

76-Year-Old Woman With Syncope and Right-Sided Weakness



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A 76-year-old woman presented to the emergency department following a syncopal episode. She was in her usual state of health when she collapsed from a seated position. She regained consciousness before Emergency Medical Services arrived. There was no witnessed seizure activity. Point of care blood glucose was 111 mg/dL (70 to 140 mg/dL). Upon presentation to the emergency department, the patient was noted to have new-onset neurologic deficits.

The patient's past medical history was significant for hypertension, hyperlipidemia, and osteopenia. She had no known neurologic or cardiovascular disease. Home medications included alendronate 70 mg weekly, cholecalciferol 1000 units daily, aspirin 81 mg daily, and lisinopril 10 mg daily.

Vital signs on presentation were as follows: blood pressure, 97/57 mm Hg; pulse, 53 beats per minute; temperature, 35.9 °C (96.62 °F); and respiratory rate, 14 breaths per minute with adequate saturation. She required atropine and vasopressor support for hypotension and bradycardia. Examination results were notable for Glasgow Coma Scale of 11, with right-sided hemiparesis, no spontaneous speech, and no response to verbal commands. She had no external injuries. Her pupils were equal in size and reactive to light.

Laboratory evaluation revealed the following (reference ranges provided parenthetically): hemoglobin, 12.8 g/dL (11.6 to 15.0 g/dL); platelet count, 195 x 10⁹/L (157 to 371 x 10⁹/L); leukocytes, 22.1 x 10⁹/L (3.4 to 9.6 x 10⁹/L); sodium, 141 mmol/L (135 to 145 mmol/L); creatinine, 0.77 mg/dL (0.59 to 1.04 mg/dL); total calcium, 7.8 mg/dL (8.8 to 10.2 mg/dL); pH, 7.21 (7.35 to 7.45);

pCO₂, 51 mm Hg (32 to 45 mm Hg); pO₂, 50 mm Hg (83 to 108 mm Hg) and HCO₃, 21 mmol/L (22 to 26 mmol/L); lactate, 2.7 mmol/L (0.50 to 2.20 mmol/L); prothrombin time, 20.4 seconds (9.4 to 12.5 seconds); activated partial thromboplastin time (APTT), 69 seconds (25 to 37 seconds); fibrinogen, <35 mg/dL (200 to 393 mg/dL); and 0-, 2-, and 12-hour high-sensitivity troponins T were 9, 46, and 262 ng/L, respectively (<10 ng/L).

Chest x-ray film showed slight tortuosity of thoracic aorta and possible widening of the mediastinum. Computed tomography (CT) of the head demonstrated extensive periventricular white matter leukomalacia, most likely secondary to small-vessel ischemic disease, with no evidence of parenchymal hemorrhage or mass effect. Tissue plasminogen activator (tPA) was administered.

1. Which one of the following diagnostic tests is most likely to reveal the underlying cause of the patient's stroke?

- CT chest angiogram
- Coronary angiogram
- Serial troponin
- Ventilation-perfusion (VQ) scan
- Electrocardiogram

This patient's presentation with ischemic stroke, shock, and widened mediastinum on chest radiograph is concerning for an acute aortic dissection (AAD). CT angiogram (CTA) is first line to confirm this diagnosis. Magnetic resonance angiography (MRA) may be used but requires more time to acquire and may not be universally available across all hospitals. In patients too hemodynamically unstable to transport, bedside transesophageal echocardiography can be considered.¹

See end of article
for correct answers
to questions.

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Coronary angiogram and serial troponin can be used to evaluate acute coronary syndrome but play no role in definitive diagnosis of AAD; VQ scan can be considered to evaluate for pulmonary embolism but would not diagnose aortic disease. Electrocardiography may be in the initial work-up but would not definitively diagnose an aortic dissection.

A CTA chest scan was obtained and demonstrated a type A acute aortic dissection of an aneurysmal ascending thoracic aorta, measuring 60 mm in greatest diameter. The dissection extended from the aortic root to the left subclavian artery, with extension into both common carotid arteries and the right subclavian artery. This was associated with a lack of perfusion in the right coronary artery, caused by occlusion from the false lumen.

