CASE REPORT

Journal of Indonesia Vascular Access (*INAVA*) 2023, Volume 3, Number 2: 19-22
E-ISSN : 2798-6780 ; P - ISSN : 2807-7032

Published By : IVAA
the Indonesian Vascular Access Association

Conservative management for type A chronic thoracic aortic dissection on a patient with Marfan Syndrome, "when surgery can't be done": a case report from the rural hospital

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Received: 2022-09-30
Accepted: 2023-11-02
Published: 2023-12-05

ABSTRACT

Introduction: In aortic emergencies, aortic dissection (AD) is one of the most common. This fatal condition affected 2-3 per 100,000 people per year. Predisposing factors initiate the tear through the aorta and create the false lumen. Chronic aortic dissection is often associated with complications that make the aortic wall become weak and can lead to the rupture of the aorta. Chronic aortic dissection also can lead to an ischemic syndrome as a result of gradual impairment of distal perfusion. Emergency surgical intervention in elderly patients with type A aortic dissection is still controversial because of the high postoperative mortality and complication rate. This study aims to discuss the case of a patient who was already diagnosed with chronic aortic dissection with underlying thoracic aortic aneurysm and Marfan syndrome who gets treated conservatively.

Case Description: We report a case of a 54 years old female who came to the emergency department with the complaint of crushed-felt-like chest pain for the last 3 days accompanied by coughing, nausea, vomiting, and epigastric pain. The patient had been diagnosed with aortic dissection with a thoracic aortic aneurysm two months ago with a history of uncontrolled chronic hypertension and she was also diagnosed with Marfan syndrome. The patient was treated conservatively due to the high risk of mortality and morbidities for cardiac surgery.

Conclusion: The decision regarding treatment options for patients with chronic aortic dissection should be considered the outcomes, mortality, and physical function. The physician needs to inform the patient and family about the condition, treatment options, long-term medical management, and the prognosis of the disease.

Keywords: Aortic Dissection, Aortic Aneurysm, Marfan Syndrome, Conservative Management.
Cite This Article: Abolla, S.M., Saputra, C. 2023. Conservative management for type A chronic thoracic aortic dissection on a patient with Marfan Syndrome, "when surgery can't be done": a case report from the rural hospital. *Journal of Indonesia Vascular Access* 3(2): 19-22. DOI : 10.51559/jinava.v3i2.43

INTRODUCTION

In aortic emergencies, aortic dissection (AD) is one of the most common. This fatal condition affected 2-3 per 100,000 people per year. Predisposing factors initiate the tear through the aorta and create the false lumen. The anatomical extent of the dissection is used to classify aortic dissection using Stanford and De Bakey classification. Aortic dissection is also classified into acute and chronic. An aortic dissection that presents in 14 days is termed acute aortic dissection and if the dissection presents after 14 days termed chronic aortic dissection. This type of dissection is often associated with complications that make the aortic wall become weak and can lead to the rupture of the aorta. Chronic aortic dissection also can lead to ischemic syndrome as a result of the gradual impairment of distal perfusion.[1]

The decision to choose between surgical or conservative management for elderly patients with type A aortic dissection should be considered the outcomes, mortality, and physical function. Emergency surgical intervention in elderly patients with type A aortic dissection is still controversial because of the high postoperative mortality and complication rate.[2] There are just a few pieces of literature that concern the management of chronic thoracic aortic dissection therefore available evidence on medical management of chronic aortic dissection is lacking.[3] We discuss the case of a patient who was already diagnosed with chronic aortic dissection with an underlying thoracic aortic aneurysm and Marfan syndrome who gets treated conservatively.

CASE DESCRIPTION

A 54-year-old female came to the emergency department complaining of her crushed-felt-like chest pain for the last 3 days and accompanied by coughing, nausea, vomiting, and epigastric pain. A history of occasional back pain, palpitation, shortness of breath, and fatigue for almost 4 years was also reported. In terms of past medical history, she suffered from chronic uncontrolled hypertension and two months ago was diagnosed with Aortic Dissection with a thoracic aortic aneurysm.

