Fatal and Near Fatal Acute Ascending Aortic Dissection: Two Case Reports with Different Cardiac Manifestations

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Abstract

Acute ascending AD remains a catastrophic disease that is liable to be missed or have a delayed diagnosis. With such diverse manifestations, the diagnosis in any atypical case is even more difficult and mortality rates are unacceptably high. We reported two cases of acute AD presenting with different manifestations. The first one was a typical Standford type A case, a 71-year-old hypertensive man presenting with chest pain and syncope. With the high risk predictors and delayed operation, he died from cardiac tamponade. In contrast, the second case was an atypical one, a young man who had no known risk factors for developing AD. He initially presented with upper back, leg pain (Standford type B) and later congestive heart failure from severe aortic regurgitation. He survived the complex aortic repair and valve replacement but the cause of AD remained unknown. We hope that these two reported cases will raise awareness of this lethal disease in clinical practice.

Acute ascending aortic dissection (AD) is a rare disease and carries a high fatality rate. Its mortality increased with time, 1-2% every hour after symptom onset. Occasionally, the diagnosis of acute AD could be missed or delayed since its presentation can mimic any arterial occlusive diseases including acute coronary syndrome, limb ischemia and stroke. To raise awareness of the medical community, we report on two acute ascending AD patients who presented with different cardiac manifestations.

Case Report # 1

A 71-year-old man, with a known history of hypertension, developed a persistent mid-chest discomfort for 12 hours. He felt dizzy while sitting up and was brought to Bhumibol Adulyadej hospital. Later, he fainted in the rest room and became hypotensive; his blood pressure (BP) was 68/43 mmHg. Since there was no palpable pulse, cardiopulmonary resuscitation was performed for seven minutes. After 2,000 ml of 0.9% NSS was given, his right and left arm BP was 90/50 and 120/60 mmHg respectively, the pulse rate was 94 per/min in sinus rhythm (SR). The echocardiogram showed moderate pericardial effusion, a hypertrophic left ventricle with normal systolic function and no wall motion abnormalities were noted. The chest film showed cardiomegaly, a dilated ascending aorta and wide mediastinum (Figure 1A). The initial electrocardiogram (ECG) showed sinus rhythm with occasional PAC and mild ST segment depression was noted in V5-6 (Figure 1B). To rule out aortic dissection, an emergent multi-slide computed axial tomogram (MDCT) was performed. It showed an acute ascending aortic dissection from the aortic root to the arch involving the brachiocephalic artery, right common carotids and right sub-clavian arteries (Figure 2). No flap was noted at the distal arch and descending aorta. The left subclavian artery, brachiocephalic artery were not involved. He became hypotensive again and improved after three liters of NSS intravenous infusion, BP was 120/66 mmHg, HR was 90 in SR. The right atrial pressure was 25 mmHg; the pulmonary capillary wedge pressure was 32 mmHg. The patient was just about to be transferred for aortic repair but he had bradycardia, hypotension and expired from cardiac tamponade. Pericardial taping obtained 300 ml of un-clotted blood. The total time from arrival to expiration was seven hours.