2. Which one of the following is most commonly seen on presentation of this condition?

- a. Syncope
- b. Hypotension
- c. Chest pain
- d. Back pain
- e. Stroke

The International Registry of Acute Aortic Dissection (IRAD) was established in 1996 and now includes >7300 cases. In this registry, syncope was reported in 13% of cases. Hypotension and shock were described in >25% of patients and were associated with neurologic deficits, ischemia, and worse prognoses. Severe chest pain was the most frequently seen presenting symptom occurring in 79% of patients with type A dissection and 63% of those with type B dissection. Back pain was seen in 43% of patients with type A dissection and 64% of those with type B dissection. Major brain injury was described in 10% of patients with type A dissection and usually indicated arch vessel involvement.²

The patient's presenting symptoms of syncope, neurologic deficits, and shock can be explained by the involvement of the carotid arteries and right coronary artery noted in CTA.

3. Which one of the following is the most commonly identifiable risk factor for the patient's underlying condition?

- a. Hypertension
- b. Tobacco use
- c. Genetically mediated aortopathy
- d. Diabetes
- e. Trauma

Hypertension is the most commonly identified risk factor for aortic dissection and represents an important variable in primary prevention. Studies indicate that up to 86% of those diagnosed with aortic dissection have histories of hypertension.³

Smoking increases the risk of aortic pathology, with smokers suffering from aortic dissection twice as frequently as nonsmokers.³

Contribution of genetic abnormalities to the development of aortic aneurysm is more prominent than previously considered. Although syndromic conditions such as Marfan or Loeys Dietz syndromes are widely recognized, nonsyndromic conditions with isolated cardiovascular abnormalities also exist, such as bicuspid valve-associated aortopathy.⁴

Diabetes is not associated with an increased risk of aortic dissection. Some studies even indicate a protective effect with lower rates of incidence and reduced mortality in patients with diabetes.³ Although blunt trauma can cause transection of the aorta at fixed points—such as the aortic isthmus, sinotubular junction, or distal aortic arch—true traumatic dissection is uncommon.⁵ Other risk factors include male sex, advanced age, inflammatory disease, and aortic aneurysm.

Our patient has a history of hypertension, which likely contributed to her presentation. However, nonsyndromic genetic conditions predisposing the patient to aortic dissection cannot be ruled out entirely.

4. Which of the following is the most appropriate immediate next step in management of this case?

- a. Blood-pressure control
- b. Pain control

- c. Surgical consult
- d. Dual antiplatelet therapy
- e. Endovascular repair

Perioperative stabilization with blood-pressure management and pain control should not delay surgical intervention for Stanford type A aortic dissections. For settings in which surgical intervention is not possible, patients should be stabilized with aggressive blood-pressure control and transferred to the nearest center with operative capability.⁵ For most patients, this means systolic blood pressure (SBP) < 120 mm Hg and heart rate < 60 bpm, preferably through the use of beta blockers.⁶

Aortic dissections are commonly misdiagnosed as myocardial infarctions, delaying proper management. Although dual antiplatelet therapy is a mainstay in treating myocardial infarction, it plays no role in treatment of AAD.

Thoracic endovascular aortic repair may be considered for patients with type B dissection; however, open surgical repair remains the gold standard for type A dissection.⁷ Of note, there are limited data on the use of endovascular repair for genetically mediated aortic disease; therefore, this is not yet recommended.⁸

The surgical team assessed the patient for operative intervention. Owing to the complexity of the presentation and the recent administration of tPA, our patient was not a surgical candidate. She remained intubated and was transferred to the cardiac intensive care unit for medical management.

