ASCENDING AORTIC DISSECTION IN THE DEVELOPING WORLD; CASE REPORT

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ABSTRACT

Aortic dissection is frequently a life threatening condition and is the most common emergency of the acute aortic syndromes. Left untreated or poorly treated; the mortality rate has been reported to increase by 1-3% per hour after presentation and is approximately 36-72% within an hour of diagnosis and 62-91% within one week. Various contributory factors related to human resources, technical resources, socio-economic constraints and other utilities in poor income countries with poor health indices also worsen the prognosis of acute aortic dissection. We have made an attempt to remind ourselves of the continued existence and challenges in managing this disease.

KEYWORDS: *Aortic Dissection, Mortality, Outcome, Underdeveloped*

INTRODUCTION

Aortic dissection occurs when a tear in the aortic intima exposes the underlying media to the hydrodynamic forces of blood within the aortic lumen leading to dissection within the media which may propagate anterogradely or less commonly retrogradely [1]. A false lumen is created by blood filling the space within the media between the intimal flap and the adventitia.

Ascertaining the exact incidence of aortic dissection is difficult because many patients die before the condition is recognized. The early mortality rate in patients with acute aortic dissection is very high, with up to a 1% per hour death rate reported in the first several hours before surgery for type A dissection [2].

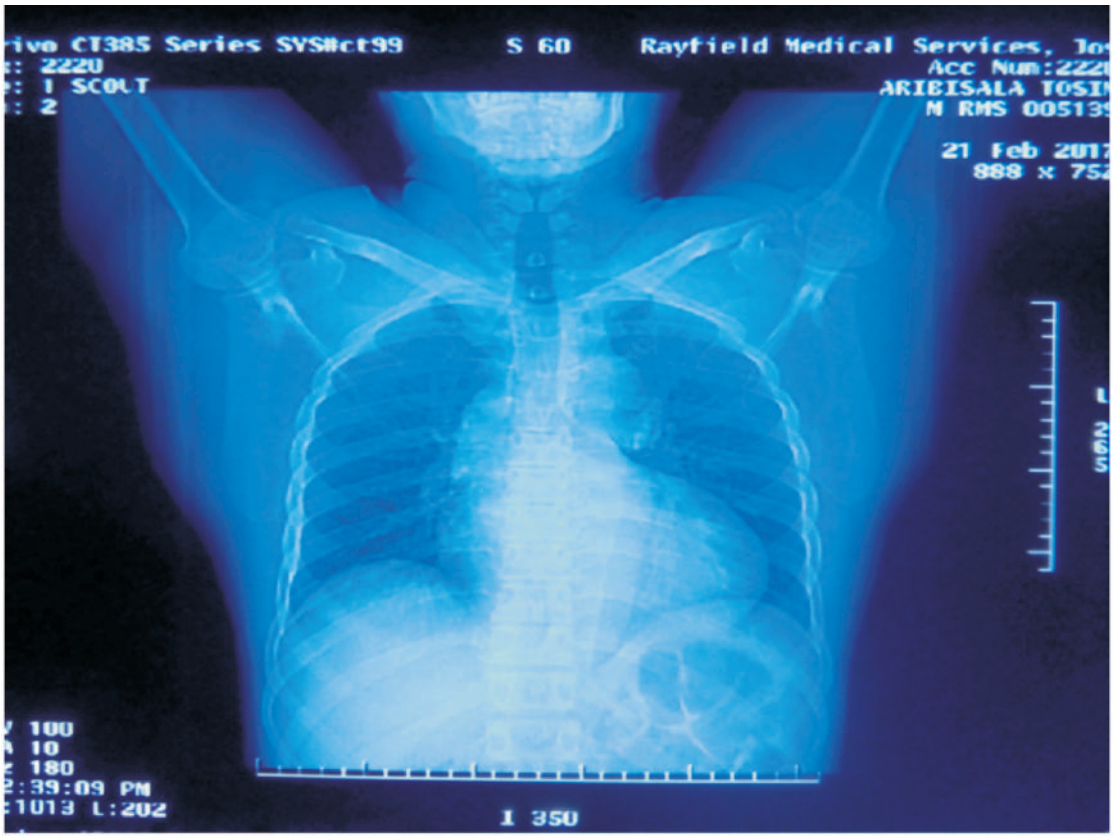
We report two cases of aortic dissection seen in the Jos university teaching hospital in February 2017.

