Case Report

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Two Cases of an Unusual Childhood Aortic Dissection Resulting in Death

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Abstract

Pediatric aortic dissection is an emergency condition that is difficult to diagnose and is associated with high morbidity and mortality. To draw attention to this issue, we present two male cases diagnosed with aortic dissection because of an autopsy. A 16-year-old man with severe new-onset chest pain had an unremarkable physical examination, electrocardiography, and laboratory values. Aortic dissection was not detected on non-contrast computed tomography or echocardiography. Type A aortic dissection was discovered during the patient's autopsy. In the second case, a 10-year-old male patient complained of fever, vomiting, and abdominal pain. The physical examination and biochemical tests were unremarkable. Type B aortic dissection was discovered during the autopsy. Genetic and congenital causes predisposing to aortic dissection and hypertension or a history of trauma were not detected in either patient. Aortic dissection, which is very rare in children, should be considered in cases of persistent chest pain, and rapid diagnosis with contrast-enhanced tomography is vital.

Keywords: Aortic dissection, autopsy, chest pain, child

Introduction

Aortic dissection (AD) is an emergency procedure with high morbidity and mortality. AD occurs as a result of an aortic intimamedia tear, which allows blood flow between the layers. The classical symptoms of AD are the sudden onset of severe chest and back pain. The mortality rate for untreated Type A AD is estimated to be 1% per hour at the beginning, 50% by the third day, and 80% by the end of the second week. Mortality rates are lower in acute type B AD (1).

According to current data, 0.67-3.5% of ADs occur in people under the age of 21. The weakness of the aortic media layer creates predisposition (2). The most common predisposing cause in children is congenital cardiovascular disease. Less frequently, it may also occur due to trauma (3).

We present two male patients, aged 16 and 10 years, who were diagnosed with AD as a result of an autopsy performed in our institution to draw attention to the fact that AD can be rarely seen at a young age and that there may be no findings in echocardiography or non-contrast computed tomography of the thorax and unusual complaints.

Case Presentation

This case presentation was approved by the Council of Forensic Medicine Education and Scientific Research Commission on May 10, 2022, in a decision with the number 407/2022 and the date May 10, 2022.

The first case, a 16-year-old male patient, presented to the emergency department with severe new-onset chest pain. His general condition was moderately good; his body weight was 83 kg (90 percent), his height was 178 centimeters (cm) (72%) and his breathing was comfortable. Vital signs were normal (arterial blood pressure: 134/60 mmHg, heart rate: 99/min, fever: 36 °C, oxygen saturation measured by pulse oximetry: 98%). Examination revealed a systolic, innocent murmur. The electrocardiogram was normal. Laboratory results showed no abnormalities. Serial troponin measurements repeated every 3 hours were normal. No pathology was detected on non-contrast

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thorax computed tomography. He was given symptomatic treatment and discharged with a preliminary diagnosis of myalgia.

The patient, whose chest pain worsened again, was admitted to the emergency unit of a different institution a few hours later. The physical examination was normal except for a 1-2/6 systolic murmur. His heart teleradiogram was normal. Creatine kinase (CK): 680 U/L, CK-MB: 16.7 U/L; C-reactive protein (CRP), 2.45 mg/L; Troponin I: <0.100 ng/mL; white blood cell, 13.130/mm^3; hemoglobin, 15.8 g/dL; platelet count, 212000/mm^3; and the Coronavirus disease-2019 polymerase chain reaction test were negative. The patient was consulted with cardiology. No ischemic electrocardiography (ECG) changes were detected. In addition, no pathological findings were detected in the transthoracic echocardiography except for mild tricuspid regurgitation. While pediatric cardiology control was planned, sudden cardiac arrest developed, and there was no response to cardiopulmonary resuscitation (CPR).

An autopsy was performed. 3500 mL of partially coagulated blood was seen within the right chest cavity. Minimal blood in the pericardial cavity and a 3-cm tear with diffuse hemorrhage around the posterior wall on the upper right of the pericardium were detected. Hemorrhage was observed around the descending and thoracic aortas in the posterior mediastinum. There is a tear in the aortic intima media extending transversely 0.5 cm above the valve (Figure 1a, 1b). Separation in the vessel wall layers and an adventitial defect behind this area, and separation along the thoracic aorta due to AD, pseudolumen formation, and hematoma were observed (Figure 2a, 2b). Histological examination revealed intimomedial separation, fibrin thrombus formation in this area, bleeding, mixed inflammatory cell infiltration in adventitial fatty tissue, and degeneration of elastic fibers in the vessel wall, consistent with AD.

