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Aortic Dissection in Young: Multiple Rare Risk Factors in a Single Patient

[1]Dr. Sithara Subair, MBBS*, [2]Prof. Dr. P Paranthaman, MD
[1]Junior Resident, [2]Professor and Head of department
Dept of General medicine, Govt Kilpauk medical college and hospital, Chennai

***Corresponding Author:**
Dr. Sithara Subair
MBBS, Junior Resident, Dept of General medicine, Govt Kilpauk medical college and hospital, Chennai

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ABSTRACT

Introduction: We describe a case of acute aortic dissection with bicuspid aortic valve, with no Marfanoid features, with accelerated premature atherosclerosis, secondary to elevated homocysteine and lipoprotein(a) levels, which hasn't been described previously in literature.

Case information: A 44 year old male, hypertensive, smoker, alcoholic presented with a history of a sudden severe sharp tearing interscapular pain lasting a few seconds, that radiated to his chest and right lower limb, following which the patient had claudication and paraesthesia in right lower limb for 15 days. He was clinically stable with echocardiogram showing an atherosclerotic flap in arch of aorta, calcified biscuspid valve, aortic stenosis and regurgitation; CT angiogram and CT abdomen with contrast showed an infrarenal aortic dissection with proximal propagation to arch of aorta. Diffuse atherosclerosis was noted in all visualised vessels. On further investigating causes of accelerated atherosclerosis, we found a highly elevated homocysteine (>50 micromol/L), elevated lipoprotein(a) (155.0 mg/dl), a borderline d-dimer (0.60 mcg FEU/ml) levels with reduced protein S activity (31%).

Conclusion: The raised homocysteine and lipoprotein(a) led to an accelerated premature atherosclerosis and dissection, compounded by a bicuspid valve with stenosis and regurgitation, in a young male and such an association hasn't been previously described.

Keywords: case report, aortic dissection, hyperhomocysteinemia, bicuspid aortic valve, dissection in young, young adult.

INTRODUCTION

The most common causes of acute aortic dissection in a young patient are bicuspid aortic valve, Marfans syndrome and Ehler Danlos syndrome. Atherosclerosis as a factor was found only in 1% of young patients according to a study[1], whereas homocysteine hasn't been associated at all, except in Marfans syndrome. Herein we describe a case with a bicuspid aortic valve, with no Marfanoid features, along with accelerated premature atherosclerosis, secondary to elevated homocysteine and lipoprotein(a) levels, which hasn't been described previously in literature.

CASE REPORT

A 44 year old farmer, previously in good health, presented with complaints of pain in right lower limb for past 15 days that worsened on walking a few steps. He described it as cramping type of pain, which diminished on rest. At the onset of pain 15 days ago, he had been hospitalised with a history of sudden severe sharp tearing interscapular pain which lasted few minutes, radiating down towards chest and right lower limb, accompanied by palpitations and syncope. These symptoms subsided within few hours

and he was referred to our hospital for further evaluation and management.

He is a known hypertensive, diagnosed 4years ago, but not on any medications currently. No history of other comorbid conditions. There is no history of rheumatic heart disease, no history of cardiac or sudden deaths in family. He is physically active with a history of regularly lifting heavy weights. He has a history of 5 pack years cigarette smoking, has weekly binges of alcohol but doesn't use cocaine or other recreational drugs.

On presentation to our emergency, patient was conscious and oriented. He was well built and of average height. He was not pale, no evidence of cyanosis, or clubbing of digits is seen. There is no pedal edema or other evidence of volume overloaded state. Muddy conjunctiva seen, but no icterus observed. No obvious features of Marfan's or Ehler Danlos seen. Pulse rate was 62 beats per minute and regular. Radial pulse was felt on both sides equally with a 'water hammer' or collapsing quality. Right lower limb had feeble femoral, popliteal, dorsalis pedis, anterior tibial and posterior tibial artery pulsations, while all pulsations were felt well in left lower limb with a pistol shot quality on auscultation of femoral artery. Patient's blood pressure was measured to be 140/70 mmHg on right upper limb and 150/80 mmHg on left with a wide pulse pressure of 70 mmHg. Left lower limb had a pressure of 160/70mmHg, while right lower limb pressures were not recordable. He had a respiratory rate of 16 per minute, room air saturation using pulse oximetry 98% and capillary blood glucose 147mg/dl.

