

Rupture of an ascending aortic aneurysm as a cause of sudden death

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ABSTRACT

An 84-year-old female patient was brought to the emergency department in cardiac arrest. Cardiopulmonary resuscitation maneuvers were performed but were unsuccessful. The patient had a past medical history of systemic arterial hypertension with target-organ lesions, including stroke and myocardial infarction. The autopsy was carried out, and the most striking finding was cardiac tamponade due to the rupture of an ascending aortic aneurysm at the site of a complex atheromatous plaque. Rupture is the most serious complication of a thoracic aneurysm and must be considered in the differential diagnosis of sudden death.

Keywords

Aortic Aneurysm; Death, Sudden; Atherosclerosis

CASE REPORT

An 84-year-old female patient was brought to the emergency department (ED) by the Pre Hospital Service in cardiac arrest (asystole). She had collapsed in the presence of relatives 30 minutes before, and the resuscitation maneuvers began 10 minutes before the hospital admission. In the ED, the cardiopulmonary resuscitation protocol for asystole lasted for a further 25 minutes, but was not successful. The relatives reported a previous medical history of systemic arterial hypertension (SAH), dilated cardiomyopathy, myocardial infarction, peripheral arterial disease, stroke, and transient ischemic attack. The autopsy was carried out with the agreement of the family.

AUTOPSY FINDINGS

The corpse weighed 78.0 kg and measured 1.69 m. The external examination was unremarkable with no signs of trauma. At the opening of the pleural and peritoneal cavities, no effusions were present. The mediastinal fat had hemorrhagic spots. The pericardial sac was bulging and tense, and after opening, a clot was found inside measuring approximately 750.0 mL in volume (Figure 1A). The heart weighed 430.0 g (mean reference value [mRV]: = 260 g) and had a thick layer of epicardial fat. The ventricular free walls were thickened, measuring 0.8 cm on the right (RV = 0.5 cm) and 2.0 cm on the left (RV = 1.5 cm), with a regular, brownish, and firm

