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CASE REPORT

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# Asymptomatic giant right coronary artery aneurysm in Kawasaki disease: A case report

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**Abstract**

Incidentally diagnosed giant coronary artery aneurysms (gCAA) exceeding 50 mm in diameter are extremely rare and carry increased risks of cardiovascular morbidity and mortality. Currently, surgery is the preferred treatment for such gCAA.

**KEYWORDS**

coronary artery aneurysm (CAA), giant CAA (gCAA), Kawasaki disease (KD)

## 1 | INTRODUCTION

We discuss the case of a 30-year-old woman with an incidentally diagnosed giant coronary artery aneurysm of the proximal right coronary artery with associated aneurysms in the left main coronary artery and the ostium of the left circumflex artery.

Discrete coronary artery aneurysm (CAA) is characterized by a localized luminal dilatation with a diameter 1.5 times that of the normal adjacent segment.[1] Variable cut-off diameters have been proposed in the medical literature to define gCAAs, and aneurysms ≥ 50 mm in diameter are extremely rare.[2] In adults, atherosclerotic coronary artery disease is the predominant cause of gCAA followed by Kawasaki disease.[2,7] The majority of patients with gCAAs present with various cardiovascular complications or sudden death.[7,8] In the relatively fewer patients with incidentally diagnosed gCAAs, timely surgical intervention would prevent potentially fatal cardiovascular complications.[2,5,7-9,19]

## 2 | CASE PRESENTATION

A 30-year-old Ethiopian woman was evaluated for dry cough associated with dyspnea and wheezing of 1-week duration. She had experienced similar symptoms intermittently, usually precipitated by a common cold, for the past 10 years. She had no angina or symptoms of heart failure and had no history of cigarette smoking or illicit drug use. She could not recall any severe childhood illness and was not aware of a family history of chronic disease. On clinical evaluation, she had no significant positive physical findings.

Laboratory blood tests showed normal values of complete blood count, serum chemistry, fasting blood glucose, lipid panel, thyroid function, coagulation panel, erythrocyte sedimentation rate, and C- reactive protein level. Serology tests for hepatitis B and C, human immunodeficiency virus, Venereal Disease Research Laboratory, rheumatoid factor, and antinuclear antibody were all negative. Her erect posteroanterior chest X-ray (Figure 1) revealed a bulge on the right side of the heart, a prominent pulmonary bay, and an inconspicuous aortic arch. Transthoracic echocardiography (Figure 2) showed a mildly dilated right atrium, mildly increased PA pressure of 37 mm Hg, a 56 × 45 mm extracardiac mass with inner spontaneous echo contrast compressing the right atrium, and a left ventricular ejection fraction of 55%-60%.

The patient was subsequently referred abroad for further evaluation and possible intervention. A transesophageal echocardiogram (Figure 3) showed a 63 × 56 mm extracardiac cyst compressing a mildly dilated right atrium and having dense spontaneous echo contrast. A cardiac CT scan with contrast

(Figure 4) showed a well-defined 58 × 60 mm globular-like mass adjacent to the right atrium. The mass showed cystic characteristics and enhancement following intravenous contrast. A CT angiogram of the aorta and its branches (Figure 5) was remarkable for saccular aneurismal dilatation of the proximal right coronary artery (RCA) immediately distal to its origin, measuring 6.8 × 6.3 × 6.3 cm. The mid and distal segments of the RCA were normal in caliber and course. The left main coronary artery (LMCA) showed fusiform dilatation measuring 2.1 × 1.2 cm with extensive calcification. The aorta and its other main branches appeared normal with no evidence of aneurysm, dissection, or mural thrombus.

Coronary angiogram (Figure 6) revealed a giant RCA saccular aneurysm with maximum dimensions of 60.8 × 58.9 mm immediately adjacent to the ostium of the RCA and extending to the proximal and mid-RCA. The distal RCA was normal in caliber and course. The LMCA was aneurismal. The left anterior descending (LAD) coronary artery wrapped around the cardiac apex into the posterior interventricular groove, and its ostium was totally occluded, probably by aneurismal compression. The LAD and its diagonals filled through collaterals. The left circumflex coronary artery (LCX) ostium was aneurismal with distal LCX plaque. Left ventricular angiogram showed left ventricular ejection fraction of 60%, no regional wall motion abnormality, and no mitral insufficiency. An aortogram at LAO 45° revealed a giant saccular aneurysm adjacent to the ostium of the RCA without thrombus.

