




# A Giant Coronary Aneurysm in a Suspected Kawasaki Disease Causing Asymptomatic Myocardial Ischemia

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Indian J Radiol Imaging 2023;33:563–566.

## Abstract

Coronary involvement in Kawasaki disease is not uncommon; however, giant coronary aneurysm exceeding 50 mm is extremely rare. In this article, we presented a case of giant coronary aneurysm involving right coronary artery with associated asymptomatic myocardial ischemia as evident by multimodality imaging.

## Keywords

- ▶ coronary artery aneurysm
- ▶ giant aneurysm
- ▶ Kawasaki disease

## Introduction

Coronary involvement in Kawasaki disease (KD) is not uncommon, but giant coronary aneurysm due to KD is relatively rare with only around 25 cases reported worldwide. We present a case of giant coronary aneurysm involving right coronary artery (RCA) with associated asymptomatic myocardial ischemia as evident by multimodality imaging along with small aneurysms in other coronary arteries.

## Case Presentation

A 22-year-old student was found to have abnormal bulge in right heart border in X-ray chest while being evaluated for upper respiratory infection (▶**Fig. 1A**). Transthoracic echocardiogram (TTE) showed a cystic lesion anterior to right atrium for which he was referred to us. His clinical examination and electrocardiogram were normal. His TTE was suggestive of huge cystic mass (▶**Fig. 1B–E**; ▶**Videos 1, 2**) arising from RCA with flow within, likely coronary aneurysm. He also had ischemic features in RCA territory with hyperechoic myocardium in inferior and inferoseptal walls with mild

hypokinesia (▶**Fig. 1D**; ▶**Video 3**). Cardiac computed tomography (CT) confirmed the mass to be a giant proximal RCA aneurysm with another aneurysm at left main coronary artery bifurcation (LMCA) (▶**Fig. 2A, B**). In view of the aneurysm in coronary arteries, a detailed history from parents was obtained. Patient had prolonged fever and mucocutaneous rashes at 5 years of age, which was suggestive of KD. As patient was asymptomatic, cardiac magnetic resonance imaging (CMRI) was advised to rule out perfusion defect or myocardial scar. CMRI showed mild perfusion defect with subendocardial ischemia at inferior and inferoseptal walls at mid segment (▶**Fig. 2C–E**). Coronary angiogram was done to plan definitive management and it showed a huge aneurysm of proximal RCA without intraluminal thrombi. Additional aneurysms involving posterior descending artery (PDA; ▶**Fig. 3A, B**; ▶**Video 4**) and LMCA bifurcation were also noted (▶**Fig. 3C–E**; ▶**Videos 5–8**).