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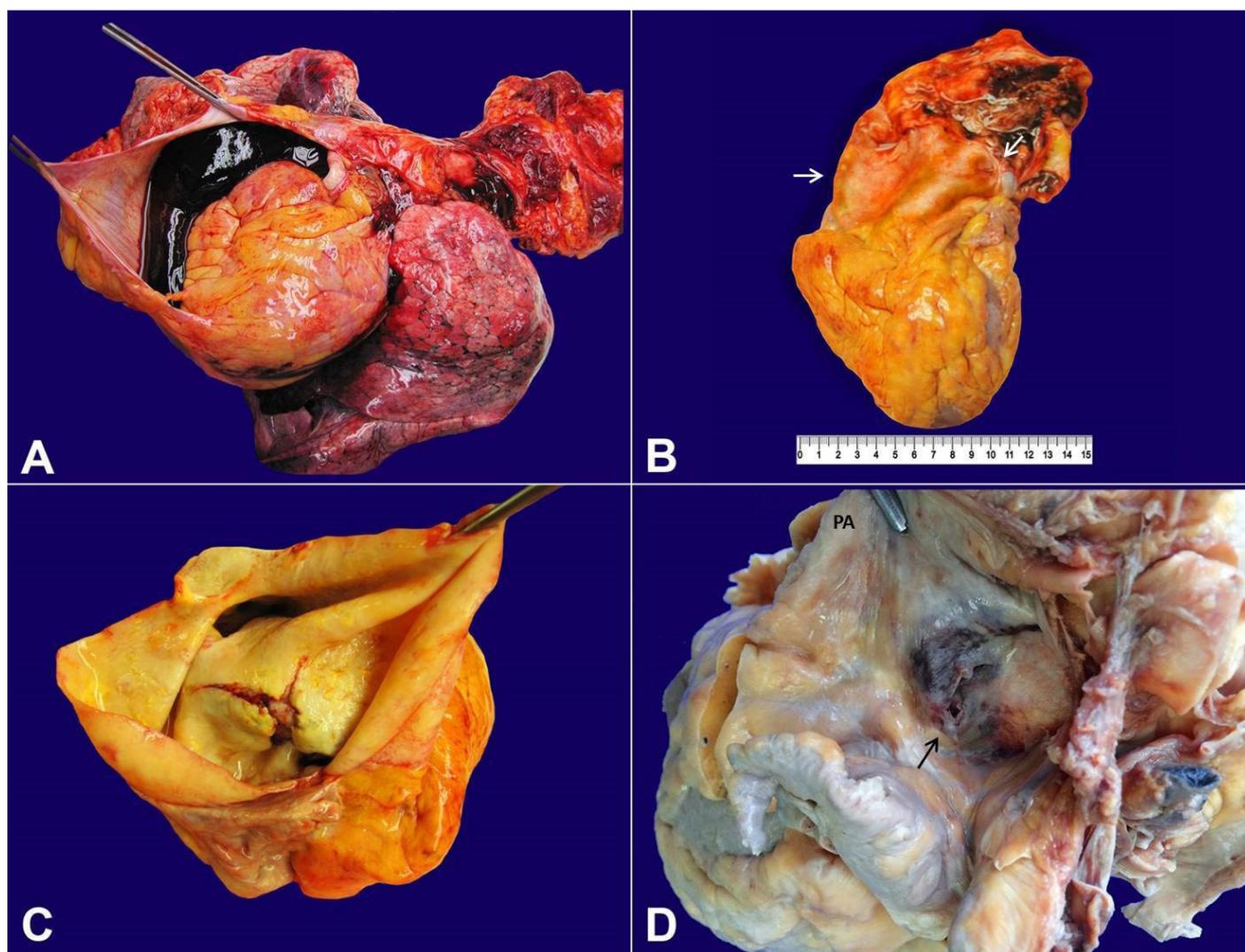


Figure 1. Gross view of the heart and ascending aorta. **A** - A clot within pericardial sac, which was responsible for cardiac tamponade; **B** - External view of the anterior aspect of the entire heart showing the aneurysm of the ascending aorta, which measured 8.5 cm in diameter (white arrows); **C** - Internal view of the aorta, showing a fractured atheromatous plaque; **D** - The heart after formalin fixation. The left lateral view of the heart base and intrapericardial segments of the pulmonary trunk (PA) and aorta, showing the orifice of rupture with a surrounding hematoma (arrow).

myocardial cut surface. The histologic exam showed cardiomyocyte hypertrophy and cytoplasmic lipofuscin pigment. The atrioventricular valves had myxoid degeneration. The main coronary artery branches had atheromatous plaques, which obstructed at least 25% of the lumen.

An ascending aortic aneurysm was detected, measuring about 8.5 cm in diameter (Figure 1B). At the opening of the arterial lumen, the endothelial surface was caked with atherosclerotic plaque. An area of the plaque was ruptured in the intra-pericardial portion of the ascending aorta (Figure 1C and 1D). On the left side, there was a dissection of the aortic wall

measuring 6.0 cm in length from the point of rupture. The microscopy showed fractured atherosclerotic plaques covered by thrombi, and bleeding in the media and adventitia producing delamination of the arterial wall with rupture and moderate neutrophilic infiltration (Figure 2A and 2B). The aortic branches had hard walls with ulcerated and calcified atherosclerotic plaques. The myocardial had interstitial fibrosis and cardiomyocyte hypertrophy (Figure 2C and 2D).