CASE ONE

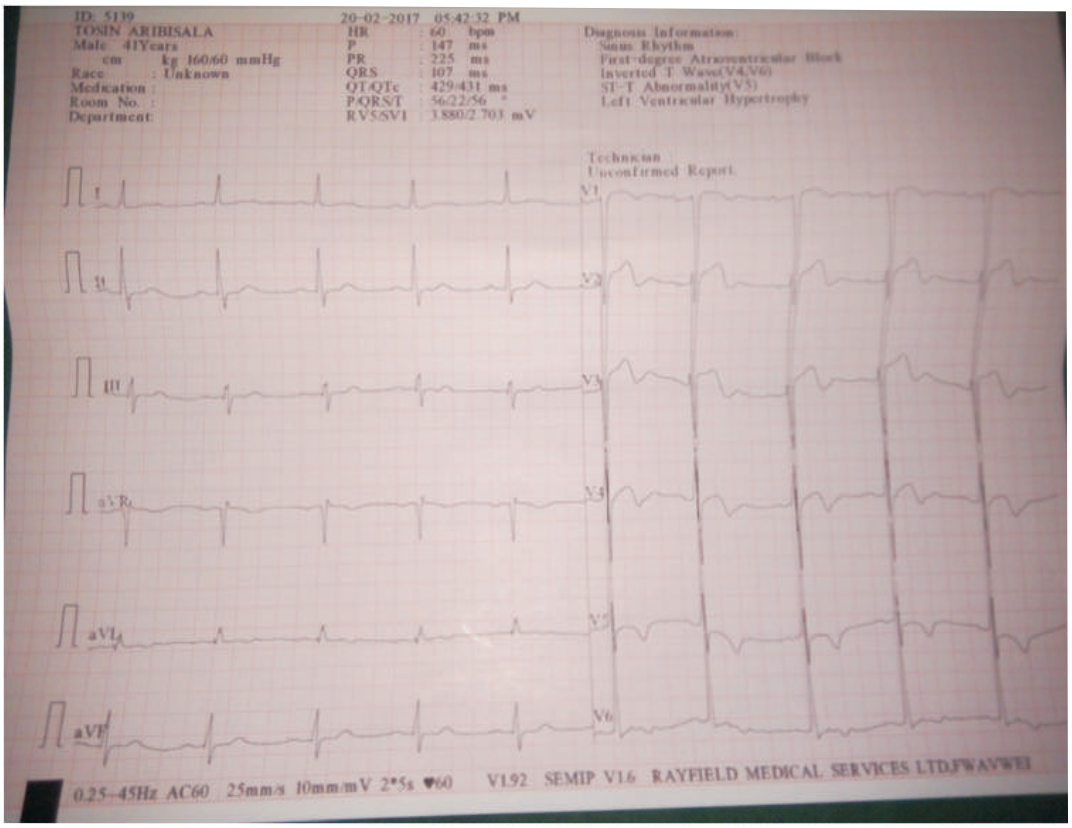
The first was Mr T.A, a 41 year old male Engineer and known hypertensive diagnosed eleven years prior to presentation but not compliant on medications, whopresented with a 4 hour history of sudden severe sharp retrosternal chest pain radiating to the back. There was associated breathlessness, a feeling of impending doom and electrical shocking sensation on the back and both lower limbs. No cough, diaphoresis, palpitation or intermittent claudication. He had an episode of severe sudden sharp retrosternal chest pain months before this episode. His mother is hypertensive but there was no family history of sudden death or stroke. He had 75mg of I.M diclofenac before presentation.

On examination, he was not in obvious painful distress, no cyanosis, no pedal oedema. Pulse rate was 60 bpm full volume and regular, the radial artery was thickened, BP was 160/60 mmHg on the right arm and 150/50 mmHg on the left arm. Apex beat was at the 6LICS, lateral to the midclavicular line and heaving. He had a normal S1, soft S2, 3/6 regurgitant murmur in the aortic area with an Austin Flint murmur at the apex. Fine crepitations were heard at the left middle and lower lung zones posteriorly.