5. In those patients who undergo successful surgical repair, what is the recommended follow-up?

- a. No additional follow-up needed
- b. Routine follow-up with primary care provider
- c. Coronary angiogram in 1 year
- d. Chest CTA or MRA at 1, 3, 6, and 12 months
- e. Annual transthoracic echocardiographic imaging

In those who undergo successful treatment of type A aortic dissection, routine surveillance

is recommended by the Society of Thoracic Surgeons (STS) and the American Heart Association (AHA) guidelines. A lack of follow-up or sole primary care follow-up would not be recommended in this case. Coronary angiography would not provide relevant data for surveillance. The current STS/AHA guidelines recommended surveillance imaging with CT or MRI at 1, 3, 6, and 12 months after intervention and yearly thereafter.⁹

Transthoracic echocardiography may be used for surveillance in patients who undergo proximal aortic surgery but is not adequate in patients treated for type A aortic dissection.⁹

In our patient, clinical status deteriorated with medical management in the intensive care setting. Ultimately, the patient was transitioned to comfort care and died.

DISCUSSION

Acute aortic dissection is an acute aortic syndrome involving the formation and propagation of an aortic false lumen, resulting in complications such as malperfusion, tamponade, and aortic-valve insufficiency. Certain variants and presentations can have catastrophic outcomes, with high mortality rates, despite prompt intervention.⁶

Studies demonstrate an incidence rate of approximately 2.6 to 3.5 cases per 100,000 person-years. Data from IRAD revealed a mean age of 63 years with a 2:1 male predominance. Hypertension was the most common predisposing factor, present in 76.6% of patients diagnosed with aortic dissection. Other common risk factors included atherosclerosis (27%), known aortic aneurysm (16%), previous cardiac surgery (16%), Marfan syndrome (5%), and iatrogenic causes (4%).²

Aortitis, or inflammation of the aorta, also increases the risk of acute aortic syndromes and may be caused by infectious or noninfectious etiologies. Noninfectious aortitis typically occurs because of systemic diseases such as giant-cell arteritis. There is also a variant without systemic features, known as clinically isolated aortitis (CIA). Traditionally, CIA is diagnosed on tissue pathology following surgical resection of aneurysm or dissection. In our case, tissue was not

available, and autopsy was not performed. Clinically isolated aortitis may also be identified radiologically with increased uptake on positron emission tomography, aortic-wall thickening with enhancement on MRI, or wall thickening or aneurysm on CT.¹⁰ Genetic predisposition may play a larger role in the pathogenesis of thoracic aortic aneurysm and dissection than previously understood. Although syndromic conditions with phenotypic abnormalities such as Marfan and Loeys-Dietz syndromes are commonly recognized, nonsyndromic conditions with isolated cardiovascular abnormalities also exist. This includes bicuspid valve-associated aortopathy, a condition commonly associated with thoracic aortic aneurysm. Genetic mutations involving the transforming growth factor beta-signaling cascade and smooth-muscle contractile apparatus have been discovered, although most presenting patients have poorly understood genetic predispositions.⁴

Acute aortic dissection can present with a variety of symptoms. Sudden-onset severe chest or back pain is the most frequently seen. Patients with abdominal pain alone have higher mortality rates, likely owing to delayed diagnosis or potential visceral malperfusion. Painless aortic dissection may also occur and is associated with syncope, congestive heart failure, stroke, and subsequently higher risk of mortality. Additional complications associated with type A AAD include shock (15%), pericardial tamponade (18%), mesenteric malperfusion (3.7%), and major brain injury (10%).² In our case, atypical presentation led to the administration of tPA, which subsequently affected treatment options. Acute aortic dissection should be in the differential diagnosis for patients presenting with acute altered mental status, syncope, and possible stroke.

Variability in AAD presentation leads to a misdiagnosis rate of 14% to 39%, with the potential for delay in care and resultant morbidity and mortality.¹¹ To reduce misdiagnosis and overtesting, the American Heart Association/American College of Cardiology 2010 guidelines proposed the Aortic Dissection Detection Risk Score (ADD-RS). This

risk stratifies patients based on the presence of 3 risk-factor categories: high-risk conditions (Marfan syndrome, family history of aortic disease, known aortic valve disease); high-risk pain features (abrupt and severe chest, back, or abdominal pain); and high-risk examination features (evidence of perfusion deficit, new aortic insufficiency murmur, hypotension or shock). This tool was validated using the IRAD database in 2011, with a sensitivity of 95.7%.¹²