The right kidney weighed 152.0 g, and the left kidney weighed 188.0 g (both RV = 120-150 g). The renal capsule was easily detached and the renal surface was finely granular, bilaterally. At microscopy,

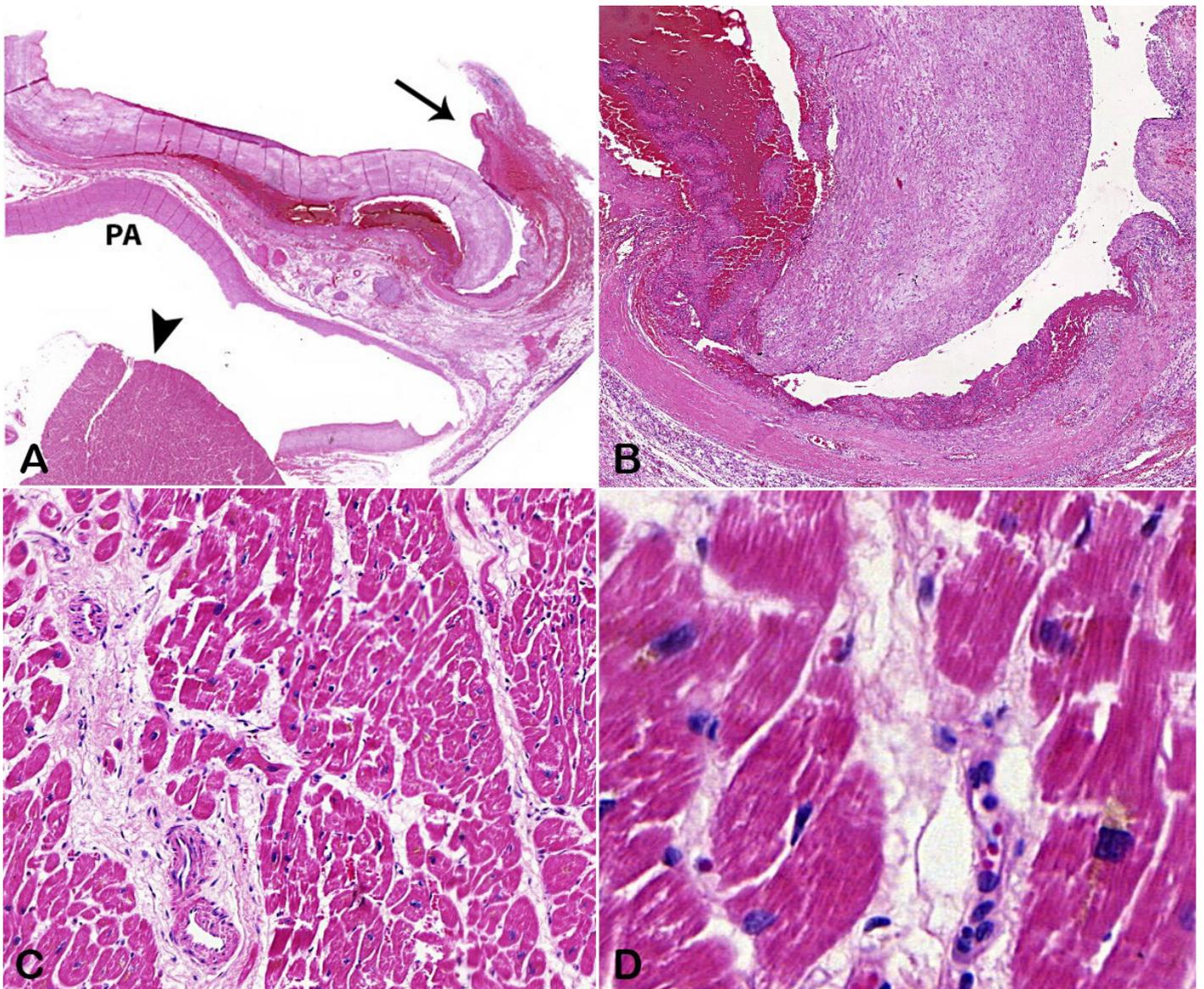


Figure 2. Photomicrographs of the aorta and myocardium. **A** - A low power view of the point of the rupture of the ascending aortic aneurysm: aorta with bleeding and clot (arrow), pulmonary artery (PA), and septal myocardium (arrow head (H&E, 5X); **B** - Detail of the fractured atheromatous plaque in the ascending aorta, with bleeding in the media and adventitia (H&E, 200X); **C** - Perivascular and interstitial myocardial fibrosis (H&E, 100X); **D** - Cardiomyocyte hypertrophy with cytoplasmic deposition of lipofuscin pigment (H&E, 400X).