Histological examination of the resected aneurysmal sac showed patchy dense hyaline collagenous thickening and myxoid areas in the intima along with scattered small myxoid and connective tissue pools, suggesting medial degeneration with focal thinning and loss of the medial layer. There was no evidence of inflammation or vasculitis.

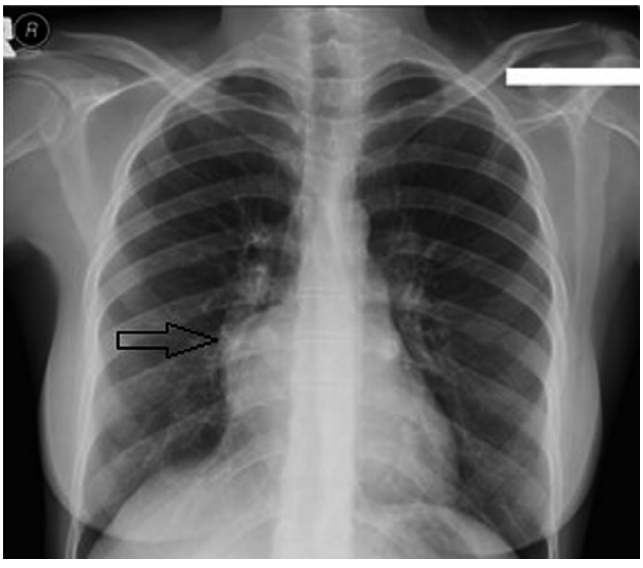

**FIGURE 1** Erect postero-anterior chest X-ray showing a bulge on the right side of the heart (arrow)

## 2.1 | Differential diagnosis

In adults, giant coronary artery aneurysm (gCAA) is predominantly atherosclerotic in origin. Other less common causes include connective tissue diseases, infections, vasculitis, Kawasaki disease, and congenital conditions. Our patient had no risk factors for atherosclerosis, history of connective tissue disease or vasculitis, and no evidence of systemic infection clinically or on workup. In addition, no supporting evidence for these etiologies of gCAA was revealed by serology tests, CT angiogram of the aorta and its branches, or coronary angiography. Coronary angiography presented no findings suggestive of a congenital source such as associated fistulous connections. Finally, histological examination of the resected aneurysmal wall did not reveal atherosclerotic changes, evidence of inflammation, or vasculitis.

Although the patient reported no childhood illness suggestive of Kawasaki disease, this disease can occasionally be manifested as a mild febrile episode, and the more characteristic mucocutaneous inflammation does not occur in all cases. Kawasaki disease is considered the most likely cause in our patient because of the multiple coronary arteries affected by aneurysms and the similar histology of the resected CAA to the late coronary histology demonstrated in Kawasaki disease complicated by CAA.

The patient's presenting respiratory complaints were attributed to bronchial asthma and allergic rhinitis.

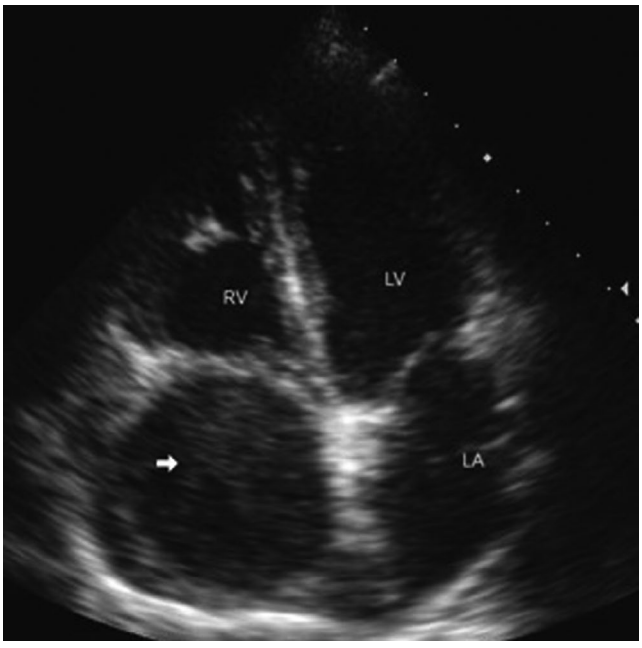


**FIGURE 2** Transthoracic echocardiography showing an extracardiac cyst with spontaneous echo contrast adjacent to and compressing the right atrium (white arrow). LA, Left atrium; LV, Left Ventricle; RV, Right Ventricle