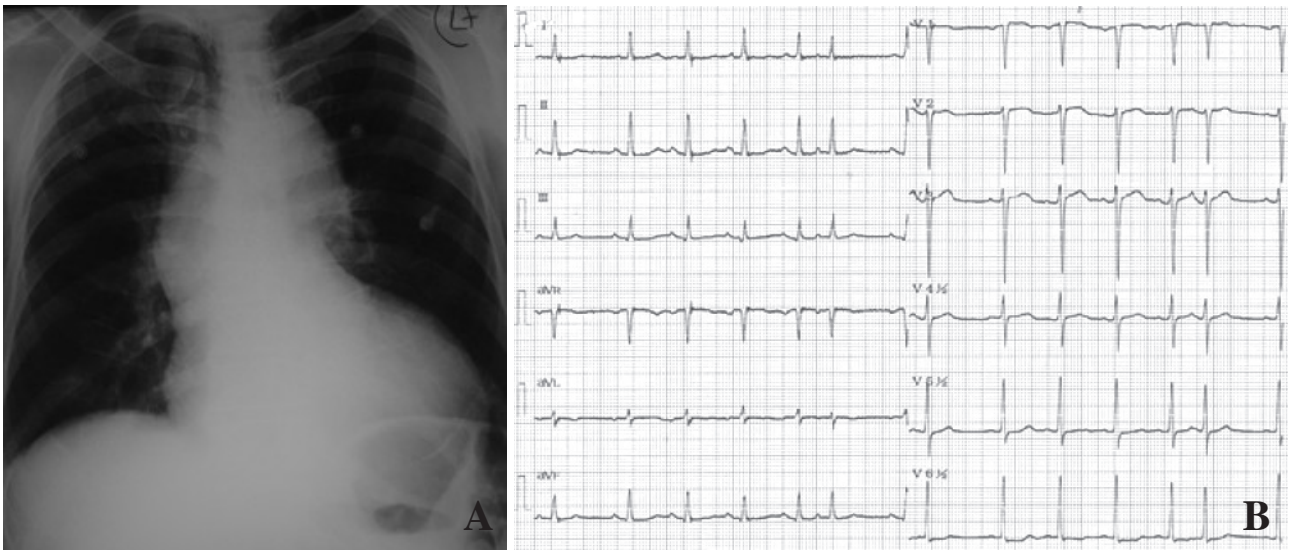

Figure 1A: Chest film showed widening mediastinum with cardiomegaly,
1B: ECG illustrated sinus rhythm with occasional PAC, mild ST depression in V5-6.