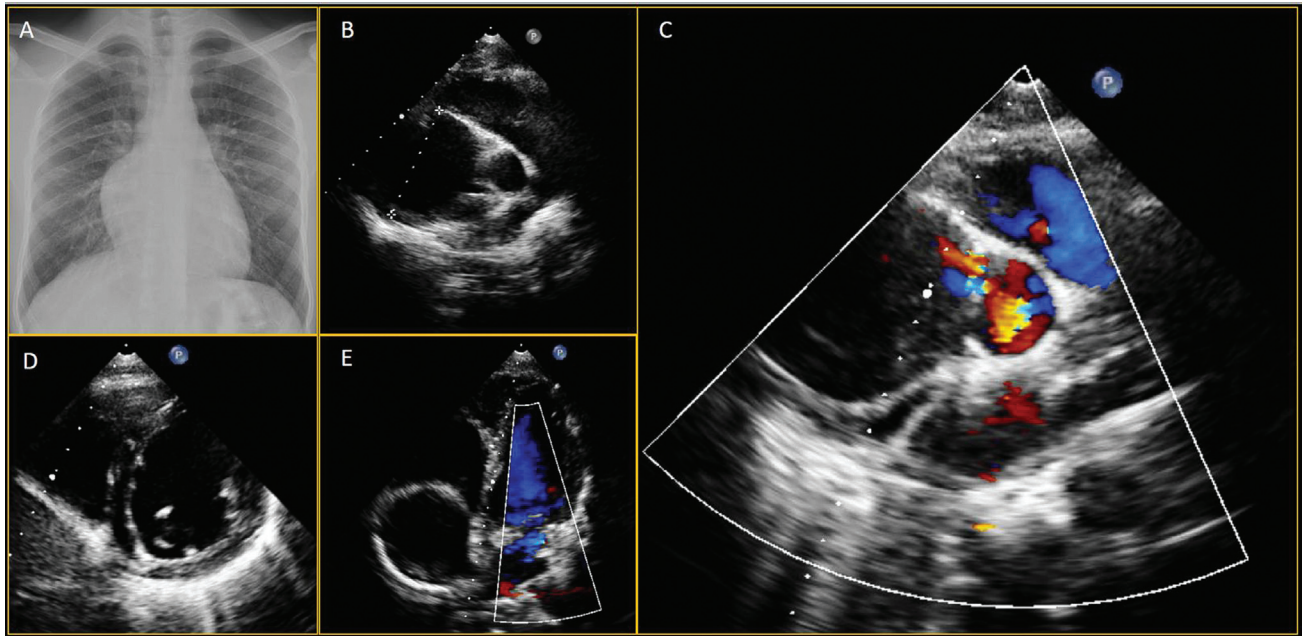
## Discussion

A giant coronary aneurysm is defined as the one with a Z-score  $\geq 10$  or a diameter  $>8$  mm for pediatric population or

article published online  
July 30, 2023

DOI <https://doi.org/10.1055/s-0043-1771363>.  
ISSN 0971-3026.

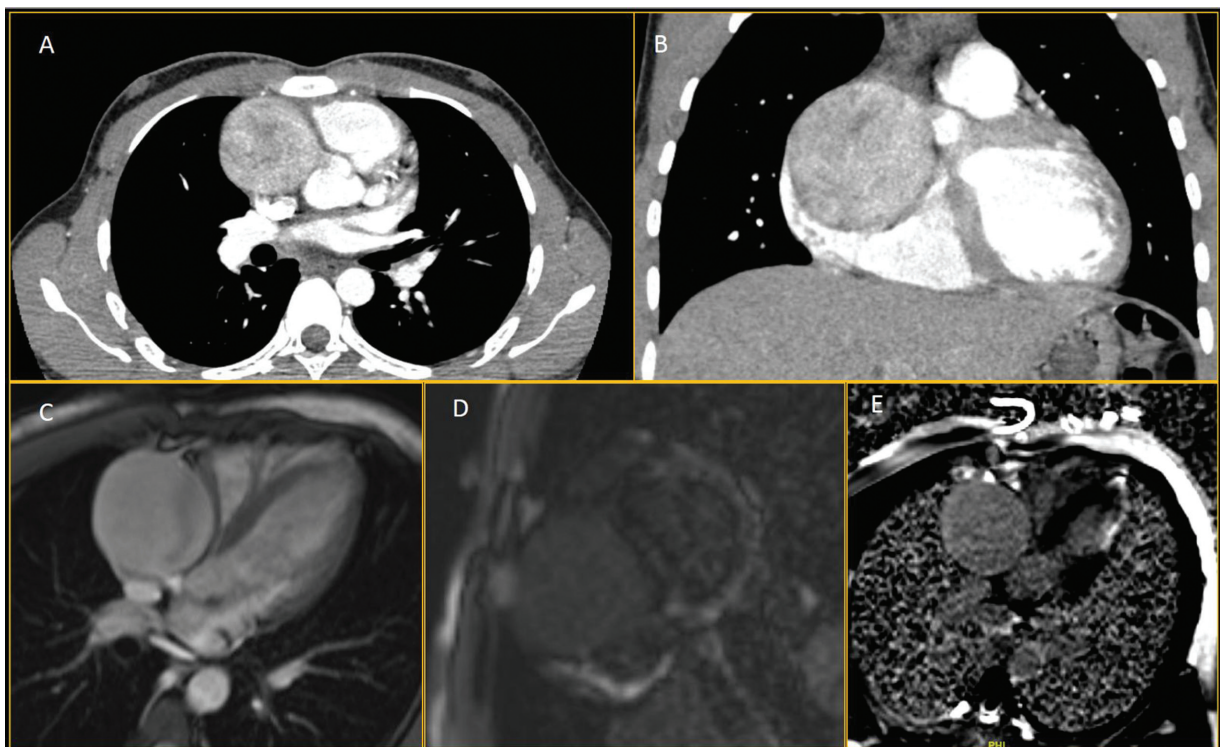
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**Fig. 1** (A) Chest X-ray showing right heart border bulging suggestive of paracardiac mass; (B, C) transthoracic echocardiogram basal short axis showing huge aneurysm just after origin of right coronary artery (RCA) abutting right atrium (RA) with color flow seen into the mass from RCA; (D) basal short axis showing hyperechogenic inferior and inferoseptal wall suggesting ischemic changes; (E) apical 5 chamber view showing huge mass abutting RA.

