Myriad cardiac manifestation of Hyperhomocysteinemia- Case Report

Dr Jai Babu[1], DrPrashanth Kumar Malkiwodeyar[2], DrShivashankara Tarikere[3], DrCholenahalliNanjappa Manjunath[4]

ABSTRACT

Hyperhomocysteinemiacan present with myriad cardiac manifestation. Here we report a young gentleman, diagnosed as stroke in young, found to have dilated cardiomyopathy, with Left ventricular dysfunction and Hyperhomocysteinemia. Now presenting with unstable angina and found to have layered Left ventricular thrombus on echocardiography and spontaneous coronary artery dissection on angiography. Our patient is being followed up on optimal medical management, as he is asymptomatic with medications.

Key Words: Hyperhomocysteinemia, dilated cardiomyopathy, Spontaneous coronary artery dissection, Left ventricular thrombus.

[1, 2, 3] Resident, [4]Director, Department of cardiology

Sri Jayadeva Institute of Cardiovascular Sciences & Research, Bangalore, India.

Corresponding author mail: jaionly@gmail.com

Conflict of interest: None

INTRODUCTION

Hyperhomocysteinemia has been recognized as a risk factor for various cardiac diseases including thrombosis of arterial and venous system, spontaneous dissection involving various vessels in the body including coronaries, aneurysms, peripheral artery disease.[1] Here we report a 38 year old gentleman, diagnosed as stroke in young, diagnosed to have dilated cardiomyopathy, with Left ventricular dysfunction and peripheral artery disease and Hyperhomocysteinemia. Now presenting with unstable angina and found to have layered Left ventricular thrombus on echocardiography and spontaneous coronary artery dissection on angiography.

CASE REPORT

A 38 year old gentleman was diagnosed as stroke in young, cardiac evaluation revealed dilated cardiomyopathy with biventricular dysfunction in the past.

Serum homocysteine was elevated and was diagnosed to have hyperhomocysteinemia.

This time he presented with unstable angina, NYHA class III symptoms; patient was non-compliant with medications prescribed earlier. Echocardiography revealed dilated chambers, with layered Left ventricular thrombus, biventricular dysfunction with LV ejection fraction of 22% (Figure 1).