## 2.2 | Treatment

The patient underwent open-heart surgery via median sternotomy and on partial bypass using aortic and single atrial cannulation technique. The RCA aneurysm was opened, J-shaped vertical aortotomy was done and the edges retracted, and the coronary ostia was identified and buttons dissected. The mid-RCA was anastomosed end-on-end to right saphenous vein graft (RSVG) using 8-0 prolene sutures. The left main aneurysm was opened, and the LCX was anastomosed end-on-end to RSVG

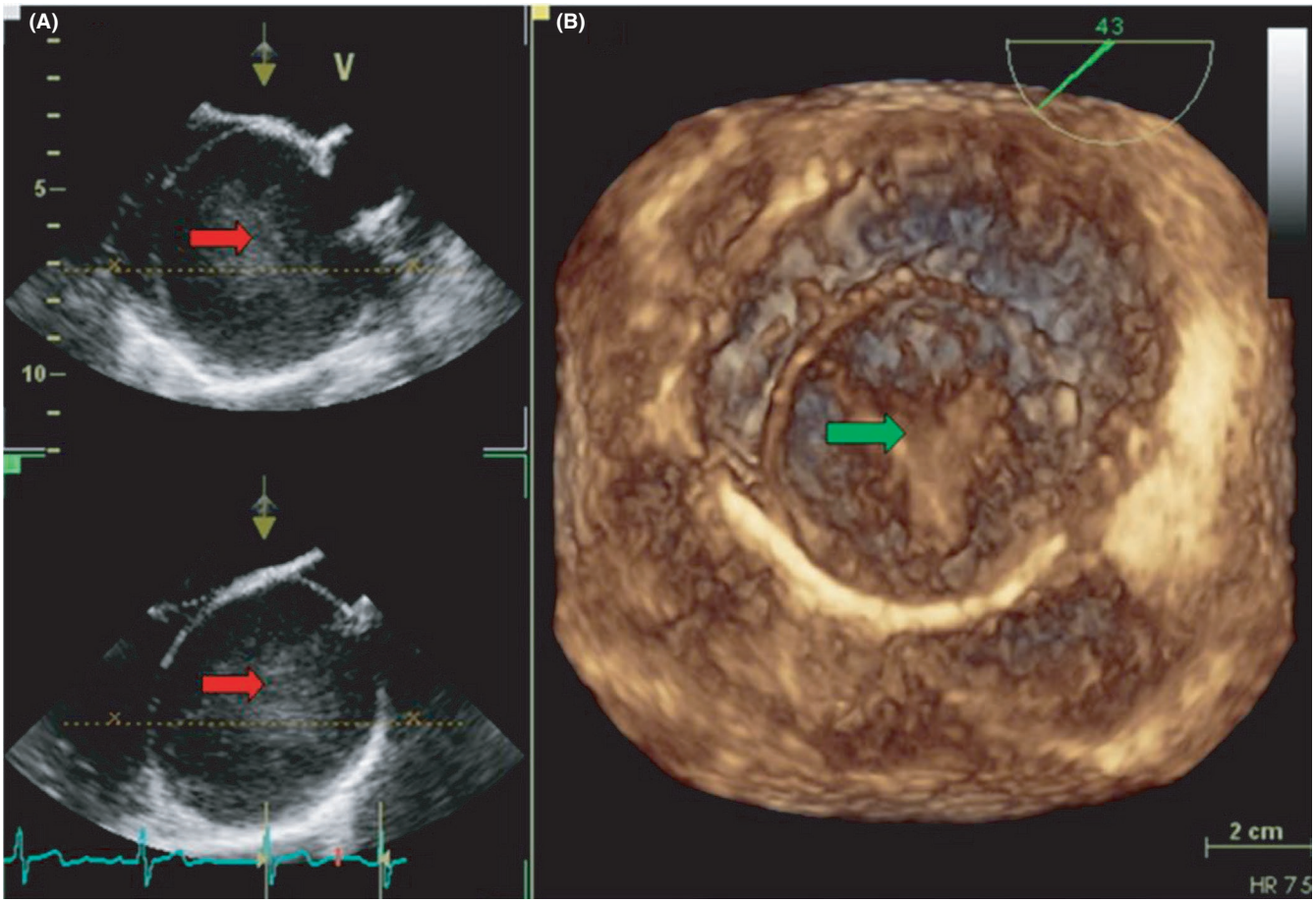