the findings were: glomerulosclerosis; focuses of interstitial chronic inflammatory infiltrate with fibrosis and atrophic tubules; and acute tubular necrosis and arteriolosclerosis.

In the brain, discrete atherosclerosis of the circle of Willis and a glial scar formation in the white matter of the temporal lobe was observed, and measured 2.0 cm in diameter. Other pathologic findings included lungs with chronic inflammatory peribronchial infiltration (lymphocytes and monocytes) and centrilobular emphysema with mild bronchitis; liver with microgoticular steatosis and intense sinusoidal

congestion ("nutmeg liver"); and chronic cystitis and an intramural uterine leiomyoma.

DISCUSSION

The autopsy case reported herein represents a death as a consequence of a complication of SAH and generalized atherosclerosis. The patient experienced sudden death due to the rupture of an atherosclerotic plaque in an ascending aortic aneurysm, with dissection, followed by hemopericardium, which produced cardiac tamponade and hypovolemic shock.

The autopsy also revealed lesions in target organs produced by the atherosclerosis and the SAH, such as myocardial fibrosis, nephrosclerosis, and a glial scar from a previous stroke. These findings revealed poor long-term control of SAH.

The aortic dissection observed at the macroscopic exam can be classified as Stanford type A (when the aortic dissection involves the ascending aorta). In this case, the dissection was associated with a fusiform aortic aneurysm. Most of the thoracic aortic aneurysms (TAA) are consequent to degenerative diseases affecting the vascular wall, and atherosclerosis is responsible for almost one-third of all cases of aortic aneurysmal disease. Without treatment, especially regarding the control of SAH, the TAA evolves slowly, with progressive dilatation. The most common complications of an aortic aneurysm are dissection and rupture, which are important causes of death in the USA.¹⁻⁴ It is important to explain that aneurysms can be complicated by wall rupture without dissection, and arterial wall dissection can occur without aneurysm.¹⁻⁴

Aortic dissection has an incidence of 2.9 per 100,000 inhabitants/year and predominantly affects men between 60 and 80 years of age. Its main risk factors are high blood pressure (hypertension), dyslipidemia, smoking, Marfan syndrome, Ehlers-Danlos syndrome, Turner syndrome, other connective tissue diseases, previous aortic valve disease, cerebral aneurysm, and family history.^{5,6} Tertiary syphilis, with cardiovascular manifestation, classically dilates the ascending aorta and causes aortic valve regurgitation due to plasmacytic vasculitis involving the vasa vasorum in the adventitia. In atherosclerotic disease, the inflammation of the arterial wall is seen in the media and surrounds the atheromatous plaque.^{7,8}

The clinical picture of aortic dissection includes chest pain radiating to the back, which, occasionally, can be associated with pulse asymmetry, syncope, stroke, myocardial infarction, aortic insufficiency, and acute heart failure. These signs and symptoms are more common in cases with a Stanford type A dissection.^{1,2,9} Rupture of an aneurysm is considered the most serious complication, and can occur because of a weakness of the wall or an atherosclerotic penetrating ulcer, which produces an intramural hematoma, dissection, and subsequent rupture. The rupture of an ascending aorta aneurysm occurs inside the pericardial sac or in the thoracic cavity (most commonly in the left hemithorax),