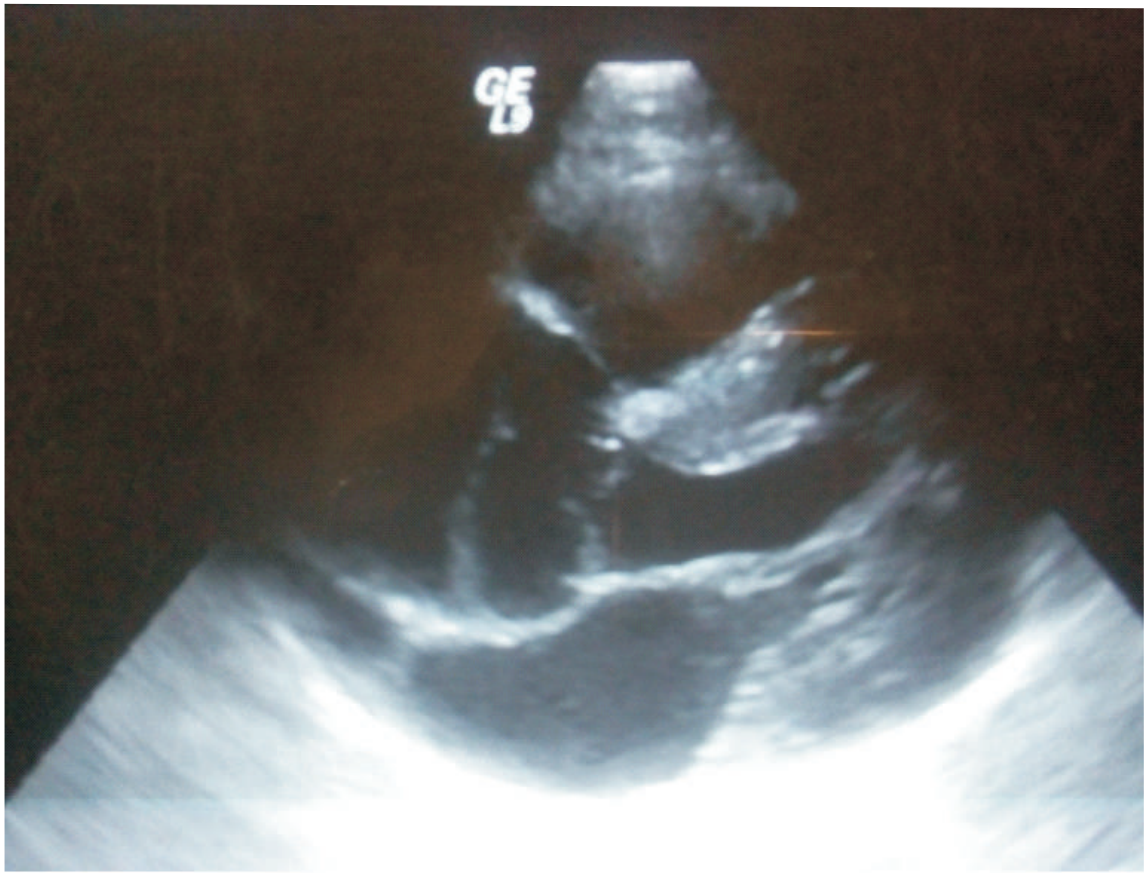
Chest X-ray (Fig 1) revealed cardiomegaly with CTR of 0.75 and widened/unfolded aortic silhouette.



