Atypical Clinical Presentation of Extensive Aortic Dissection at Mulago Hospital: Case Report

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Introduction

Aortic diseases contribute to the high overall cardiovascular mortality [1]. Aortic dissection (AD) shows a worldwide incidence of 5 to 30 cases per million people per year. The incidence of AD depends on the prevalence of risk factors in the study population. Aortic dissection is a rare cardio-vascular disease with fatal outcome reason why most of the patients with aortic dissection die before their admission in the hospital or prior to diagnosis [2]. The overall in hospital mortality rate for acute aortic dissection is reported to be 12%-27% [3].

The occurrence of aortic dissection is usually preceded by diseases that have weakened the aortic wall. Medial degeneration tends to be more extensive in older adults with chronic hypertension, in cystic medial necrosis associated connective tissue disorders such as Marfan’s syndrome and with atherosclerosis causing occlusion and injury of the vasa vasorum [4].

Aortic dissection typically presents with sudden onset of exsanguinating, tearing, anterior, or interscarpular chest pain that tends to migrate along the course of the dissection. Clinical presentation is dominated by dissection-related side branch obstruction or malperfusion syndrome due to aortic obstruction by the flap.

The Stanford classification of aortic dissection distinguishes between types A and B. Type A means the dissection involves the ascending aorta while type B dissection does not involve the ascending aorta. The De Bakey classification subdivides the dissection process further: A type I dissection involves the entire aorta; a type II dissection involves the ascending aorta, and a type III dissection the descending aorta [5].

Imaging studies confirm the presence of the disease in patients clinically suspected with thoracic aortic dissection. Imaging findings are helpful in classification of aortic dissection, identifying the entry and reentry sites and evaluating the patency of the false lumen. The presence or absence of aortic branch involvement, the involvement of the coronary ostia; the aortic valve competency and the presence or absence of extravasated blood in mediastinal, pleural or pericardial spaces are also detected by imaging studies. In addition, imaging should help distinguish classic aortic dissection from other causes of “acute aortic syndrome” such as acute intramural hematoma and penetrating aortic ulcer [3].

Acute aortic dissection can be treated surgically or medically [6].

Case Report

This was a fifty four year old male who was admitted at Mulago Hospital in Uganda with sudden onset chest pains, became unconscious for few minutes then recovered the consciousness with ability to walk, stand and sit. Clinical diagnosis prior to radiological investigation was acute coronary syndrome. Surprisingly cardiac Echo showed AD with aortic regurgitation. Computed tomography aortogram showed aortic dissection De Bakey type I. The immediate medical management included intravenous morphine and an oral beta blocker, with a target of maintaining the systolic blood pressure <100 mmHg and the resting heart rate between 60-70 bpm. The patient was discharged from the hospital after eleven days on medical therapy including oral bisoprolol, oral losartan and furosemide. Arrangements for Bentall procedure and Thoracic Endovascular Aortic Repair (TEVAR) were planned in two months at the time of discharge from hospital; unfortunately the patient has never been operated. He has been maintained on medical therapy for three years. The aim of this clinical report is to appreciate the role of radiological investigations in the diagnosis of atypical clinical presentation of extensive aortic dissection.
and surgical history was unremarkable. On physical examination, the patient was fully conscious, afebrile with no palor or edema. The chest had bilateral coarse crepitations posteriorly. The cardiac auscultation showed pan systolic murmur and tachycardia with a heart rate of 110 per minute. The blood pressure was 115/98 mmHg. The saturation of oxygen (SaO₂) was 95% on room air. The abdomen was mildly distended, no tender with no pulsating mass noted. No ascites or hepatomegaly was noted. The diagnosis hypothesis was acute coronary syndrome. The laboratory tests requested showed hypercholesterolemia and leucocytosis. Cardiac Echo done at Mulago Hospital showed aortic dissection with aortic regurgitation (Figures 1 and 2).

Computed tomography aortogram done at Saint Francis Hospital Nsambya showed extensive aortic dissection involving the aortic root, ascending aorta, aortic arch, descending aorta and bilateral common iliac arteries (Figures 3-7). On admission, vital parameters were: BP of 115/58 mmHg and HR of 110 bpm. The immediate medical management included intravenous morphine to reduce the pain, and an oral beta blocker, with a target of maintaining the systolic BP <100 mmHg and the resting HR between 60-70 bpm. The patient was also given azithromycin for treatment of possible pulmonary infection and bisacodyl for constipation. Surprisingly the patient was able to walk and sit while awaiting aortic dissection surgical repair.