On detailed cardiovascular examination, apical impulse was undisplaced, with a heaving character. A midsystolic harsh murmur of grade 3 was heard on auscultating the aortic area with patient in sitting and leaning forward position, in full expiration. A soft diastolic murmur heard in neoartic area. Second heart was soft, no third or fourth heart sounds were heard, no ejection click or opening snap heard. Systolic bruit heard in both carotids. No abdominal bruit heard. Patient had prominent pulsations seen in jugular notch and both carotids. Locomotor brachii was observed on both arms in semiflexed position. Dilated veins were observed over superior vena cava territory.

Clinically we narrowed down differential diagnoses for severe interscapular and chest pain associated with absent pulse in right femoral, which included aortic dissection with reduced blood flow to right lower limb causing claudication, acute coronary syndrome with an an embolus to right femoral, or acute pulmonary embolism with deep vein thrombosis, requiring us to rule out thrombophilias.

Electrocardiogram showed features of left ventricular hypertrophy by voltage criteria, no changes suggestive of ACS seen. Echocardiogram showed an atherosclerotic flap in arch of aorta, calcified bicuspid aortic valve with aortic stenosis and regurgitation. Concentric left ventricular hypertrophy was seen with grade 1 diastolic dysfunction. Contrast enhanced CT abdomen and angiogram showed aortic dissection in the infrarenal abdominal aorta for a length of about 4.8cm, extending to bilateral common iliac arteries seen, with reduced attenuation in right common and external iliac arteries. Coeliac artery, superior mesenteric, right renal artery and inferior mesenteric arteries arose from true lumen, whereas left renal artery arose from false lumen. No evidence of aneurysms, other organ involvement or free fluid seen. Doppler scans showed normal renal artery resistive indices on both sides, and no thrombosis or stenosis. Routine investigations were normal except for an elevated ESR and mild leukocytosis.

Owing to the young age of the patient, we considered the bicuspid valve to be one of the causes of dissection. He also had risk factors of chronic smoking and hypertension, and the presence of atherosclerotic flap in echo as well, prompting us to consider accelerated premature atherosclerosis as another possibility. We looked into blood parameters which promote atherosclerosis and thrombosis. We found a highly elevated serum homocysteine of >50 (normal range 5.46-16.40 micromol/L), an elevated lipoprotein(a) levels of >155 (normal <20 mg/dl), a borderline d-dimer value of 0.60 (normal <0.50 mcg FEU/ml) and reduced protein S activity (31%). Protein C activity was normal and there was no factor V Leiden mutation detected. Patient remained clinically stable and endovascular stent grafting and aortic valve replacement was planned for him.

DISCUSSION

Aortic dissection overall has an incidence of 2.6 to 3.5 per 100,000 person-years. There is a male

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preponderance, with mean age of around 63 years. Risk factors for aortic dissection in a young patient include Marfan's, seen in 50%[2], Ehler Danlos syndrome, bicuspid aortic valve [3], preexisting aneurysm (19% of patients), aortic coarctation, prior aortic surgery. Only 1% of young patients with dissection had atherosclerosis according to a study[1]. Familial thoracic aortic aneurysm and dissections (TAAD) [3], strenuous activity, cocaine use, arteritides (Giant cell, Takayasu, syphilitic, rheumatoid), Turner's syndrome and pregnancy in females, trauma and flouroquinolone use [4] are other risk factors.

Prevalance of bicuspid aortic valve is around 1% of population, with a male preponderance. Presence of bicuspid valve was associated with 9-fold increased risk of dissection[7], while prevalence of bicuspid valve in population with aortic dissection is 4-12%[1]. Genetic inheritance patterns maybe autosomal dominant with variable penetrance, and have been associated with Turners, Loeys-Dietz syndrome and familial TAAD.

Patients with bicuspid aortic valve develop aortic stenosis more frequently than regurgitation. They are at risk for aortic dilation and dissection, caused by underlying aortopathy with cystic medial degeneration. There is increased apoptosis, more intense elastic fragmentation, and higher matrix metalloproteinases[7]. Increased collagen related stiffness and decreased fibrillin-1 content has been described. Abnormal hemodynamic effects and associated regional aortic wall shear stress from bicuspid aortic valve flow patterns.

Disease associations of hyperhomocysteinemia include cardiovascular and cerebrovascular diseases, but unlike other modifiable risk factors, lowering homocysteine will not prevent cardiovascular events. Also a risk factor in premature arteriolosclerosis, venous thromboembolism. It is not a known risk factor for aortic dissection except in Marfan's syndrome. Homocysteine is known to weaken the elastic fibres of vessel walls, and lipoprotein(a) promotes atherosclerosis, while in patients with a bicuspid aortic valve, there is evidence of aortopathy with cystic medial degeneration along with the effects of abnormal flow patterns and shear stress.