producing intense chest pain, hypotension, shock, and sudden death.^{1,2} Cardiac tamponade is observed when the aneurysm rupture goes into the pericardial sac, as observed in this reported case. Occasionally, the aneurysm rupture passes into the esophagus through an aorto-esophageal fistula.¹⁰ In a cohort study on acute aortic dissection with 1,079 patients enrolled, Nienaber et al.³ found that women were less likely to have aortic dissection (346 cases [32.1%]). However, the women were older and arrived later at the hospital to get medical assistance than the men, and were more likely to have neurologic signs (coma), aneurysm rupture (periaortic, pericardial, or pleural), shock, cardiac tamponade, higher in-hospital mortality, and a worse surgical outcome. In a similar cohort study, Gilon et al.¹¹ found 126 cases of cardiac tamponade (18.7%) among 674 patients with Stanford type A aortic dissection, with a 54% mortality rate. In a retrospective study evaluating ruptured TAA in Stockholm, Sweden, Johansson et al.¹² found a total of 158 cases in 1980 and 1989, with 41% of the cases arriving alive in the ED, but with an overall mortality rate near to 100%. In the city of Malmö, Sweden, Svensjö et al.¹³ found 63 cases of death due to the rupture of the thoracic aorta in the autopsy records from 1958 to 1985, with an incidence of 0.9/100,000 for men and 1.0 per 100,000 for women.

The size of the thoracic aneurysm can predict the risk of rupture. Davies et al.¹⁴ analyzed data from 721 patients with TAA; those patients with aneurysms greater than 6.0 cm in diameter had a 27-fold risk increase for rupture in a 5-year follow-up, with rupture and death occurring at a rate of 3.7% and 11.8% per year, respectively. In the case reported here, the patient had an aneurysm measuring 8.5 cm in diameter.

For medicolegal reasons, or to evaluate the quality of medical assistance, a pathologist must give an estimate of the extent, localization, and time of the rupture, which are revealed during the autopsy of a deceased person from an aortic aneurysm rupture. This issue was competently addressed by Ma and Ang¹⁵ in the case report of an 82-year-old woman who died from spontaneous rupture of the descending thoracic aorta distal to the left subclavian artery ostium through an atheromatous plaque, without exhibiting a local aneurysm. Deaths immediately after such a rupture show an acute inflammatory response with neutrophils in the vessel wall. Signs of healing indicate that the

rupture is old: tissue granulation and fibrosis appear after a week; and chronic pericarditis, fistula formation, and false aneurysm appear after 2-3 weeks.¹⁵⁻¹⁸ In the case reported herein, we could find hemorrhage and a discrete leukocyte infiltration in the adventitia by the area of rupture, indicating immediate death after the event. In many cases, the diagnosis of an aneurysm and its rupture is only made at autopsy. Vázquez Muñiz and Delgado Osorio¹⁹ evaluated 16 patients with acute dissection of the thoracic aorta. The type A dissection was more common (88%) and the overall mortality was 93%, with eight cases (50%) diagnosed at autopsy. Nine cases had cardiac tamponade (56%). Young and Ostertag²⁰ found 114 cases of aortic aneurysms, 95.6% due to atherosclerosis and SAH, among 3,375 autopsied cases during 5 years in Germany. The percentage of thoracic aneurysms, thoracic aneurysms with rupture, and rupture as the immediate cause of death were 22.8%, 41.2%, and 65.4%, respectively. SAH was a high risk factor for aneurysm rupture in this study.²⁰

The present case represents how a thoracic aneurysm can be the cause of sudden death and must be listed in the differential diagnosis made by all doctors working in an ED. The diagnosis in the emergency room can be performed with imaging auxiliary methods, such as a bedside ultrasound or computed tomography, and the thoracic and cardiac surgery team must be immediately activated.²¹ However, the mortality rate is very high, even when the patients arrive alive at the hospital.^{11,12,14,19} Preventive health measures should be implemented for people with aortic aneurysms, such as blood pressure control, smoking cessation, and close observation of the aneurysm using imaging exams with surgical intervention, when properly indicated, to avoid lethal outcomes.^{1,2}

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