The second case was a 10-year-old male patient weighing 70 kg (99 percentile) who was given ibuprofen by his family after he complained of fever and weakness at 07:00. He was brought to the hospital at 13:00 after he vomited 2-3 times and had abdominal pain. Except for a fever (38 °C), arterial blood pressure of 120/65 mmHg, no other pathological findings were found during his physical examination. He was discharged with a pre-diagnosis of acute gastroenteritis after the administration of intramuscular methaclopropamide and ranitidine.

He was brought back to the hospital at approximately 20:00 because he vomited again at home and his abdominal pain continued. His general condition was good, and there were no abnormal findings on physical examination. The oxygen saturation was 97%, his pulse was 104 bpm, and the fever 36 °C. Except for CRP 11.0

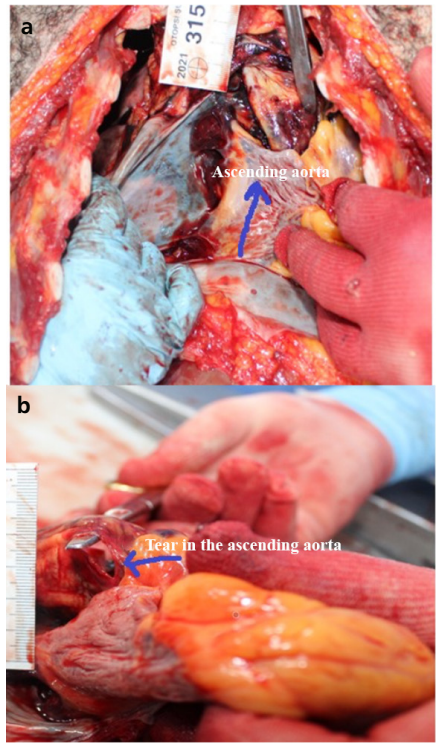

Figure 1. a) Ascending aorta, b) tear in the ascending aorta

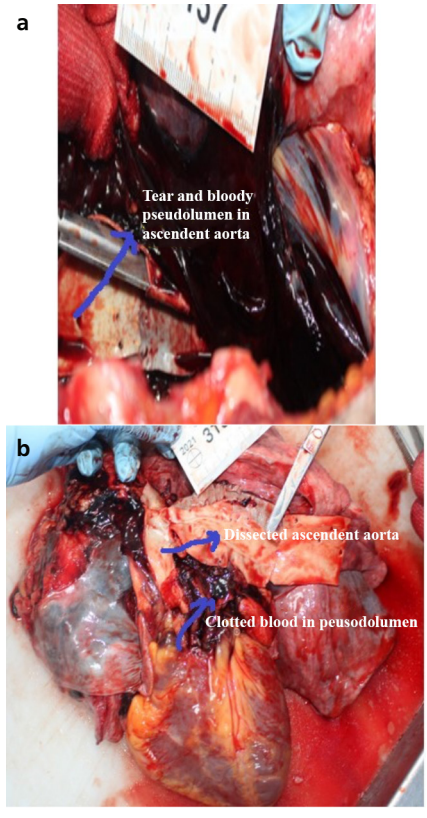

Figure 2. a) Tear and bloody pseudolumen in ascendent aorta, b) dissection and pseudolumen formation in the thoracic aorta

mg/L (ref: <0.5), blood tests were normal. Protein and ketones were positive in the qualitative urine test. At 00:08, a direct abdominal X-ray was taken. Increased gas was seen. General surgery and pediatrics are consulted on the patient. A rectal enema was applied. He was discharged at 3:15. According to his expressions, at 7:30, he said that his stomach hurt a lot and he felt like his heart would stop. He was found lying on the ground, pale yellow, around 7:50. When the emergency aid team arrived, the patient was in cardiopulmonary arrest, and there was no response to CPR.

An autopsy was performed. 100 cc of blood in the right thoracic cavity and 2000 cc of partially coagulated blood in the left thoracic cavity were observed. When the pleura was scraped over the vertebrae, there was a 1.5-cm-long aortic rupture open to the posterior chest wall on the left side. The heart was opened, and rare atheromatous plaques were observed in the aortic lumen and valves. The thoracic aorta was removed from the arch to the level of the diaphragm. There was bleeding around and between the layers. The abdominal aorta appeared intact. When the aorta was opened, it was ruptured at the level of the 7th vertebra of the thoracic spine. There was bleeding between the layers of the aorta downwards and upwards, and there was bleeding to the left side of the chest with a left opening 1.5 cm below the rupture. The aortic diameter was measured at 1.2 cm at the rupture part. In the histopathological examination of the aorta and surrounding connective tissue, medial degeneration in the aorta, laceration forming a cleft in the medial wall, old bleeding areas organizing in this area, hematoma formation in the medial layer, fresh bleeding in the paraaortic soft tissue, an increase in band-type connective tissue, and medial hypertrophy in medium-sized vascular structures were detected. Subendocardial valvular myxoid degeneration was detected in the heart tissue.