CONCLUSION

Accelerated atherosclerosis secondary to hyperhomocysteinemia, increased lipoprotein(a) along with risk factors of hypertension and smoking, along with preexistent bicuspid aortic valve, caused the development of acute aortic dissection, of type B which proximally propagated to type A. Fortunately the patient is clinically stable despite having combined type A and B dissections. Such a case has never been described before in literature.

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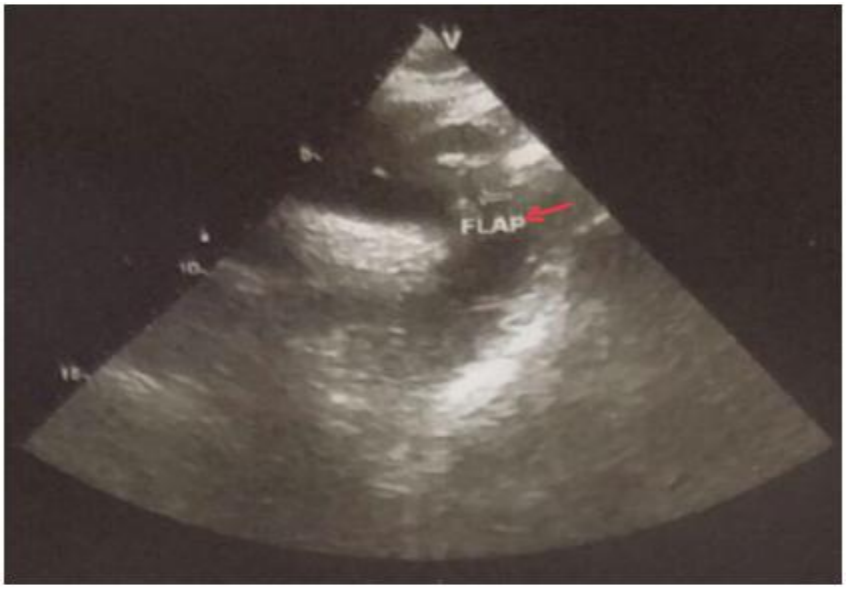
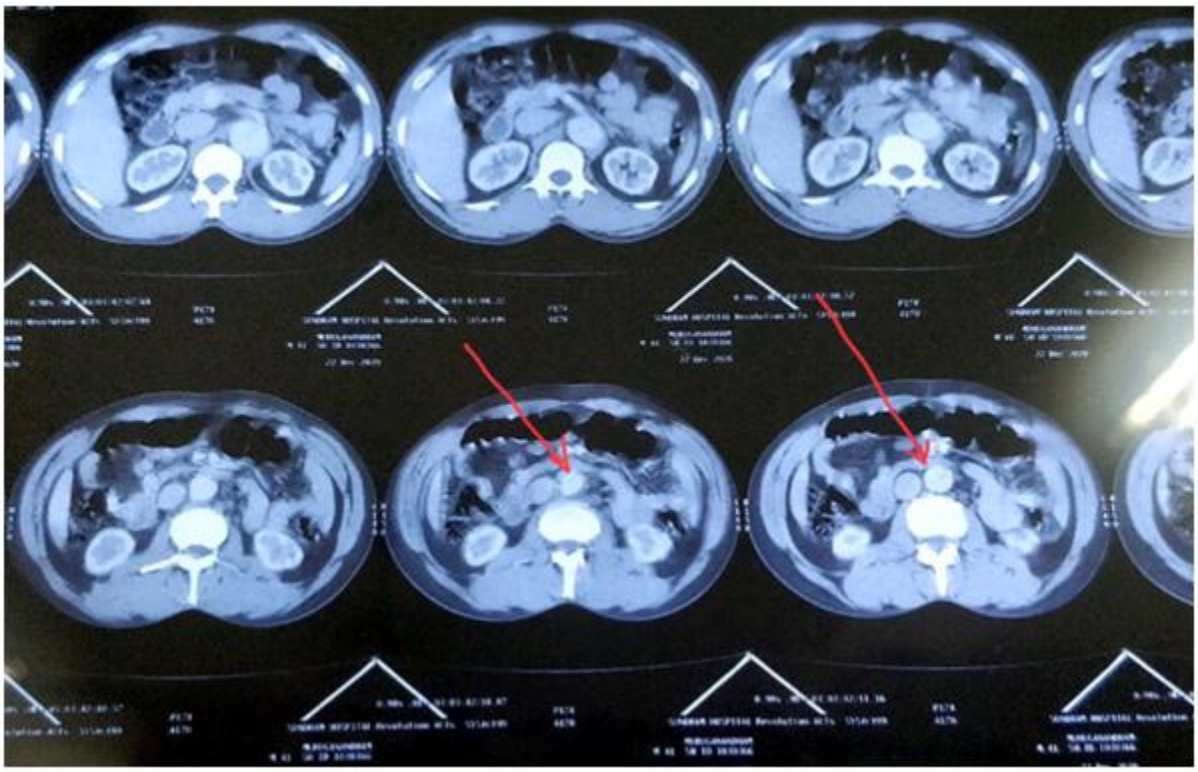