The patient has been treated with 5

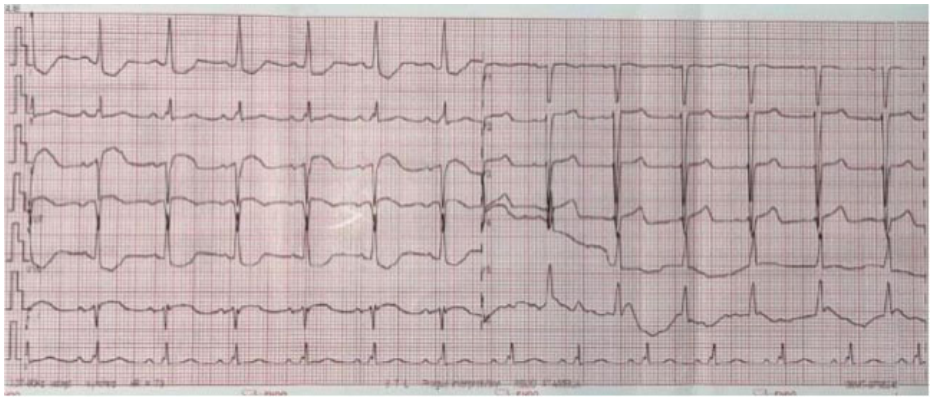

Figure 1. 12-leads ECG on admission showed showed sinus rhythm, heart rate 75 bpm, poor r wave progression, ST Elevation in lead III & aVR, and ST depression in lead I & aVL.

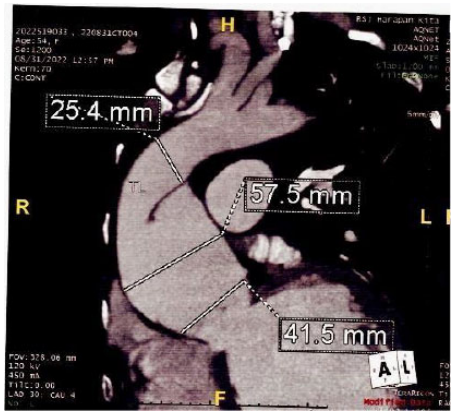

Figure 2. CT Angiography showed revealed aortic root dilatation (45,1 mm), proximal ascending aorta aneurysm (57,7 mm).

mg of Amlodipine once daily, 40 mg of Furosemide twice daily, 5 mg of Bisoprolol once daily, 25 mg of Spironolactone once daily, 10 mg of Ramipril once daily and 20 mg of Simvastatin once daily for the past year. On examination on admission, the patient was alert, body mass index of 15.6, normotension with blood pressure 123/43mmHg on her right arm and 125/39mmHg on her left arm, Heart rate of 75 beats per minute, Respiratory Rate 32 times per minute, Temperature 37,2°C and pulse oximetry was 84%.

Physical examination revealed elevated jugular vein pressure, early diastolic murmur at the upper right sternal border, pectus carinatum, positive wrist & thumb sign, and skin striae. X-Ray showed widening of the mediastinum, and cardiomegaly with elongation, dilatation, and calcification of the aorta. 12-Leads ECG showed sinus rhythm, heart rate 75 beats per minute, poor r-wave progression, ST Elevation in lead III & aVR, and ST depression in lead I & aVL.

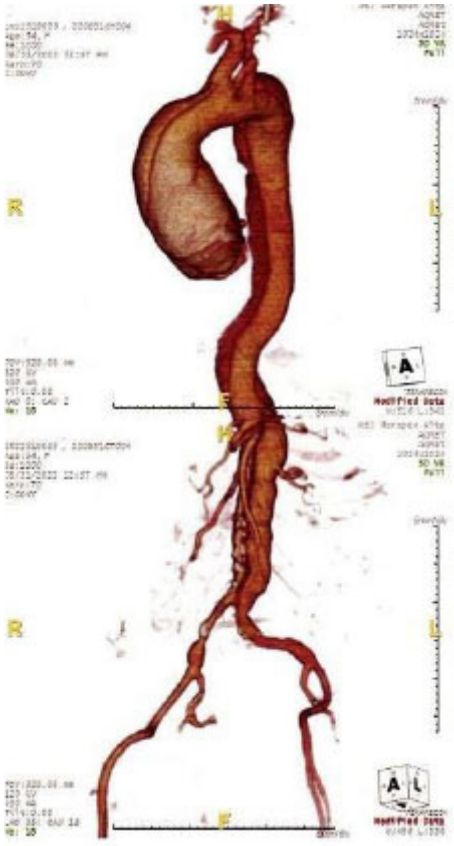

Figure 3. CT Angiography showed aortic dissection as high as the mid ascending aorta, aortic arch, descending aorta, aorta abdominals up to bifurcation aorta with multiple entry tear and re-entry tear before bifurcation aorta.

Laboratory examination revealed mild anemia (10.5 g/dL), elevated liver enzymes (SGOT: 435 U/L & SGPT 305 U/L), and decreased renal function (Ureum: 88 mg/dL & Serum creatinine: 1.5 mg/dL). Her last CT angiography 2 months ago revealed aortic root dilatation (45,1 mm), proximal ascending aorta aneurysm (57,7 mm), aortic dissection as high as the mid ascending aorta, aortic arch, descending aorta, aorta abdominals up to bifurcation aorta with multiple entry tear and re-entry tear before bifurcation aorta.