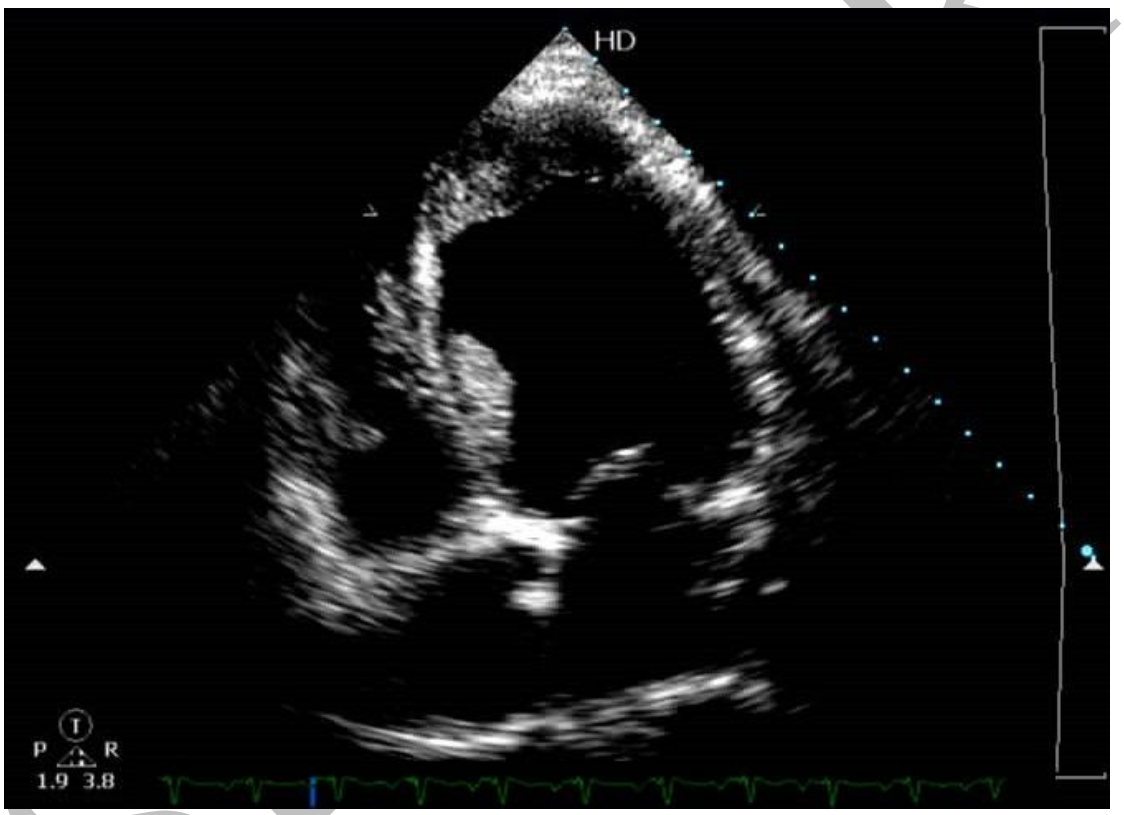

Figure 1: Echocardiography (Apical 4 chamber view) showing dilated chambers, with layered Left ventricular thrombus.

Coronary angiography showed spontaneous dissection in proximal Left Anterior Descending artery & distal Left circumflex artery (Figure 2). Patient is being followed up on optimal medical management & regular follow up.

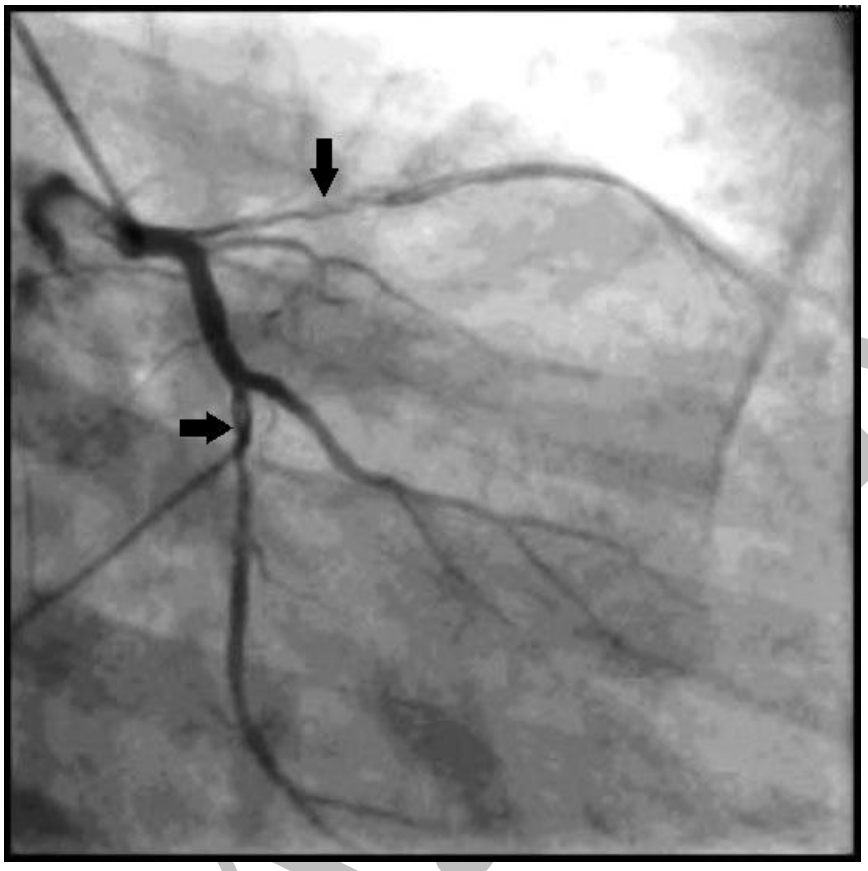

Figure 2: Coronary angiography showing spontaneous dissection in proximal Left Anterior Descending artery & distal Left Circumflex Artery