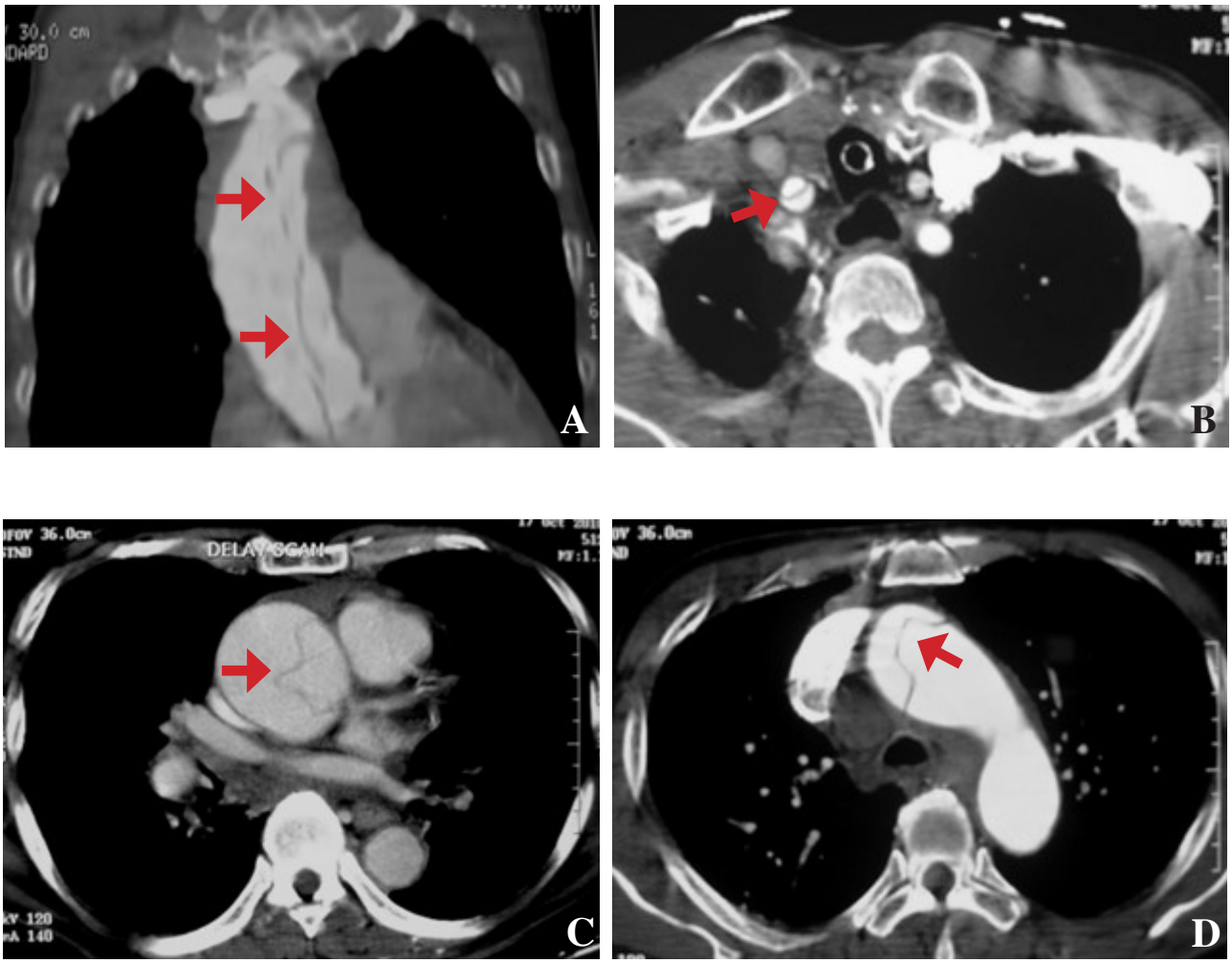

Figure 2: Computerized axial tomogram (CAT) showed an ascending aortic dissection, Standford type A (arrow 2A, C, D) extended to aortic arch involving brachiocephalic artery (arrow 2B), rt. common carotids and right sub-clavian arteries.

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Case Report # 2

A previously healthy 27-year-old man experienced pain in the upper back and shoulder region two weeks prior to admission. He had never had a physical check-up before but his systolic blood pressure was high; 170 mmHg when he was seen. After taking pain medication from the clinic, he felt better within a week but began to experience tightness and numbness of the right leg while walking. Three days prior to admission, he had progressive dyspnea and orthopnea and was brought to Chandrubeksa hospital (located in Nakorn Pathom, 100 km away from Bangkok). It was found that he had congestive heart failure from severe aortic valve leakage. His BP was elevated, ranging from 150/70 to 180/80 mmHg. The ECG showed sinus tachycardia with incomplete right bundle branch block (Figure 3B). The echocardiogram revealed three leaflet aortic valves with severe aortic regurgitation and fair left ventricular systolic function, LVEF of 0.50. The cardiac troponin T was elevated, 222ng/ml. The CAT scan showed Standford type A, acute aortic dissection, starting from the ascending aorta to the descending part so he was urgently referred to Bangkok Heart hospital. Atenolol and nitroglycerin were given to control the BP and the heart rate. On arrival, he was conscious with no palpable pulse in all extremities and only faint Doppler signal was seen at the left foot. The chest film showed an obvious widening superior mediastinum, diffuse interstitial infiltration of both lungs and minimal left pleural effusion (Figure 3A). The repeat 256 slide MDCT scan showed two entry sites at the proximal ascending aorta, just above the origin of the left main and right coronary arteries, and distal arch, with an extending flap to the aortic bifurcation and involved right common iliac artery (Figure 4A-E). After the risks and benefits of surgical intervention were explained and informed consent obtained, an emergent operation was performed for 10.7 hours. The time taken for a total aortic cross clamp and bypass was 293 and 348 minutes respectively. There were two ruptured sites, the first at the ascending aorta, 1 cm above the left coronary sinus and the second between the left subclavian and common carotid arteries. There was malcoaptation of the aortic valve causing severe regurgitation. The maximal diameter of the ascending aorta was 4 cm and no bloody pericardial effusion was found. The ascending aorta, aortic sinus and aortic valves were excised and replaced with no.25 St. Jude composite graft (Bentall procedure). Both coronary ostia were reattached to the ascending aorta. A total arch replacement (24/10/8/8 mm Galweave 4 branches plexus) and re-implantation of innominate, left common carotid, left subclavian arteries were performed, see Figure 5. The patient recovered well. Initially he had myonecrosis with a peak of creatinine phosphokinase (CPK) of 43,871 unit/L which compromised the kidney function (the serum creatinine rose from 1.3 to 4.33 mg/dl). With a forced diuresis and alkalinizing urine, the urine flow was fairly good and CPK was normalized.