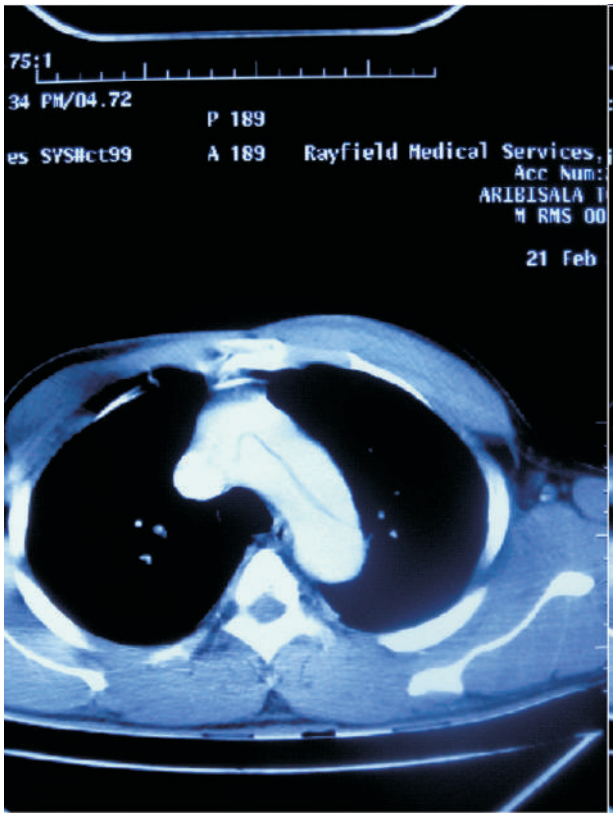
ECG (Fig 2) showed first degree AV block, left ventricular hypertrophy and T wave inversion in V4-V6.



Echo (Fig 3) revealed a dilated aortic root (Ao 48mm), severe aortic regurgitation, a mobile dissecting flap around the sinus of valsava and thickened interventricular septum and LV posterior wall.



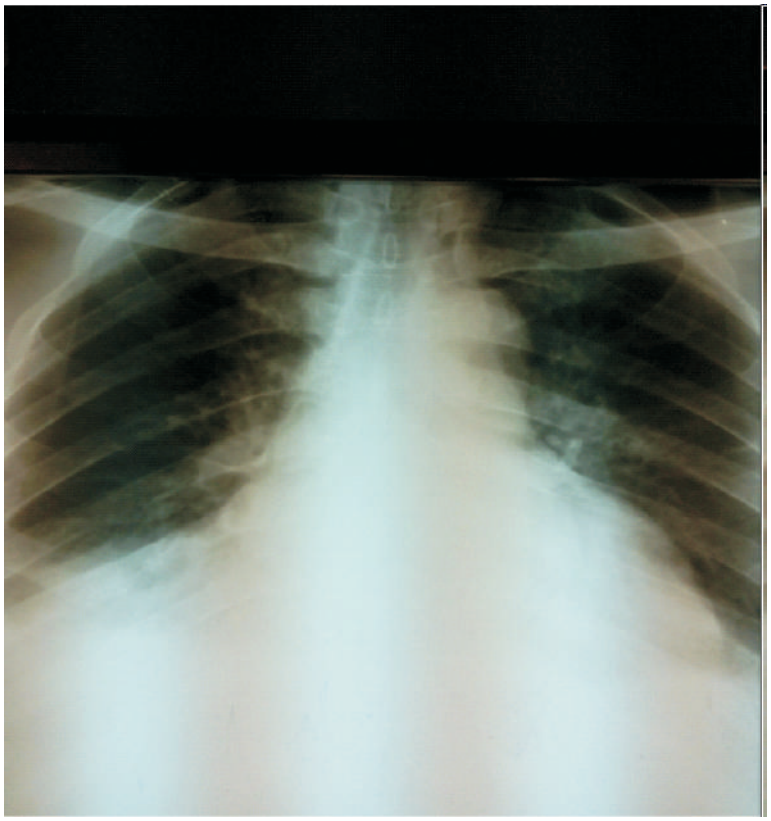
CT angiography (Fig 4,5,6,7) showed an extensive intimal flap in the ascending aorta up to the tip of the left subclavian artery.