DISCUSSION

Hyerhomocysteinemia incidence is estimated to be 5% to 7% in general population; it is a known risk factor for various vascular disorders which includes coronary artery disease, venous & arterial thrombosis, peripheral vascular disease, & cerebrovascular disease.[1-2] Hyperhomocysteinemia is associated with raised serum levels of homocysteine in general with concentrations more than 14 µmol/L but with no elimination of homocysteine in the urine.[3]

Homocysteine is reflected to be noxious to vascular endothelium leads on to arterial intimal thickening and fibrous rich plaques in smooth muscle cells and collagen relatively than typical fatty plaques noted in atherosclerosis. Further homocysteine promotes endothelial injury, postulated to be

facilitated by pro-inflammatory changes, oxidative stress, and endoplasmic reticulum stress, which accordingly leads to endothelial cell injury and dysfunction.[4]Further hyperhomocysteinemia increases thrombotic tendency leading on to further endothelial cell injury, result in the early infarctions and death associated with this condition.[4]

Hyperhomocysteinemia is associated with arterial & venous thrombus formation. Left ventricular thrombus formation can be seen to be independently associated with hyperhomocysteinemia & LV dysfunction.[5] Hyperhomocysteinemia can even present as left atrial thrombus in sinus rhythm, pedunculated left ventricular mass in different case reports reported in literature.[6-7] Our patient had layered left ventricular thrombus on echocardiography, which has not been reported in literature to our knowledge. Although biventricular dysfunction with reduced ejection fraction may be an antecedent factor in the cause of thrombus formation, nonetheless homocysteinemia cannot be ruled out as one of the causative factor in this case.

There have been numerous causes associated with spontaneous coronary artery dissection like atherosclerotic plaque, peripartum period, connective tissue disorders (Marfan's syndrome), vasculitis, after intense exertion, blunt chest trauma, cocaine abuse, hyperhomocysteinemia& idiopathic. [8-12] Hyperhomocysteinemia is related to spontaneous dissection involving aorta (aortic aneurysm), coronary, intra & extra cranial arteries & peripheral arteries. [9, 13-16]

Spontaneous coronary artery dissection in coronary arteries are most frequently located in the left anterior descending coronary artery (45-75%), followed by the right coronary artery (20-33%), left circumflex coronary artery (4-19%) and left main coronary artery involved in < 1%. Left coronary artery dissections are more common in women and right coronary artery dissections occur more frequent in men. [9-10, 17] Spontaneous coronary artery dissection can present as acute coronary syndrome – unstable angina, non ST elevation myocardial infarction, ST-elevation myocardial infarction, or chronic

stable angina.[18] And can even present as refractory congestive heart failure with attempt to bridge to cardiac transplantation.[19-20]

In our case, patient had spontaneous dissection in proximal Left Anterior Descending artery & distal Left circumflex artery. This being unique case report in that, both spontaneous dissection of coronary arteries present at the time of clinical presentation with unstable angina.

Recognized guidelines are not present to the treatment of patients with spontaneous coronary artery dissection at present. Treatment options include optimal medical management & revascularization with CABG or PCI. Medical management includes antiplatelets, anticoagulation, nitrates, and beta-blockers.[21-22] Revascularisation is contemplated with ongoing ischemia and is frequently treated with percutaneous intervention or surgery and is individualized.[9]

Our patient is being followed up on optimal medical management, as he is asymptomatic with medications. He may be considered for myocardial perfusion imaging for inducible ischemia in future for possible coronary revascularisation.

CONCLUSION

Myriad manifestations of Hyperhomocysteinemia have been reported in the literature. We have reported this case as it was unique with layered left ventricular thrombus on echocardiography and spontaneous coronary artery dissection on angiography. The current case illustrates the relationship of increased levels of homocysteine and the hypercoagulable syndrome. Layered Left ventricular thrombus was managed with anticoagulants. Spontaneous left main coronary artery dissection is a rare event and has mostly been managed by CABG. In our case, patient was asymptomatic and was managed with optimal medical management.

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