**FIGURE 3** Transesophageal echocardiogram showing an extracardiac cyst with dense spontaneous echo contrast (panel A, red arrows) and its corresponding three-dimensional image (panel B, green arrow)

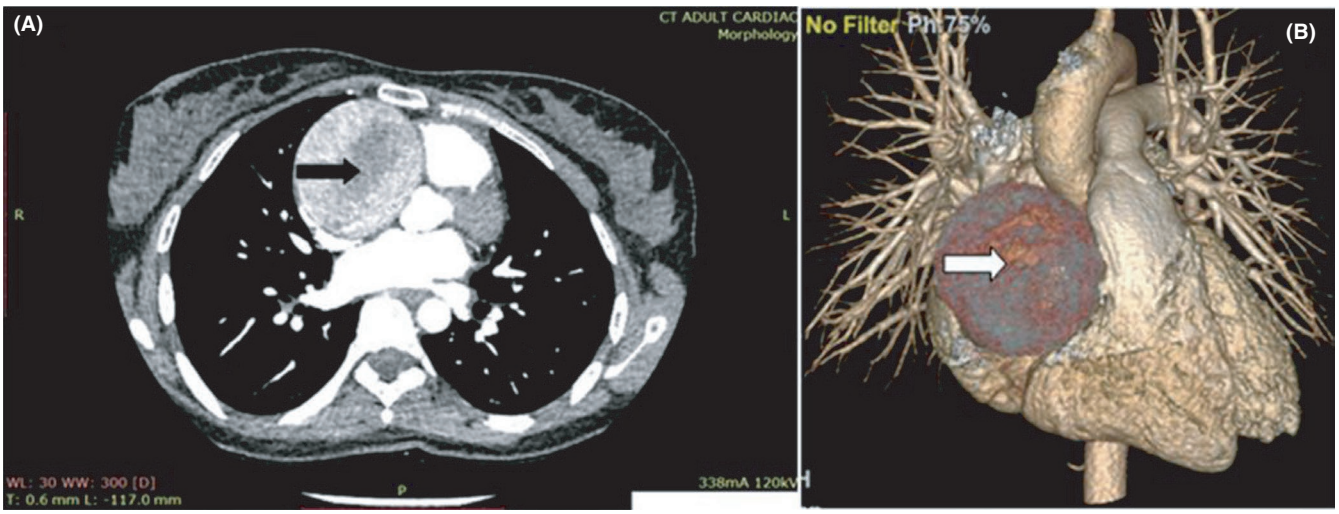


**FIGURE 4** Cardiac CT with contrast showing a well-defined, cystic globular-like mass adjacent to the right atrium (Panel A, black arrow). Three-dimensional reconstruction of cardiac CT images shows a giant right coronary artery aneurysm (Panel B, white arrow)