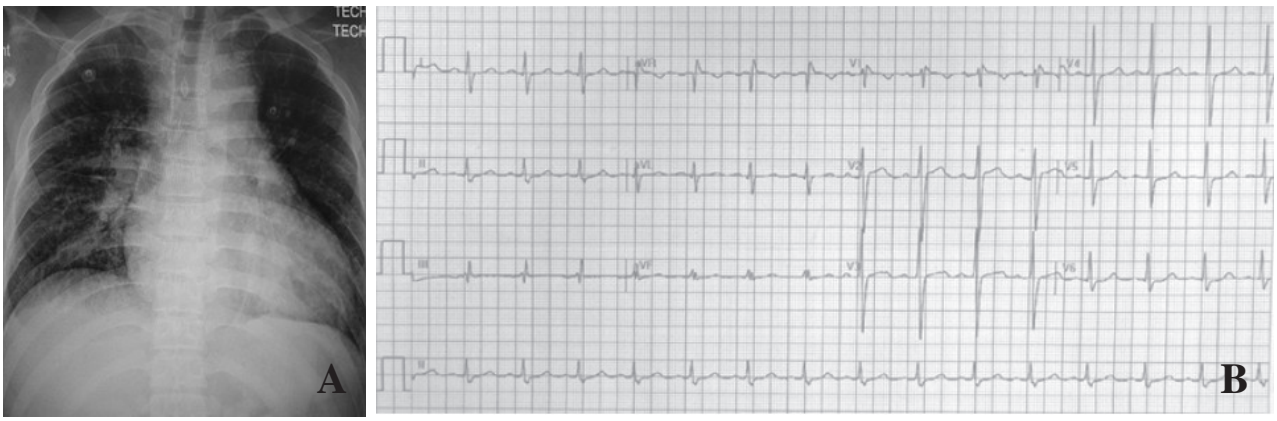

Figure 3A: Widening superior mediastinum with cardiomegaly was seen in chest film of case no.2,
3B: ECG showed sinus rhythm with incomplete right bundle branch block