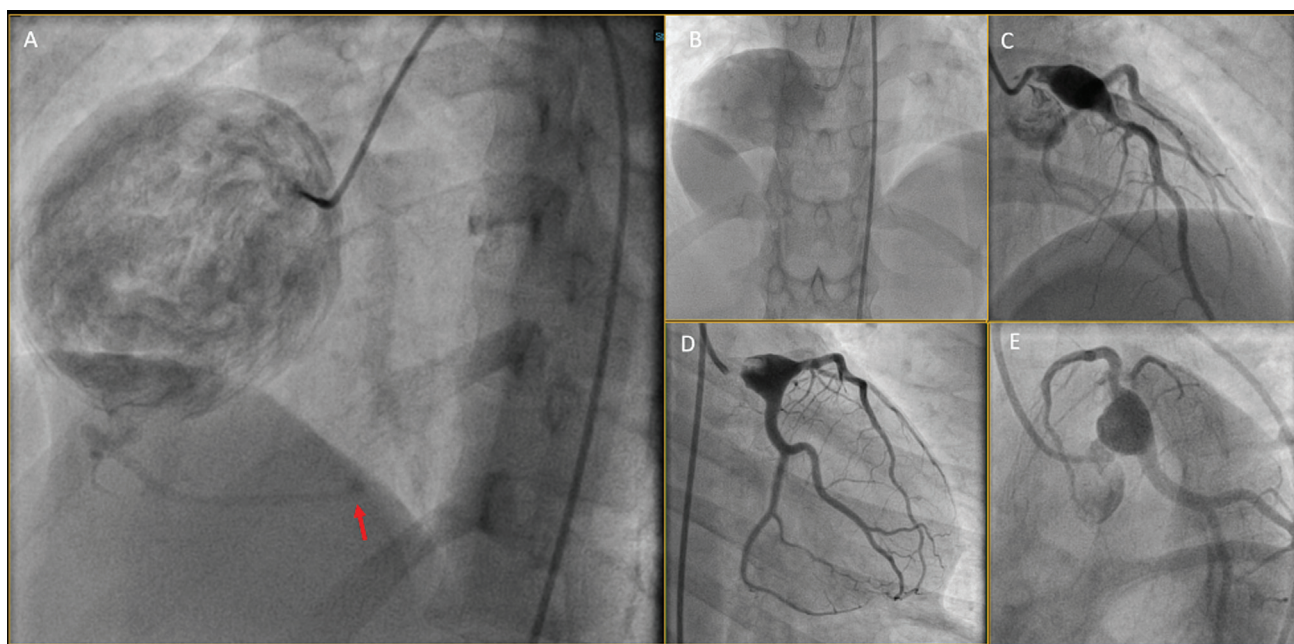
diameter exceeding the reference vessel diameter by  $>4$  times in adults.<sup>1</sup> Postulated mechanisms are destruction of arterial media, thinning of the arterial wall, increased wall stress, and progressive dilatation of the coronary artery segments due to high concentration of matrix metallopro-

teinases (1, 2, 3, 9, and 12). KD, coronary atherosclerosis, vasculitis, infections, congenital coronary cameral fistula, and connective tissue disorders are important factors contributing to aneurysm formation. In this case, a past history suggestive of KD, younger age of presentation, no



**Fig. 2** (A, B) Cardiac computed tomography showing huge right coronary aneurysm (67 × 69 mm) just after origin; (C) Contrast magnetic resonance imaging showing huge right coronary artery (RCA) aneurysm, distal RCA can also be seen distal to the aneurysm; (D, E) 15 minutes Late gadolinium scan showing subendocardial delayed enhancement in inferior and inferoseptal wall suggesting ischemia.