The systemic score (Z Score) for Marfan's syndrome was 7 (Wrist & thumb sign, pectus carinatum, skin striae, and moderate mitral regurgitation). The patient was then treated with oxygen supplementation, fluid management in heart failure condition, adequate analgesia, gastroprotection, Intravenous diuretic, and continue other routine oral medication.

Then patient undergo echocardiography which revealed Type A Aortic Dissection De Bakey I, Severe Aortic regurgitation, Moderate Mitral Regurgitation, and Tricuspid Regurgitation with a high probability of Pulmonary Hypertension and reduced left ventricular ejection fraction 42%. After three days of admission, all symptoms have improved, and BP is stable at 107/31 mmHg with HR of 60 bpm, then the patient was discharged with 500 mg of paracetamol three times a day, 5mg amlodipine once daily, 2.5 mg bisoprolol once daily, 25 mg spironolactone once daily, 10 mg ramipril once daily, 20 mg simvastatin once daily and 20 mg omeprazole twice daily as her routine oral medication. Surgical intervention is still considered the best treatment option for patients with chronic type A aortic dissection but bad nutritional status and other comorbidities like anemia, low ejection fraction, and decreased renal function make these patients more likely to have bad outcomes if undergo surgical intervention. The patient was then treated conservatively and has been planned for CT evaluation next year at National Cardiovascular Center Harapan Kita.

DISCUSSION

Classic aortic dissection results when the layers of the aortic media are separated by blood, commonly through intimal tears,

thus forming a false lumen. Frequent predisposing factors are atherosclerosis and hypertension. Degeneration of the medial layer is also harmful to the aortic wall, especially in hereditary defects like Marfan's and Ehlers-Danlos' syndromes. A degeneration of the arterial wall is present also in most other cases of AD and is more severe than that expected as part of the aging process.[4]

This could be the result of both genetic factors which enhance continuing irreversible destruction of the aortic wall.[4] According to research, ascending aortic dissection occurs almost twice as often as descending aortic dissection.[5] Chronic AD can be classified into two groups. Patients who had medical, endovascular, or surgical intervention for acute AD and survived for the next 90 days after the initial event gets into the chronic phase, and the other group consists of patients who get diagnosed with chronic AD for the first time.[3]

Complete history taking and clinical examination should be done in the initial evaluation. Sudden severe onset chest or back pain are the classic symptoms of the hallmark of thoracic dissection. It is important to thoroughly question the pain onset, duration of the pain, extent of the pain, pain characteristics, and history of an aneurysm. Neurological deficits like vision change, headache, and stroke can be present in AD patients and compromised mesenteric artery can cause abdominal pain and extremity pain due to femoral artery dissection.[6] The majority of the remaining clinical presentations of subacute/chronic Type A AD were non-specific for aortic dissection, such as shortness of breath, vomiting, coughing, palpitation, calve edema, dizziness, insomnia, fatigue, and hemoptysis.[7]

In a patient with proximal aortic dissection, half of them can be presented with a diastolic murmur that indicates an aortic regurgitation. Periodically, the murmur can be very faint, and peripheral signs of severe aortic regurgitations may be absent. Physicians should call for rapid surgical intervention if pericardial involvement signs like jugular vein distension, pericardial friction rub, or paradoxical pulse exist. If the aorta ruptures into the pleural space, it may cause pleural effusion, the left side is usually involved.[8]

The patient was then diagnosed with chronic type A De Bakey I aortic dissection with thoracic aortic aneurysm and Marfan syndrome with hypertension as the main risk factor and needed to be treated immediately. ECG evidence of acute ischemia or acute myocardial infarction may show in about 20% of type A aortic dissection patients. Patients that have ECG evidence of ischemia with suspected aortic disease must undergo diagnostic imaging before administering the thrombolytic therapy.[8] In this case, 12-Leads ECG showed sinus rhythm, heart rate 75 beats per minute, poor r wave progression, ST Elevation in lead III & aVR, and ST depression in lead I & aVL.