Discussion

As in our two cases, 76% of the cases in the literature were male. In addition, risk factors such as trauma, connective tissue diseases, Marfan syndrome, and hypertension were absent in our first case (4). However, in our second case, subendocardial valvular myxoid degeneration and medial degeneration of the aorta were found in postmortem histopathology.

AD is classified according to duration and localization (Stanford Classification). If the elapsed time is less than 14 days, it is acute; if it is more, it is chronic. If the dissection includes both the ascending and descending aorta, it is defined as type A, and if it includes only the ascending aorta, it is defined as type B (1,5). While our two cases were acute dissection, the first case was type A, and our second case was type B dissection. The worldwide incidence of acute AD is approximately 2.6-3.6 cases per 100,000 people per year; however, in China, the rate may reach 5 cases per 100,000 people per year (6). The disease progresses rapidly and is fatal if untreated immediately. Unfortunately, despite recent advances in diagnostic techniques, rapid diagnosis of AD remains challenging, largely because of the heterogeneity of symptoms.

In a series of 1351 ADs for which 9-year data from 12 centers were analyzed, only 9 patients were under 21 years of age. In the projection made, it is predicted that 50 pediatric patients with AD will be seen in the United States between 2005 and 2040, excluding those after trauma (2). In another series, the median age was 19 years (15-21) in 45 patients with AD under the age of 21: 82% were male, 42% were due to trauma, 24% were due to Marfan syndrome, 22% were without predisposition, 8% had aortic valve disease, 6.7% had hypertension, 4.4% had mitral valve disease, 2.2% had Takayasu's arteritis, and 2.2% had fibromuscular dysplasia (7). Strenuous exercise and drug use could cause high blood pressure, a sudden blood pressure increase, and AD (8). In the medical records of both cases, there was no strenuous exercise or drug use. The postmortem weights in both cases were above average. There was also no history of violence or emotional stress. While there was no known predisposing factor in the 16-year-old patient of our two cases, aged 16 and 10 years, medial degeneration of the aorta was detected in the second case.

In cases of traumatic AD, there is a structure that makes dissection easier. Dissections that occur on their own are also common in these patients, and their family members should also be checked for dissection (9). Although Q waves and ST elevation can be seen on the ECG, they may be normal in 25% of patients. Two ECGs taken from our patient were also found to be normal (10). Aortic and mitral regurgitation can be seen on echocardiography, and an increase in aortic diameter can be seen. However, in the first case, echocardiography performed shortly before the patient's death did not reveal any features other than mild tricuspid regurgitation. Again, no pathological features were detected in the non-contrast thorax computed tomography examination performed 8 hours before his death.

Aortography, magnetic resonance imaging, or echocardiography are helpful, but thoracic computed tomography (CT) is the first-choice diagnostic method. Contrast-spiral CT is more sensitive. Transesophageal echocardiography is highly sensitive and is considered a specific imaging modality for detecting intimate flap AD. The gold-standard diagnostic method is angiography (2). In our patient, aortic enlargement was not observed

with non-contrast CT. In our second case, because the symptoms presented as acute gastroenteritis and did not suggest a cardiac cause, AD was not considered in the differential diagnosis, and these tests were not requested.

Although rare, AD can be observed in the pediatric patient group. As in our first case, if there is severe chest pain that cannot be explained by standard diagnostic methods and does not respond to painkillers, it should be referred without delay to an advanced cardiology center, where it should be carefully evaluated in terms of AD and emergency treatment should be performed. Sometimes, AD patients may be masked by accompanying nonspecific findings such as vomiting and abdominal pain, as in our second case, which may cause delays in diagnosis and increase mortality. In severe chest pain that does not respond to painkillers, AD should be considered, and the necessary examinations should be made quickly. Referral to a center where it can be operated may reduce mortality.

Ethics

Informed Consent: Consent was not obtained because the cases were dead and anonymized.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: M.K., A.A., C.E., Concept: K.S., Design: B.O., F.M., M.E., Data Collection or Processing: M.K., A.A., K.S., C.E., Analysis or Interpretation: K.S., B.O., Literature Search: K.S., F.M., M.E., Writing: K.S., B.O.

Conflict of Interest: The authors have no conflicts of interest to declare.

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