**Fig. 3** (A, B) Angiogram showing right coronary artery giant aneurysm just after origin, distal vessel likely filling normally, note the posterior descending artery aneurysm near crux (arrow); (C, D, E) Different angiographic view showing large left main coronary artery bifurcation aneurysm at point of confluence.

involvement of coronary ostium, absence of aortic calcification, and absence of markers of infection pointed toward KD as the likely etiology.

Kato et al first reported the development of coronary aneurysms after acute KD. Coronary artery sequelae, usually aneurysms, occur in 20 to 25% of cases.<sup>2</sup> Common sites of coronary aneurysms in KD are proximal left anterior descending artery and proximal RCA > LMCA > left circumflex artery > distal RCA. Regression is likely to occur within 1 or 2 years after onset, and it is unlikely to occur more than several years after onset. In a series of 26 patients with giant coronary aneurysms, 12 developed stenotic lesions and none showed regression. Risk factors for developing aneurysm are late diagnosis and delayed treatment, age <1 year and >9 years, male sex, long duration of fever ( $\geq 14$  days), failure to respond to initial immunoglobulin therapy, platelet counts <3 lakhs/mm, and low serum albumin.<sup>3</sup> In a large single-center study, Suda et al followed 76 patients with giant aneurysms and reported a survival rate of 88% at 30 years.<sup>4</sup> TTE may not be adequate to identify coronary artery abnormalities (CAAs) of KD that often require multimodality imaging.<sup>5,6</sup> Distal coronary aneurysms can be seen in patients with KD that are often missed on TTE as seen in our case (LMCA bifurcation and PDA aneurysm).<sup>7</sup> Limitation of TTE can be overcome by cardiac CT and CMRI that can clearly show coronary artery aneurysm, the distal coronary arteries, as well as calcifications and thrombus. CMRI can also evaluate myocardial fibrosis/viability. Options for managing coronary aneurysms are still ill-defined. There are no randomized trials to evaluate different management strategies and their outcomes. However, coronary artery bypass grafting (CABG) is preferred over percutaneous coronary intervention for giant aneurysms. Keyser et al recommended

surgery for super-giant CAAs > 50 mm.<sup>8</sup> In most studies, resection of the aneurysm and CABG is performed simultaneously.<sup>9</sup>

## Conclusion

On the background of fever and mucocutaneous rash in the childhood, a likely diagnosis of coronary aneurysm caused by KD was made.<sup>10-12</sup> Giant coronary aneurysm >65 mm in diameter due to KD is extremely rare with around 25 cases reported worldwide and possibly none from India till date. The index case has been planned for CABG with grafts to PDA with aneurysm ligation.

### Video 1

2D-Echocardiogram basal short axis view. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-1771363>.

### Video 2

2D-Echocardiogram apical 4-chamber view. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-1771363>.

**Video 3**

2D-Echocardiogram mid-segment short axis view. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-1771363>.

**Video 4**

Selective right coronary angiogram showing giant coronary aneurysm. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-1771363>.

**Video 5**

Left coronary injection in antero-posterior caudal view. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-1771363>.

**Video 6**

Left coronary injection in antero-posterior cranial view. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-1771363>.

**Video 7**

Left coronary injection in left anterior oblique cranial view. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-1771363>.

**Video 8**

Left coronary injection in left anterior oblique caudal view. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-1771363>.

**Note**

Patient consent for publication was duly obtained for this study.

**Funding**

None.

**Conflict of Interest**

None declared.

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