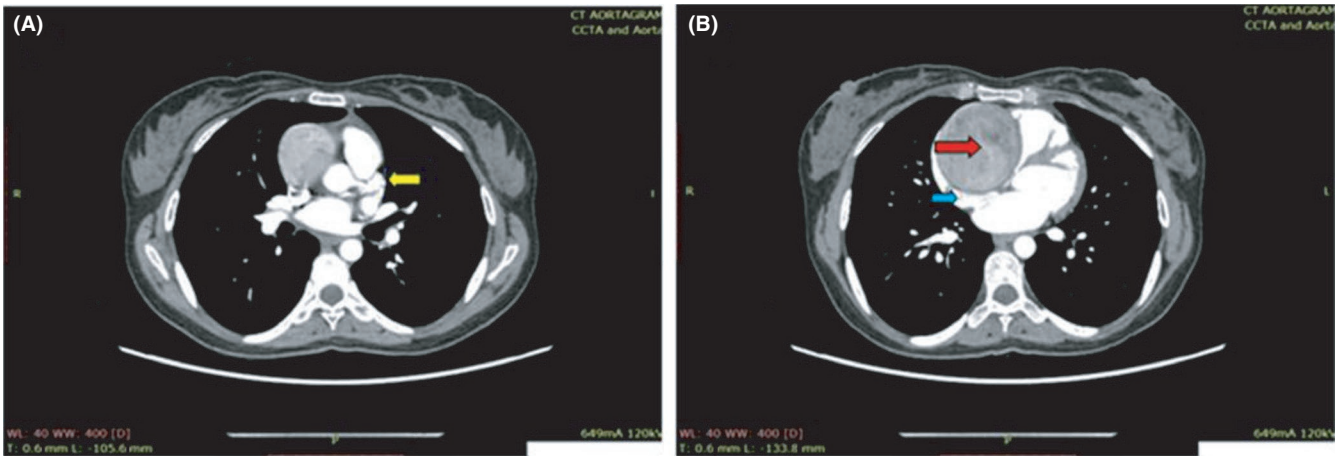


**FIGURE 5** CT angiogram of the aorta and its branches showing fusiform dilatation of the LMCA (panel A, yellow arrow) and a large saccular aneurismal dilatation of the proximal RCA (panel B, red arrow) compressing the right atrium (panel B, blue arrow)

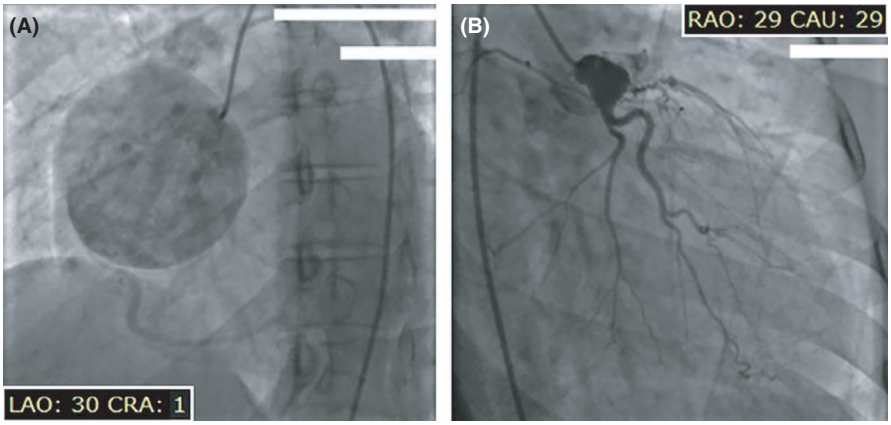


**FIGURE 6** Coronary angiogram showing a giant right coronary saccular aneurysm involving the proximal and mid-RCA (panel A) and aneurismal LMCA, LAD totally occluded at its ostium, and aneurismal LCX ostium (panel B)

using 8-0 prolene sutures. The left internal mammary artery was taken through A rent in the pericardium and anastomosed to the LAD using 8-0 prolene continuous sutures. The coronary ostia were then closed with pericardium. The aortotomy was closed in single layer using 4-0 prolene sutures. The proximal ends of RSVG were anastomosed to the ascending aorta.

Postoperatively, she was started on 150 mg oral aspirin daily, clopidogrel 75 mg orally daily for up to 1 year, atorvastatin 10 mg orally daily, montelukast 10 mg orally at night, and MDF Budamate (formoterol fumarate & budesonide) two puffs every 12 hours.

### 2.3 | Outcome and follow-up

Postoperative transthoracic echocardiogram showed left ventricular systolic function of 50%, mild to moderate eccentric tricuspid insufficiency jet, increased pulmonary artery pressure with pulmonary artery systolic pressure of 40 mm Hg, upper normal-sized right atrium, and no regional wall motion abnormality. The patient had an unremarkable postoperative hospital course and was discharged 2 weeks after admission.

One year postsurgery, the patient is well and has no limitations in her daily activities.

