelialisation with risk of thrombotic occlusion.^{19 21} BMS may be preferable to covered stents if flexibility is demanded by tortuosity within the aneurysm and the peri-aneurysmal vessel.²⁰ In one novel case report, a drug-eluting stent was deployed co-axially within a covered stent for treatment of acute CAA thrombosis in an attempt to reduce the likelihood of stent re-stenosis.²²

In cases where stenting is considered, imaging with intravascular ultrasound (IVUS) may yield useful information regarding the morphology, wall composition, thrombus burden and distribution within the CAA,¹⁴ especially as large and highly irregular aneurysms may compromise stent apposition.²⁰ However, the decision to gain additional information using IVUS must be balanced against the need for timely definitive management. In our patient, IVUS assessment was neither necessary nor appropriate, given the patient's instability, persistent thrombus and very large aneurysm size, which precluded consideration of stenting.

Although no clinical guidelines exist, immediate surgical revascularisation on first discovering CAA thrombosis has been advocated when there is involvement of the left main coronary artery.⁷ A surgical approach may also be sought after initial stabilisation with antiplatelet and antithrombotic therapy, and when PCI is not technically feasible or has a high likelihood of complication, such as distal embolisation of large thrombus burden, or expected stent malapposition in very large, irregular CAAs. We recognised early in our patient's treatment that definitive revascularisation with PCI would not be feasible and contacted the surgical team accordingly. Notably while in the cardiac catheterisation laboratory, her coronary flow grade and clinical stability were best after we initially crossed the culprit lesions with the first guide wire. While waiting for surgical transfer, we made various attempts to improve thrombus burden by aspiration thrombectomy, and then tried to pass a buddy wire that would allow upsizing of the guide catheter and aspiration system. This further instrumentation of the culprit vessel was associated with persistent thrombus, distal embolisation, and deterioration of coronary flow and clinical state, requiring further resuscitative measures. This highlights the complex nature of decision-making in these cases, and provides a cautionary lesson of the delicate balance between accepting a clinically stable, albeit suboptimal coronary flow grade before surgery, and trying to perfect the situation.

American Heart Association guidelines pertaining to the acute and long-term management of KD describe a method of risk stratification for likelihood of coronary ischaemia to inform on ongoing cardiac investigation and antithrombotic therapy.²³ Although long-term treatment with aspirin and warfarin has been recommended for thrombotic prevention in the case of giant aneurysms (≥ 8 mm) (level C evidence),^{23 24} oral anticoagulation was not adopted in our patient despite her remaining LAD aneurysm, at the discretion of her regular cardiologist.

CONCLUSION

Thrombosis of a CAA, a sequela of KD, is an uncommon but important cause of acute STEMI, especially in young patients of Asian background. It provides a challenging management problem for interventional cardiologists in the emergency setting, particularly in the absence of randomised controlled

data and a lack of international consensus guidelines regarding optimal treatment strategies. On initial presentation, pharmacological therapy and emergency angiography should proceed as per standard STEMI management, although in the presence of known aneurysmal disease there may be a case for delaying the second antiplatelet agent until a decision regarding surgery is made. At angiography, acute management should be guided by coronary anatomy, thrombus burden, ongoing ischaemia and haemodynamic instability, in order to select an appropriate revascularisation strategy or conservative management. Early consultation with the cardiothoracic surgical team is essential.

Competing interests None declared.

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Learning points

- Acute thrombosis of a coronary artery aneurysm should be suspected in young patients presenting with ST-elevation myocardial infarction (STEMI), with or without a definitive history of Kawasaki disease.
- Percutaneous intervention may be challenging due to difficulty in passing guide wires across complex aneurysms, and may not be appropriate or feasible due to persistent thrombus burden, large aneurysm size and irregular morphology.
- Thrombus burden within aneurysms may be reduced by the use of intravenous or intracoronary antithrombotic agents and aspiration thrombectomy.
- Surgical revascularisation should be considered early after angiographic diagnosis especially in the setting of large, complex aneurysms, heavy thrombus burden and ongoing ischaemia despite attempted percutaneous coronary intervention.

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