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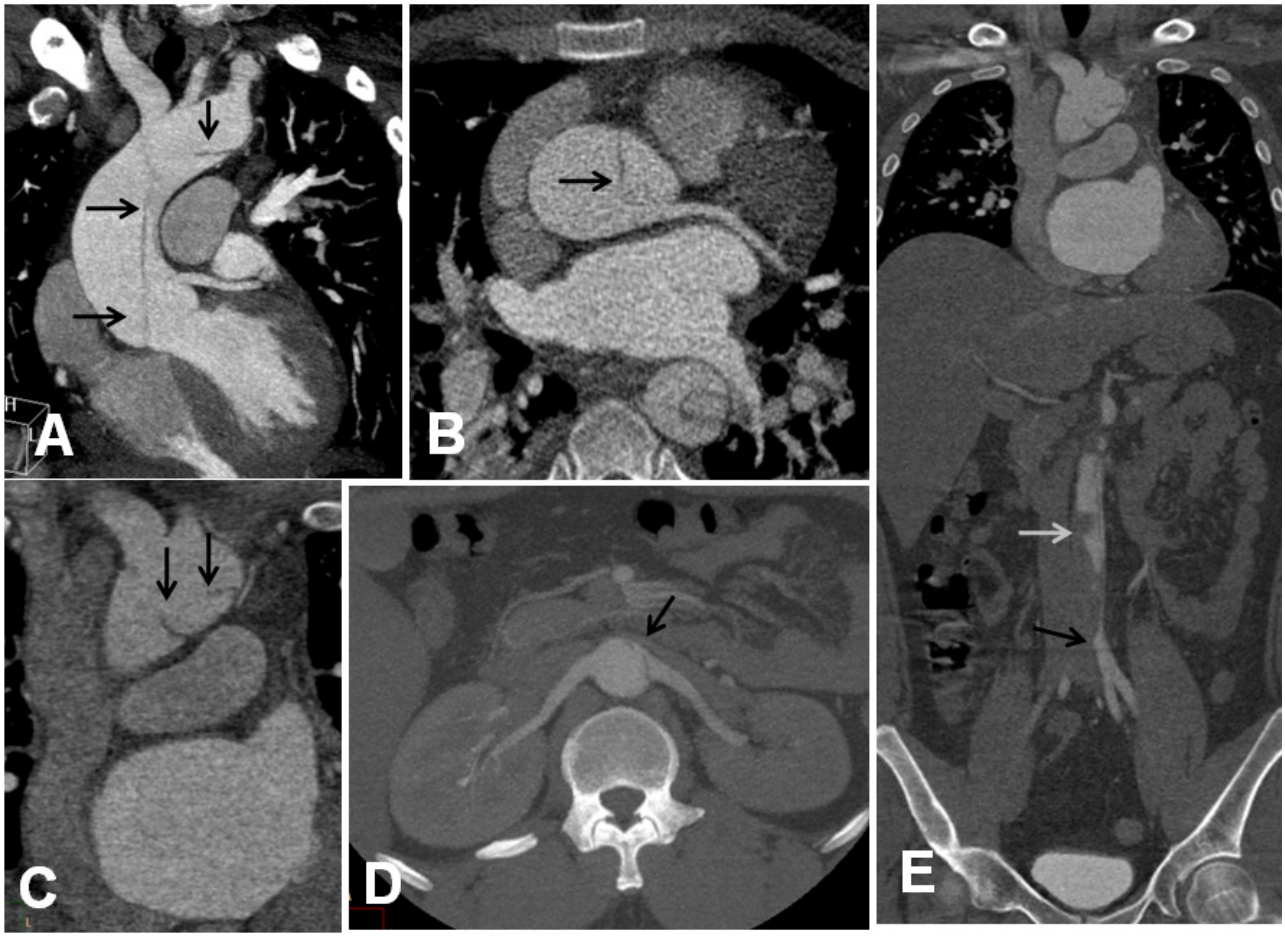

Figure 4A-E: Computerized axial tomogram (CAT) of case no.2 showed an ascending aortic dissection, Standford type A (arrow A), extending down to iliac artery. The proximal entry site was just above the left main origin (B, arrow). The distal entry site was between innominate and subclavian arteries (C, arrow). The dissection flap compromised the left renal (arrow D) and right common iliac arteries (E, black arrow). Intramural thrombus was also noted (white arrow, E).