## 3 | DISCUSSION

The Coronary Artery Surgery Study (CASS) registry defines discrete coronary artery aneurysm (CAA) as a localized luminal dilatation with a diameter 1.5 times that of the normal adjacent segment.[1] There is no uniformly accepted definition of giant CAA (gCAA), and diameters exceeding 20, 40, 50 mm, and four times the reference vessel diameter have all been proposed in the medical literature.[2]

In the CASS registry, the largest angiographic study of its kind, comprising 20 087 patients, the combined incidence rate of discrete CAA and diffuse ectasia was 4.9%.[1] The overall incidence of discrete CAA in angiographic studies ranges from 0.2% to 2.2%, and the mean incidence rate from pooled analysis is 1.1%.[3,4] Giant CAAs are rare, with a reported prevalence of 0.02%-0.2%. Giant CAAs ≥ 50 mm in diameter are extremely rare with prevalence of 0.02%.[5,6] For the purpose of our case report, gCAA refers to an aneurysm diameter ≥ 50 mm. Giant CAAs are more prevalent in males than

in females, affect the proximal RCA in the majority of cases, and involve multiple coronary arteries in one of four cases.[7-9]

In adults, gCAA is predominantly atherosclerotic in origin.[2,7,9] Kawasaki disease (KD) is an acute inflammatory syndrome affecting children of all races throughout the world and is the second most common cause of CAA in adults.[10] It may result in acute vasculitis of the coronary arteries with infiltration by inflammatory cells, destruction of the internal elastic lamina, smooth muscle cell necrosis, and myointimal proliferation.[11] Disruption of the internal elastic lamina leads to subsequent coronary artery dilatation and aneurysm formation in around 10%-25% of untreated cases.[12] Treatment of KD with intravenous immunoglobulin lowers the rate of CAA formation to 3%-5%.[13] Coronary aneurysms in KD usually develop 6-12 months following the acute illness.[14] About half of these aneurysms subsequently regress, and approximately 1% of patients who recover from the acute illness develop gCAAs, coronary artery thrombosis, or stenosis.[15] Kawasaki disease can uncommonly manifest as a mild febrile episode in childhood, and coronary artery aneurysms may be incidentally discovered in adulthood as a late complication, as in our case.[16,17]

A review by Burns of the pathology of four hearts from young adults with late complications of childhood KD demonstrated that the vascular lesions of KD are different and distinct from atherosclerosis.[18] The late coronary histological changes of KD include intimal thickening, replacement of the medial smooth muscle cells with fibroblasts and extracellular matrix, and destruction of the internal elastic lamina.

Review of the literature revealed that, although CAAs are mostly asymptomatic, the majority of patients with gCAA present with cardiac symptoms.[8] A review by Keyser and colleagues reported 25% of the patients with gCAAs to have experienced no cardiac symptoms.[9] Patients may present with acute coronary syndromes, congestive heart failure, a mediastinal mass, compression of adjacent structures, sudden death, or complications such as thrombus formation, distal embolization, fistula formation, or rupture into surrounding structures.[6-10]

While gCAAs can be diagnosed with relative ease using noninvasive imaging modalities like echocardiography, CT angiogram, or magnetic resonance imaging, invasive coronary angiography remains the gold standard as it provides complete information regarding the size, shape, and location of the aneurysm as well as associated anomalies including coronary artery disease, multiple coronary aneurysms, and fistulous communications.[19] Occasionally, the initial finding may be an abnormal cardiac or mediastinal bulge on standard chest X-ray taken for other reasons, as in our case.

Because of the rarity of gCAAs, there are no standardized management guidelines, and current practice is based on reported experience rather than controlled trials.[7,8,20] A literature review suggests that surgical correction is generally accepted as the preferred treatment for gCAAs, since it allows excision of the aneurysmal sac and management of associated anomalies and prevents the aforementioned complications.[2,5,7-9,19] Surgical approaches include aneurysm resection with distal coronary artery bypass grafting, isolated coronary artery bypass grafting, aneurysm plication, and saphenous vein patch repair of the aneurysm.[2,5,7,21] Additional procedures including thrombectomy, fistula closure, and coronary artery reconstruction can be performed in patients with complications or concomitant lesions.