The use of D-dimer was recently added to this algorithm as a secondary filter to those who had $ADD-RS \leq 1$. The ADVISED study (Aortic Dissection Detection Risk Score Plus D-Dimer in Suspected Acute Aortic Dissection) evaluated the efficiency and failure rate of AAD rule out with the use of ADD-RS and D-dimer. It suggested that although patients with $ADD-RS > 1$ should receive conclusive imaging regardless of D-dimer, those with $ADD-RS \leq 1$ could be further risk stratified. In this group, those with D-dimer ≥ 500 ng/mL should receive conclusive imaging, whereas those with D-dimer < 500 ng/mL can have AAD ruled out without further imaging. The study suggested that use of this method could avoid up to 1 in 2 CTA examinations in all patients presenting with suspected AAD.¹¹ In our case, D-dimer was not collected for risk stratification.

In those cases that warrant imaging, CTA is typically first line, given its availability and reliability with sensitivity and specificity approaching 95% to 98%. In the IRAD cohort, CT imaging was used to diagnose 63% of patients. This modality provides rapid diagnosis of aortic disease and additional information regarding large-vessel and side-branch involvement, allowing for surgical planning. Drawbacks include need for transportation to radiology suite, radiation exposure, and risk of contrast nephropathy.

MRI or MRA can also be used to image thin intimal flaps, intramural processes, and morphology of aortic-wall inflammation for vascular-disease classification but is limited in the acute setting by increased examination times and a lack of general availability. Imaging can take upward of

60 minutes and requires that patients hold their breaths to best capture dissection flaps.

Although the use of echocardiography has waned over time,² it can be considered for those patients who are too hemodynamically unstable for transport or who have severe contrast allergy. Transthoracic echocardiogram (TTE) alone has a sensitivity of only 59% to 83% in diagnosing all forms of AAD and is therefore not the modality of choice in most settings. Transesophageal echocardiogram (TEE), however, can reach a sensitivity of 99%, with a specificity of 89%. Echocardiography is also limited by an inability to evaluate anatomic details for endovascular intervention planning.¹

Aortic dissections are classified by the location of the lesion. The Stanford classification criterion distinguishes between type A dissections, involving the ascending aorta with or without descending aorta involvement, and type B dissections, which involve only the descending aorta.⁶ These classifications help to determine treatment and prognosis.

Type A aortic dissections are surgical emergencies, typically requiring resection of the aorta with the intimal tear involvement and consideration of aortic root/arch replacement. Medical management should not delay surgical intervention.⁵ Patients with uncomplicated type B dissections can be managed medically. Management consists of reduction of systemic arterial pressure and cardiac contractility in addition to adequate analgesia. Hemodynamic goals include SBP < 120 mm Hg and heart rate < 60 bpm, preferably through the use of beta blockers. Endovascular intervention should be considered in those with persistent or recurrent pain, aortic expansion, progression of dissection, or end-organ malperfusion.⁶

All patients should receive life-long treatment of hypertension with beta blockers as the preferred choice and a recommended goal blood pressure of < 120/80 mm Hg.⁶ Some guidelines suggest follow-up assessment of the aorta at 1, 3, 6, 9, and 12 months after discharge and yearly thereafter.⁹ In approximately 12% to 30% of patients, repeat surgical intervention is required for complications such as extension or recurrence of

dissection, formation of aneurysm, graft dehiscence, aortic regurgitation, or infection.⁶

Prognosis of AAD is dependent on classification and presentation. Without immediate surgical management, type A dissections have a mortality rate of 24% at 24 hours and 49% at 14 days. With surgical intervention, mortality rates remain as high as 10% at 24 hours and 20% at 14 days.⁶ Long-term rates of survival have been reported as high as 75% at 10 years by some studies.⁵ Type B dissections are less lethal, with a 30-day mortality rate of approximately 10%. Mortality rates are highest in those with complications of pericardial tamponade, myocardial infarction, and stroke. Other factors contributing to increased mortality rate include older age, hypertension, cardiac tamponade, renal failure, and pulse deficits.⁶

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CORRECT ANSWERS: 1. a. 2. c. 3. a. 4. c. 5. d.