Echocardiogram showed atherosclerotic flap in arch of aorta. Calcified bicuspid aortic valve seen with aortic stenosis and regurgitation. Concentric LV hypertrophy with normal LV systolic function and grade 1 diastolic dysfunction.



(A)

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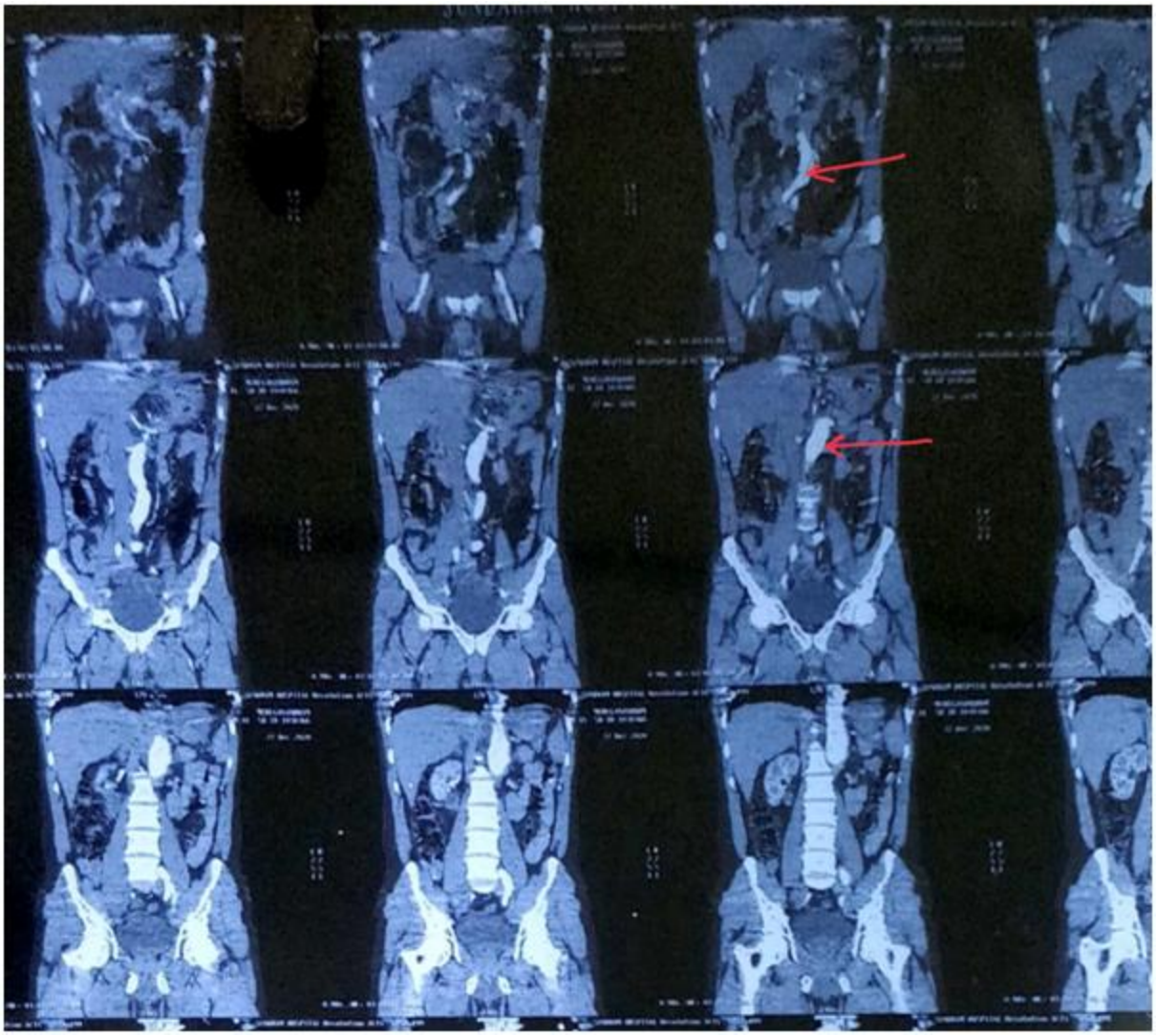