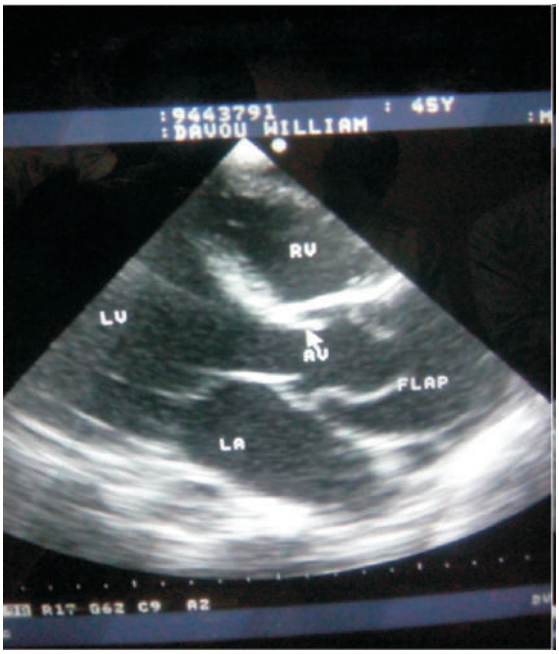
A diagnosis of Type Aortic dissection was made and he was placed on metoprolol, frusemide, lisinopril and referred to the cardiothoracic/vascular surgeon.

CASE TWO

Mr D.W, a 45 year old man who presented with abrupt sharp chest pain with breathlessness and cough. Chest X ray showed dilated aortic silhouette and increased CTR.



Echo revealed a dilated aorta (30mm at the root and 53mm at the bulb). There was a swinging intimal flap in the ascending aorta with prolapse of the aortic cusps into the LVOT. The left ventricle was dilated (LVIDD 69mm) and hypo contractile.



CT angiography could not be done due to financial constraints.

DISCUSSION

Population studies in the United States have estimated the incidence of aortic dissection to range from 2 to 3.5 cases per 100,000 person years [3]. In Sweden, the incidence of aortic dissection in men is reported to be 16 per 100,000 yearly [3]. In autopsy series, the prevalence of aortic dissection ranges from 0.2 to 0.8 % [4]. Aliyu and coworkers reported a case of intramural aortic haematoma in an 11 year old girl with Marfan's syndrome in Kano, Northwest Nigeria [5]. Sule AZ et al reported a case series of infrarenal abdominal aortic aneurysm (without dissection) in Jos, Nigeria [6].

Aortic dissection is rare in individuals younger than 40years of age [7]. Ascending aortic dissection occurs most commonly in individuals between 50 and 60 years of age while descending aortic dissections are more commonly encountered in older individuals with a peak at 60 to 70 years of age [8]. The typical aortic dissection patient is a male in his sixth decade of life. However, the presentation may be variable and a high index of suspicion should be entertained. It is an uncommon condition and several other conditions predispose the aorta to dissection. Most resulting from disruption of the normal architecture and integrity of the aortic wall.

The interplay of three factors are thought to be responsible for the pathogenesis of most cases of aortic dissection [9]. First is an abnormality or weakening of the aortic media which may occur with aging, in some congenital cardiovascular anormalies and with the fibrinillopathies. The second is an agent of intimal injury or tear as observed in atherosclerosis or hypertension. And the third is the systemic blood pressure that is responsible for a pressure head that drives blood to dissect the aortic wall [9]. Hypertension occurs in approximately 72% of all patients who suffer aortic dissection [10]. Hypertension causes alterations in the elastic properties of the arterial wall and increases stiffness thereby predisposing to aneurysm or dissection. However, hypertension alone is not usually associated with significant aortic root dilation [8]. Cystic medial degeneration commonly underlies aortic dissection and several genetically triggered disorders of connective tissue like Marfan's syndrome, Loeys-Dietz syndrome, familial Thoracic Aortic Aneurysm syndromes and vascular Ehlers-Danlos syndrome [11]. Apart from hypertension, the patient did not have features suggestive of these genetically triggered disorders like skeletal or craniofacial abnormalities. The complication of aortic dissection in Marfan's syndrome however, usually presents between 30-50 years of age [12] ,the age group into which these patients fall. Syphilitic aortitis can cause aortic dissection but the clinical features suggesting primary, secondary or tertiary syphilis were lacking in these patients and the short/acute/emergent presentation of the first case didn't give room for investigating syphilis as a probable cause of the dissection. None of the patients volunteered history of ingestion of cocaine of other psychosocial drugs. Cocaine abuse (particularly crack cocaine) accounts for less than 1% of cases of aortic dissection [8].