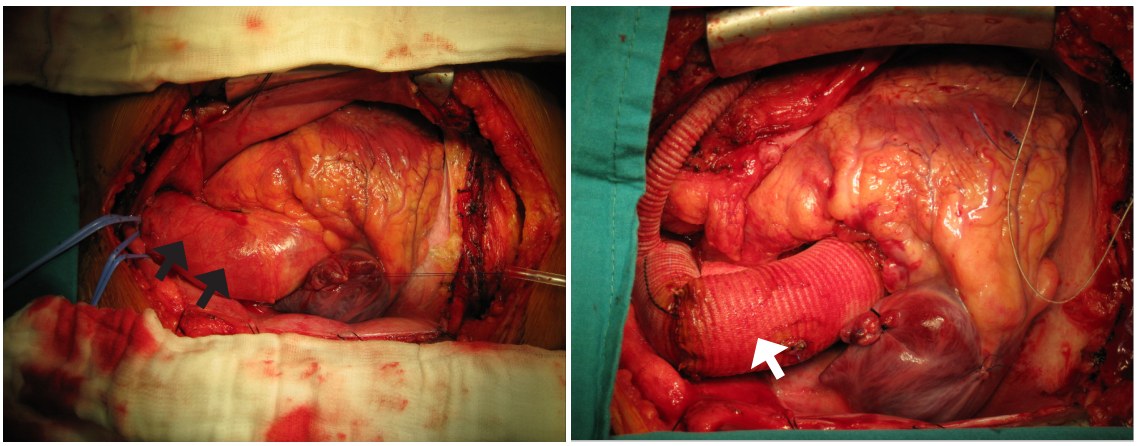

Figure 5: A dissected ascending aorta of case no. 2 was shown before (black arrow, A) and after resection with conduit graft replacement (white arrow, B).

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Discussion

Acute aortic syndrome, the highly fatal disease

Acute aortic syndrome refers to a group of lethal aortic diseases that share a common pathological background, acute aortic dissection (AD), intramural hematoma (IH) and symptomatic penetrating aortic ulcer (PAU).[1] Of these, acute AD is the most dangerous condition and carries the highest fatality rate. Its incidence ranges from 2.6 to 3.5 cases per 100,000 person-years.[1-4] The mortality rate of acute ascending AD (Stanford type A) patients increases with time after the onset of symptoms, by a factor of 1-2% per hour[5,6] while the 30-day mortality rate of uncomplicated descending AD (Stanford type B) cases is lower, at around 10%.[7] It is clear that acute ascending AD requires a rapid diagnosis and emergent surgical repair (1-7). In IH, the mortality is highest in the aortic sinus part (60%), followed by ascending aorta (50%), proximal arch (33%) and decreasing in distal arch (7%), descending thoracic aorta (13%) and abdominal aorta (0%).[8] Recently, we reported on a fatal penetrating aortic ulcer and aorto-iliac occlusion in a 56-year-old man who had a cardiac arrest and died from severe metabolic acid and multi-organ failure.[9]

Diverse clinical manifestation made the diagnosis of AD more difficult

The clinical manifestation of acute AD patients varies widely, depending on which arterial branches are involved. Therefore the presenting symptoms may be overlapping with any acute arterial occlusion such as acute coronary syndrome, and stroke.[1] Data from the international registry of acute AD (IRAD), based on 464 cases, indicated that most victims were men (65%) with a mean age of 63 years.[7] Other predisposing risk factors of developing AD were: hypertension (72%); history of atherosclerosis (32%); prior history of cardiac surgery (18%); Marfan syndrome (5%); and iatrogenic causes (4%, i.e. postvalvular, aortic surgery and vascular intervention).[1,7] The majority of acute AD cases presented with acute severe chest pain and only 4.5% of them experienced no pain.[1,10] Other presenting symptoms could be syncope, TIA, stroke[11], angina or myocardial infarction[12,13], and ischemic limbs with pulse deficit.[1] On rare occasions, an AD patient might come with supra-sternal bruising and neck swelling.[14]