Laboratory examinations such as complete blood count (CBC), serum chemistry, and cardiac marker had to be done. From CBC we may find leukocytosis which usually represents a stress state. We may also find decreases in hematocrit and hemoglobin indicating leaking or rupture of the dissection. Decrease renal function may indicate prerenal azotemia as a result of blood loss or associated dehydration, elevation of creatinine and blood urea nitrogen (BUN) can also indicate involvement of the renal arteries besides exhibiting haematuria, oliguria, or even anuria (<50mL/day). If the dissection involved the coronary arteries and caused myocardial ischemia, the level of myocardial muscle creatine kinase isoenzyme, myoglobin, troponin I, and troponin T will be elevated. If there is hemolysis in the false lumen, lactate dehydrogenase (LLDH) levels may be elevated.[9]

Studies showed that D-dimer levels are highly sensitive to aortic dissection, with 97% negative predictive value, except in young patients and thrombosed false lumen.[10] We didn't have the facility to check D-dimer, LDH, CKMB, Troponin I, and T in this patient. From CBC we found leukocytes within normal limits, mild anemia, elevated liver enzymes, and elevation of BUN and creatinine. Widening superior mediastinum and aorta, the difference in the size between the ascending and descending aorta, change in aortic configuration between successive examinations, and calcified plaque displacement by 10mm or more is the most common finding in chest radiographs in patients with proven aortic dissection. There is often cardiomegaly and pleural effusion may be present, usually on the left side.[11]

The most commonly used modality for imaging patients with acute and chronic aortic dissection is CT angiography. The wide availability of CT scanners and the capability to acquire high-resolution volumetric data sets that can be represented in another wide range format of 2D and 3D angiographic images. Magnetic Resonance Imaging (MRI) can be accepted as an alternative to CT angiography for assessing patients with chronic aortic dissection. Transthoracic Echocardiography (TTE) is commonly performed in chronic aortic dissection patients with concomitant dilatation of the unrepaired aortic root.[12] Recent chest x-ray showed widened mediastinum, and cardiomegaly with elongation, dilatation, and calcification of the aorta.

The last CT angiography revealed aortic root dilatation (45,1 mm), proximal ascending aorta aneurysm (57,7 mm), aortic dissection as high as the mid-ascending aorta, aortic arch, descending aorta, abdominal aorta up to bifurcation aorta with multiple entry tear and the current TTE found are Severe Aortic regurgitation, Moderate Mitral Regurgitation and Tricuspid Regurgitation with high probability of Pulmonary Hypertension and decreased left ventricular ejection fraction 42%. All these findings indicate an immediate repair surgery, but due to high comorbidity and high mortality because of bad nutrition, surgery can't be done and the patient is treated conservatively with medication. The available evidence on the medical management of chronic aortic dissection is lacking.[3]

Medical treatments are given to make the dissection more stable, prevent it from being rupture, accelerate healing, and minimize the risk of complications. The main goal of medication therapy are maintaining systolic blood pressure (BP) to reduce aortic wall stress and controlling the pain with adequate analgetic, morphine sulfate is recommended to use as pain

control.[13] The blood pressure needs to be controlled to approach systolic <110mmHg and heart rate (HR) < 60 beats per minute.[14] To determine antihypertensive agents for patients with aortic dissection, we should consider the contraindications. Monotherapy antihypertensive agents are rarely adequate, a combination of several antihypertensive agents is required to control massive hypertension.[15]

Beta-blockers showed their potentially beneficial role in treating chronic aortic dissection by effectively lowering the heart rate and blood pressure and also reducing the peak ejection rate of the left ventricle. Beta-blockers decreased left ventricle dP/dt and lowered shear stress on the aortic wall.[3] Some studies showed the effectiveness of ACE Inhibitor (ACE-I) or Angiotensin Receptor Blocker (ARB) in decreasing aortic dilatation progress and reducing complications in patients with Marfan syndrome.[16] Statins are suggested to minimize complications for patients with aortic aneurysms in some studies. The availability of data regarding statin's role in chronic aortic dissection is still scarce.[3]

The patient was then treated with pethidine to relieve pain, a combination of Beta-blocker, ACE-I, and diuretics to control blood pressure and heart rate, and statin. After three days of admission, all the symptoms improved, and blood pressure and heart rate had been controlled. The patient then discharges with analgetic, Beta-blocker, ACE-I, diuretics, and statin as routine oral medication. The prognosis of all heritable thoracic aortic diseases is mainly determined by the progressiveness of aorta dilatation, which can lead to aortic dissection/rupture. The mean age mortality of untreated Marfan syndrome patients is <40 years, but can approach the general population if the diagnosis is on time and treated properly.[14]

CONCLUSION

Decision-making regarding treatment options for patients with chronic aortic dissection should consider the outcomes, mortality, and physical function. Chronic type A aortic dissection has high mortality if left unrepaired, but in some cases, surgery or endovascular therapy can't be done. Physicians need to inform the patient and family about the condition, treatment options, long-term medical management, and the prognosis of the disease.

CONFLICT OF INTEREST

None.

FUNDING

This study did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

ETHICAL STATEMENT

The patient has given informed consent regarding the publication of this case.

AUTHOR CONTRIBUTION

All authors contributed equally to this study.

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