(B)

CT abdomen with contrast showing aortic dissection in abdominal aorta, extending to bilateral common iliac arteries. Celiac, SMA, right renal artery, IMA arise from true lumen. Left renal artery from false lumen. (A) axial view (B) coronal view

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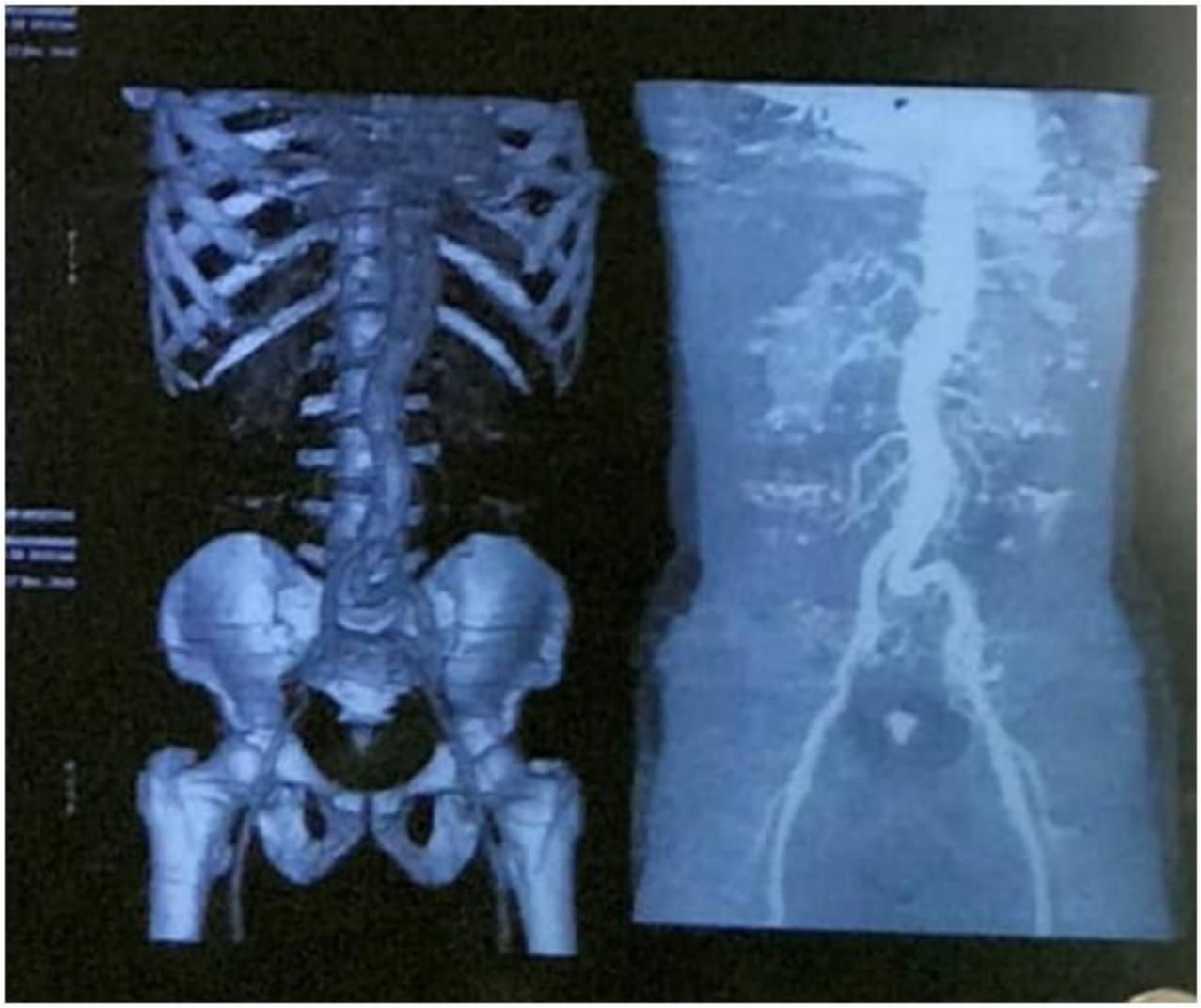

CT angiogram 3D reconstruction showing abdominal aortic dissection with reduced attenuation in right common and external iliac arteries. Diffuse atherosclerotic changes in all visualised vessels. Infrarenal aorta is involved for a length of 4.8cm, with extension into bilateral common iliac arteries for a short length.

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