Case Report

Fatal Rupture of Descending Thoracic Aortic Aneurysm in a Nigerian Patient

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Abstract

Aortic aneurysms are rare conditions in black patients and carry a high mortality if detected late. Unfortunately, the rarity of the condition makes detection a difficulty for many physicians as diagnosis in the early stages requires a high index of suspicion. The varied symptoms presented by the patient often mimic other less severe, but more common conditions and may cause a misdiagnosis. We report a 79-year-old male known hypertensive patient who presented a day before demise with a 4 h history of severe epigastric pain. He was managed for acute exacerbation of peptic ulcer disease to rule out pancreatitis. However, all investigations done were within normal limits. His clinical condition deteriorated, and he died 18 h after admission. An autopsy revealed massive left hemothorax and a tear in the descending aortic aneurysm. Microscopic examination confirmed complicated atherosclerosis in the descending thoracic aorta. The misdiagnosis, in this case, leads to wrong management with the patient receiving treatment for severe peptic ulcer. There is a need to consider this disease in patients who present with severe nonspecific abdominal symptoms.

Keywords: Aortic aneurysm, autopsy, left hemothorax, rupture

INTRODUCTION

An aortic aneurysm refers to a pathological dilation of the normal lumen.^[1] It is defined as a permanent localized dilation of the aorta, having a diameter at least 1.5 times of the expected normal diameter of that given aortic segment.^[2] It is a vascular emergency. Risk factors include systemic hypertension, coronary artery disease, hypercholesterolemia, Ehlers–Danlos Type IV, and Marfan's syndrome.^[3] The most common cause of aortic aneurysms is atherosclerosis in 80% of cases. They are usually asymptomatic except when dissection or rupture occurs, but occasionally present with symptoms such as abdominal and back pain.^[4-6]

Thoracic aortic aneurysms (TAAs) and ruptures are less common than abdominal aortic aneurysms accounting for 25% of all aneurysms.^[7-9] TAAs are more common in males with a male to female ratio of 1.7–3:1 and a mean age of 65 years at presentation.^[7-9] The incidence of TAAs is estimated to be 5.9 compared with 350 cases for abdominal aortic aneurysms per 100,000 person-years.^[10] Of the TAAs, the ascending aorta is affected in 50% of cases; the aortic arch in 10% and the descending thoracic aorta (DTA) in 40%.^[9] Rupture of an aortic

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aneurysm has a high mortality even with prompt treatment only 10%–25% of patients survive. Globally, mortality figures from aortic aneurysms have risen from approximately 100,000 in 1990–152,000 in 2013.^[11]

CASE REPORT

We report a 79-year-old male patient who was a known hypertensive for several years but has not been compliant with prescribed medications. He was also on medications for peptic ulcer diseases diagnosed years earlier. He presented a day before demise with a 4 h history of severe epigastric pain. He was managed for acute exacerbation of peptic ulcer disease to rule out pancreatitis and was subsequently placed on intravenous fluids, antibiotics, and antiulcer medications. Results of hematological and serological investigations

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were within normal limits. Eighteen hours later, his clinical condition deteriorated rapidly and he had a cardiopulmonary arrest and was confirmed dead.

At autopsy, general examination revealed only severe pallor. The thoracic cavity, however, revealed 3,300 ml of left hemothorax with collapse of the left lung and bulging left pleura [Figures 1 and 2]. There was a 6-cm aneurysmal dilatation of the DTA with a 4 cm \times 2 cm tear/rupture [Figure 3] in the wall of the aneurysmal sac. Atheromatous plaques and extensive intimal hemorrhage were also seen.

Microscopic examination confirmed complicated atherosclerosis in the DTA. Hypovolemic shock from massive hemothorax due to rupture of a descending TAA was implicated as the cause of death.

DISCUSSION

A high index of suspicion is needed in diagnosing aortic aneurysm due to its ability to mimic other more common conditions as was seen in this case.^[12,13] The disease has been dubbed "a silent killer" due to the fact that only about 5% present with symptoms while 95% of emergency patient with aortic aneurysm were previously asymptomatic. The previous history of peptic ulcer disease had totally masked any suspicion of an aortic aneurysm. The lack of evidence of peptic ulcer in the stomach at autopsy suggests that the epigastric pain may be due to the weakening aneurysmal sac.^[14]

Cystic medial degeneration, usually due to aging, has been implicated as the main cause of aortic dissection and aneurysm in the ascending part of the thoracic aorta while atherosclerosis is the most common cause in the descending part of the thoracic aorta.^[15] This is due to the weakening of the tunica media by deposition of atherosclerotic plaques causing derangement of layers of elastic fibers and smooth muscle cells as was seen in the index patient with complicated atherosclerosis. Although many connective tissue diseases, such as Marfan and Ehlers–Danlos syndrome, are known causes of aortic aneurysm, familial clustering of diseases have also been reported with several genes which code for fibrillin being implicated particularly MYH11 and ACTA2.^[16]

The silent nature of the disease stresses the importance of doing appropriate examinations and tests in the elderly and in patients with family history of the disease. Despite the high fatality rate in the event of a rupture, the disease is curable using surgical and medical therapy if discovered early.^[16] Typically, most aneurysms are treated surgically when the diameter of aneurysmal sac exceeds 5 cm.^[15,16] Medical treatment is usually instituted before the aneurysm getting to the critical value. The aneurysmal sac in our index patient was 6 cm which indicates that the aneurysm had gone beyond the critical value and was already a candidate for a catastrophic event as at the time of presentation.



Figure 1: Massive left hemothorax on opening the body up on the autopsy table



Figure 2: Blood clots removed from the left thoracic cavity at autopsy



Figure 3: Ruptured aneurysmal dilatation of the descending thoracic aorta

Over 50% of aortic aneurysms are undetected before death which suggests that the actual incidence of this disease is not truly known.^[17] The autopsy that was done in the index patient was key to arriving at the definitive cause of death. The importance of autopsy is further illustrated by this case and will again remind the medical community about the need to be aware of a "silent killer" and take appropriate preventive measures in their patients.

CONCLUSION

The findings at the autopsy of this index patient is a reminder of the postmortem examination in determining the cause of death and thus making us have a true incidence of many "Silent killers" such as aortic aneurysms which are underreported. There is a need to look out for this disease in susceptible patient populations.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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