Other disorders associated with aneurysm and dissection include Bicuspid or unicuspid aortic valve, Noonan syndrome, supravalvular aortic stenosis, aneurysm-osteoarthritis syndrome, aberrant right subclavian artery (Komerrell diverticulum), right sided aortic arch, polycystic kidney disease and Alport's syndrome in males [3].

The presentation of aortic dissection is that of sudden onset chest or back pain that migrates as the dissection progresses [13]. Both cases had abrupt onset chest pain as reason for presentation. Although tearing is the classic description, the pain is often described as sharp. Aortic dissection can also be painless in about 10% of patients especially those with neurologic complications from the dissection, those with Marfan's syndrome, diabetes mellitus, previous aortic aneurysm or prior cardiac surgery [9,13]. Patients can present with hypertension or hypotension (due to cardiac tamponade or haemopericardium). The first case had hypertension but the second case did not have hypertension neither were there echo features suggestive of pericardial effusion. He however had a dilated heart that was hypo contractile and unlikely to sustain an elevated blood pressure. Acute congestive failure related to ascending aortic dissection generally results from acute severe aortic regurgitation which both cases had.

Despite our ability to recognize this life threatening condition, the absence of such facilities like transoesophageal echocardiography and lack of facilities for appropriate surgical intervention and the cost of such intervention if sourced abroad are severe limiting factors to the management of this condition in our setting.

There is the need to improve on the level of service delivery which will reduce the need for medical

tourism and the preventable death sentence(s) meted on most patients who come down with this condition in our setting.

REFERENCES

1. Solomon D. Scot. In Essentials of echocardiography. Human Press. Totowa, New Jersey. 2006.
2. LeMaire SA, McDonald ML, Guo DC et al: Genome-wide association study identifies a susceptibility locus for thoracic aortic aneurysms and aortic dissections spanning FBNI at 15q21.1. Nat Genet 43:996,2011
3. Hiratzka LF, Bakris GL, Beckman JA et al: 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/ST/SVM guidelines for the diagnosis and management of patients with thoracic airtic disease: A report of the American College of Cardiology Foundation/American Heart Associatin Task force on practice guidelines, American Association for Thoracic surgery, American College of Radiology, American stroke Association, Society of cardiovascular Anaesthesiologist, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Soceity of Thoracic surgeons and society for vascular medicine. Circulation 121;e266,2010.
4. Tsai TT, Trimachi S, Nienaber CA: Acute aortic dissection: Perspectives from the International Registry of Acute Aortic Dissection(IRAD). Eur J Vasc Endovasc Surg 37:149,2009.
5. Aliyu I, Akhiwu HO. Intramural aortic haematoma in an 11 year old girl with Marfan's syndrome. Nig Card J 2014;11(2):139-141
6. Sule AZ, Ardill B, Ojo EO. Abdominal aortic aneurysm and the challenges of management in a developing country: A review of three cases. Anals Afri Med J. 2012:11(3);176-181.
7. Ngan KW, Hsueh C, Hseih HC, Ueng SH. Aortic dissection in a young patient without any predisposing factors. Chang Gung Med J 2006:29:419-12.
8. Rodney HF, Ray EH. In Douglas LM, Douglas PZ, Libby P, Bonow R. editors. 10th edition. Braunwald's Heart Disease. Elsevir Saunders. 2015;1572.
9. Edwin F, Tetty MM, Frimpong-Boateng K. eComent: Acute Aortic dissection in children and young adults- the role of silfenafil. Interact Cardiovasc Thorac Surg 2009;9:143
10. Hagan PG, Nienaber CA, Isselbacher EM et al: The International Registry of Acute Aortic Dissection(IRAD): New Insights into an old disease. JAMA 283:897.2000
11. Doyle JJ. Gerber EE, Dietz HC: Matrix-dependent perturbation of TGFbeta Signalling and disease. FEBS LeH 586:2003,2012.
12. Ruiz ME, Sty JR, Wells RG. Aortic Dissection in a 5-year old girl with Marfan's syndrome. Arch Pediatr Adolesc Med 1996;150:440-2
13. Petre R, Von Segesser LK. Aortic dissection. Lancet 1997:349:1461-4