Less-invasive percutaneous interventions can be used to exclude CAAs in selected patients with suitable anatomy. Percutaneous techniques include stenting using Polytetrafluoroethylene (PTFE)-covered stents with or without coil embolization.[10,22,23] In a review by Szalat and colleagues comparing patients with CAA treated surgically (18 patients) vs via less-invasive percutaneous approach using PTFE-covered stents (24 patients), the restenosis rate was higher in stent patients who had CAAs larger than 10 mm than those with lesser diameters.[24] Accordingly, percutaneous exclusion of CAAs using PTFE-covered stents seems reasonable for CAAs with diameters between 5 and 10 mm; however, this review was limited by its small size, lack of controls, and lack of homogenous data.[10]

If surgical or percutaneous correction is not feasible, medical literature supports the use of antiplatelet and/or antithrombotic therapy for all large aneurysms in order to reduce the risk of in situ thrombus formation or distal embolization.[2,9,10,19]

No prospective data are available regarding postoperative anticoagulation regimens in adult patients who underwent surgery for gCAA in the late stages of KD. Treatment decisions depend on the extent and severity of coronary involvement and the type of surgical intervention. Therapeutic regimens include antiplatelet therapy with aspirin, with or without dipyridamole or clopidogrel; anticoagulant therapy with warfarin or low-molecular-weight heparin; or a combination of anticoagulant and antiplatelet therapy, usually warfarin plus aspirin.[25]

Two retrospective review studies of institutional databases of patients with KD complicated by large CAA have shown moderate long-term survival, although patients have a tendency to require multiple transcatheter or surgical interventions with increasing age.[26,27] The first of these studies showed cumulative coronary intervention rates of 33.7% at 20 years postonset of KD, and a second study showed 28%, 43%, and 59% cumulative coronary intervention rates at 5, 15, and 25 years postonset, respectively, with survival rates of 95%, 88%, and 88% at 10, 20, and 30 years, respectively.[28] Accordingly, the authors recommended careful follow-up of these patients, with routine

coronary angiography in order to allow timely intervention and to pre-empt complications.

To the best of the authors' knowledge, there is no published case report from Ethiopia of gCAA in an adult patient due to late complication of KD.

## 4 | CONCLUSIONS

Giant coronary artery aneurysms (gCAA) ≥ 50 mm in diameter are extremely rare and could be initially diagnosed as a late complication of Kawasaki disease (KD) in adults without apparent childhood illness suggestive of KD. They usually present with cardiovascular complications or sudden death. Surgery is the preferred treatment for gCAAs, even in the absence of symptoms.

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### CONFLICT OF INTEREST

None declared.

### AUTHOR CONTRIBUTIONS

DKB: contributed to collecting all the clinical data and writing of the draft manuscript. FC: contributed to literature review and revision of the manuscript.