The patient was discharged from the hospital after eleven days on medical therapy including oral bisoprolol, oral losartan and furosemide. The BP on discharge was 105/62 mmHg and the HR was 65 bpm. He was free of chest and abdominal pain. Arrangements for Bentall Procedure with Aortic Arch replacement and an elephant trunk about 2 cm to be left floating into the descending aorta were planned at the time of discharge from hospital. There was also plan for Thoracic Endovascular Aortic Repair (TEVAR) of the descending aorta, to be done 2 months after arch replacement surgery. The patient has never
been operated. He has been maintained on medical therapy for three years.

**Figure 5:** CT angiogram shows intimal flap in the thoracic portion of descending aorta, suprarenal and infrarenal portions of the descending aorta and common iliac arteries.

**Figure 6:** CT aortogram 3D reconstruction image shows intimal flap in the aortic root, ascending aorta, aortic arch and proximal portion of the descending aorta.

**Figure 7:** CT aortogram shows the intimal flap involving the aortic root, ascending aorta, aortic arch and proximal portion of the descending aorta.

**Discussion**

The above presented patient had sudden onset chest pain leading to provisional diagnosis of acute coronary syndrome. Abdominal pain shows extension of the disease in abdominal aorta and branches. There is high mortality rate noted in patients with extensive aortic dissection. Pain is the most common presenting symptom of aortic dissection. Pain in aortic dissection is midline, experienced in the front and back of the trunk, depending on the location of the dissection. Sudden onset of pain is usually life threatening, and it reaches a maximum level in a shortest time. Patients with dissections of the ascending aorta and arch more frequently experience anterior chest pain, whereas patients with dissections of the descending aorta more frequently experience posterior chest, back, and abdominal pain. These clinical symptoms explain the confinement to bed of patients with extensive aortic dissection contrary to atypical clinical presentation in this case where the patient didn't present any abdominal or back pain and was still able to stand, to walk and to sit down. Extension of the pain down to the back, abdomen, hips, and legs indicates the extension of the dissection process distally. Clinical presentation helps the clinician to make a provisional diagnosis of aortic dissection and to plan the investigations. Our patient had clinical signs of acute coronary syndrome reason why the clinician requested initially cardiac echo than CT scan Aortogram even if echocardiography is still widely available and helpful in the diagnosis of aortic dissection. Cardiac Echo is useful in the diagnosis of dissection involving the ascending aorta and can diagnose the hemodynamic significance of pericardial effusions, the degree of aortic regurgitation, and left ventricular function. In the present case, Echocardiography showed aortic root dilatation with a false lumen extending from the aortic root to the descending aorta (Figures 1 and 2). Aortic regurgitation was also noted. Echocardiography played a good role in the diagnosis of aortic dissection however it couldn't demonstrate the extension of the intimal flap in the entire aorta, in the branches of the aorta and associated lesions of the surrounding tissues. The sensitivity of Echocardiography in detecting descending aortic dissections was previously reported to be lower (31%-80%) than contrast-enhanced CT and MRI, recent technical innovations such as harmonic imaging and microbubble contrast enhancement have been demonstrated to improve the sensitivity of Trans Thoracic Echocardiography (TTE) in detecting descending dissections to 84% [3].
CT angiogram was then indicated to give more details. CT angiography confirmed the diagnosis and showed extension of the lesion in the entire aorta, left subclavian artery, and common iliac arteries, external and internal iliac arteries (Figures 3-7). This was extensive aortic dissection De Bakey type I. Patients with such findings on CT angiogram have a high mortality rate; the diagnosis is usually made after sudden death due to aortic rupture. However, the above presented patient was still able to stand, to walk and to seat, this was atypical manifestation of extensive aortic dissection. CT angiography was helpful for treatment plan. Follow-up of such patients is mandatory. These imaging findings show how helpful is CT scan in the diagnosis of AD.

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Conflict of Interest

The authors declare that they have no competing interests. The article contains case description and figures needed.

Ethics Approval and Consent to Participate

Approval to do this case report was sought by Radiology department of Mulago Hospital. The purpose of this case report was explained to the patient and informed consent was sought.

References


Authors’ Contributions

All listed authors have contributed all the time in writing this case report.

All authors read and approved submission of this case report.

Conclusion

Aortic dissection is still one of the life-threatening cardiovascular diseases with high mortality rate. Patients with extensive aortic dissection may present with sudden onset chest pain and abdominal pain. However, atypical presentation may also occur in few patients with aortic dissection. Echocardiography is helpful for the diagnosis of proximal aortic dissection and evaluation of aortic valve competency and determining the presence or absence of extravasated blood in mediastinal, pleural or pericardial spaces. Echocardiography may not demonstrate clearly the extension of the disease in branches of abdominal aorta. CT aortogram remains the gold standard for diagnosis of AD and treatment plan. Patients with acute coronary syndrome should also benefit of cardiac Echo and CT aortogram for prompt radiological diagnosis leading to better treatment plan in order to reduce the mortality rate of AD.