Typical case presentation and high risk predictors

The first patient truly represented a typical case of Stanford type A, acute AD. This 71-year-old hypertensive man experienced chest pain which is common in proximal AD cases.[1] In addition, he had a syncopal attack and hypotension which were the risk predictors of death. In the IRAD registry, syncope was regarded as a poor prognostic indicator and was found in 13%. Syncopal cases trended to have proximal AD, more serious complications (i.e. cardiac tamponade, stroke and neurological deficit), and carried higher in-hospital mortality, 34%, vs 23% of cases who had no syncope.[15] In this particular case, moderate pericardial effusion was detected by echocardiogram suggested that the blood was dissecting down into the pericardial space, causing cardiac tamponade and shock. According to Mehta et al, tamponade, shock and hypotension are the strong independent predictors of in-hospital mortality with the odd ratio of 2.97 (95% CI 1.8-4.8), see Table 1.[16] In addition, the difference in systolic pressure between right and left arms (in this case 90 and 120 mmHg), as coded in the ACC/AHA guidelines of diagnosis and management of patients with thoracic aortic disease, is also indicated as a high-risk examination feature.[17] In cardiac tamponade cases, pericardiocentesis is still a matter for debate issue since it can make the patient's symptoms worse.[18] Owing to the late presentation (12 hours), all high-risk predictors and the delayed diagnosis, the patient died before referral.

Atypical proximal AD case with congestive heart failure

In contrast, the second case was of a quite young man who had no known predisposing factors of developing AD. Generally speaking, the younger AD cases (age < 40 years) are more likely to have Marfan syndrome, bicuspid aortic valve or prior aortic surgery[1] and none of these were found in this case. He smoked 1-4 cigarettes/day, had a small intake of beer or whisky but denied using cocaine or other drugs. It is possible that he might have asymptomatic hypertension as it is present in both sides of his family. He denied any history of blunt chest injury or vascular surgery in the past. On physical examination, there was no evidence of Marfan syndrome or other connective tissue disease and overt congestive heart failure from aortic regurgitation murmur was appreciated. The echo-cardiogram confirmed severe aortic valve leakage with three leaflets and no other valvular pathology was detected.

Unlike the first case, he did not have chest pain but had back and right ankle pain for two weeks. In type B, AD, back and/or abdominal pain were quite common.[1,7] It is possible that the dissecting blood first entered the wall at the distal arch, moving down to the abdominal aorta and occluded the right common iliac artery, see figure 4E. Operative findings confirmed two ruptured sites, one at the distal arch and another at the aortic root just above the left coronary sinus. As the blood entered the proximal entry site, it dissected into the aortic valve, causing severe regurgitation and congestive heart failure. A pulse deficit and murmur of aortic regurgitation were also the high risk features in this case.[13] The patient underwent complex surgical repair with no complication but the cause of AD remained unclear. Screening for family members was carried out as suggested in the current guidelines.[13]

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Conclusion

Acute ascending AD is one of the most catastrophic diseases that requires rapid diagnosis and surgical treatment. Missed or delayed diagnosis is quite possible and results in poor surgical outcomes or eventually death. We reported two cases of acute AD who presented with two different cardiac manifestations. The first one was typical Stanford type A AD who had syncopal attack from cardiac tamponade and died within 12 hours before surgery. The second case was a relatively young man, had no underlying disease, initially had Standford type B AD which later involved ascending aorta, causing severe aortic regurgitation and congestive heart failure. He survived complex surgical repair, aortic valve replacement and was discharged home. We hope that our reported cases will raise clinical attention for the early detection and treatment of this high fatality disease.

Table 1: Independent predictors of in-hospital death[12]

Variables	Overall Type A,%	Among survivors,%	Among death,%	Parameter Coefficient	*p*	Odd ratio for death (95%CI)
Age >70 yr	35.2	30.0	46.1	0.53	0.03	1.70 (1.05-2.77)
Female gender	34.5	30.7	42.7	0.32	0.20	1.38 (0.85-2.27)
Abrupt onset of pain	84.5	82.3	89.0	0.96	0.01	2.6 (1.22-5.54)
Abnormal ECG on presentation	69.6	65.2	79.5	0.57	0.03	1.77 (1.06-2.95)
Any pulse deficit on presentation	30.1	24.7	41.1	0.71	0.004	2.03 (1.25-3.29)
Kidney failure on presentation & before surgery	5.6	2.9	11.9	1.56	0.002	4.77 (1.80-12.6)
Hypotension/shock/tamponade	29.0	20.1	47.1	1.09	<0.0001	2.97 (1.83-4.81)

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