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### REFERENCES

1. Swaye PS, Fisher LD, Litwin P, et al. Aneurysmal coronary artery disease. *Circulation*. 1983;67(1):134-138.
2. Crawley PD, Mahlow WJ, Huntsinger DR, Afiniwala S, Wortham DC. Giant coronary artery aneurysms: review and update. *Tex Heart Inst J*. 2014;41(6):603-608.
3. Hartnell GG, Parnell BM, Pridie RB. Coronary artery ectasia. Its prevalence and clinical significance in 4993 patients. *Br Heart J*. 1985;54:392-395.
4. Tunick PA, Slater J, Kronzon I, Glassman E. Discrete atherosclerotic coronary artery aneurysms: a study of 20 patients. *J Am Coll Cardiol*. 1990;15:279-282.
5. Li D, Wu Q, Sun L, et al. Surgical treatment of giant coronary artery aneurysm. *J Thorac Cardiovasc Surg*. 2005;130:817-821.
6. Ipek G, Omeroglu SN, Goksedef D, Balkanay OO, Basar I, Ayan F. Giant right coronary artery aneurysm associated with coronary-cameral fistula. *Tex Heart Inst J*. 2012;39(3):442-443.
7. Jha NK, Ouda HZ, Khan JA, Eising GP, Augustin N. Giant right coronary artery aneurysm. *J Cardiothorac Surg*. 2009;4:18.
8. Morita H, Ozawa H, Yamazaki S, et al. A case of giant coronary artery aneurysm with fistulous connection to the pulmonary artery: a case report and review of the literature. *Intern Med*. 2012;51(11):1361-1366.
9. Keyser A, Hilker MK, Husser O, Diez C, Schmid C. Giant coronary aneurysms exceeding 5 cm in size. *Interact Cardiovasc Thorac Surg*. 2012;15(1):33-36.
10. Abou Sherif S, Ozden Tok O, Taşköylü Ö, Goktekin O, Kilic ID. Coronary artery aneurysms: a review of the epidemiology, pathophysiology, diagnosis, and treatment. *Front Cardiovasc Med*. 2017;4:24.
11. Amano S, Hazama F, Hamashima Y. Pathology of Kawasaki disease: I. Pathology and morphogenesis of the vascular changes. *Jpn Circ J*. 1979;43(7):633-643.
12. Eleftheriou D, Levin M, Shingadia D, Tulloh R, Klein NJ, Brogan PA. Management of Kawasaki disease. *Arch Dis Child*. 2014;99:74-83.
13. Newburger JW, Takahashi M, Beiser AS, et al. A single intravenous infusion of gamma globulin as compared with four infusions in the treatment of acute Kawasaki syndrome. *New Eng J Med*. 1991;324:1633-1639.
14. Onouchi Z, Hamaoka K, Kamiya Y, et al. Transformation of coronary artery aneurysms to obstructive lesion and the role of collateral vessels in myocardial perfusion in patients with Kawasaki's disease. *J Am Coll Cardiol*. 1993;21:158-162.
15. Suzuki A, Kamiya T, Yasuo O, Kuroe K. Extended long term follow-up study of coronary arterial lesions in Kawasaki disease. *J Am Coll Cardiol*. 1991;17:33A.
16. Kawasaki T, Kosaki F, Okawa S, Shigematsu I, Yanagawa H. A new infantile acute febrile mucocutaneous lymph node syndrome (MLNS) prevailing in Japan. *Pediatrics*. 1974;54:271-276.
17. Porter GF, Gentles TL. Giant coronary-artery aneurysm in Kawasaki's disease. *N Engl J Med*. 2001;345:98.
18. Burns JC. Kawasaki disease update. *Indian J Pediatr*. 2009;76(1):71-76.
19. Pahlavan PS, Niroomand F. Coronary artery aneurysm: a review. *Clin Cardiol*. 2006;29:439-443.
20. Al Salihi S, Jacobi E, Hunter R, Buja M. Multiple giant coronary artery aneurysms: a case report. *Cardiovasc Pathol*. 2016;25:203-207.
21. Matsubayashi K, Asai T, Nishimura O, et al. Giant Coronary artery aneurysm in the left main coronary artery: a novel surgical procedure. *Ann Thorac Surg*. 2008;85(6):2130-2132.
22. Eshtehardi P, Cook S, Moarof I, Triller HJ, Windecker S. Giant coronary artery aneurysm: imaging findings before and after treatment with a polytetrafluoroethylene-covered stent. *Circ Cardiovasc Interv*. 2008;1(1):85-86.
23. Briguori C, Sarais C, Sivieri G, Takagi T, Di Mario C, Colombo A. Polytetrafluoroethylene-covered stent and coronary artery aneurysms. *Catheter Cardiovasc Interv*. 2002;55(3):326-330.
24. Szalat A, Durst R, Cohen A, Lotan C. Use of polytetrafluoroethylene-covered stent for treatment of coronary artery aneurysm. *Catheter Cardiovasc Interv*. 2005;66(2):203-208.
25. Newburger JW, Takahashi M, Gerber MA, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the committee on rheumatic fever, endocarditis and kawasaki disease, council on cardiovascular

disease in the young, American Heart Association. *Circulation*. 2004;110:2747-2771.

26. Bang JS, Kim GB, Kwon BS, Song MK, An HS, Song YW. Long-term prognosis for patients with Kawasaki disease complicated by large coronary aneurysm (diameter ≥6 mm). *Korean Circ J*. 2017;47(4):516-522.
27. Suda K, Iemura M, Nishiono H, Teramachi Y, Koteda Y, Kishimoto S. Long-term prognosis of patients with Kawasaki disease complicated by giant coronary aneurysms: a single-institution experience. *Circulation*. 2011;123:1836-1842.
28. Baman TS, Cole JH, Devireddy CM, Sperling LS. Risk factors and outcomes in patients with coronary artery aneurysms. *Am J Cardiol*. 2004;93:1549-1551.

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