Abdominal aortic aneurysm

JAMES K. DAVID, MD, SUE S. BORNSTEIN, MD, AND LYNDAKAY G. MYERS, MD

CASE PRESENTATION

JAMES K. DAVID, MD: A 68-year-old retired draftsman with chronic obstructive pulmonary disease (COPD), type 2 diabetes mellitus, and systemic hypertension presented to a hospital emergency department with a 5-day history of worsening dyspnea and abdominal distention and a 1-day history of a nonproductive cough and wheezing. One week earlier he had been discharged from the hospital with a diagnosis of COPD exacerbation, and his prednisone dose had been gradually decreased. His current dyspnea occurred at rest and worsened with minimal exertion. He denied chest pain, abdominal pain, fever, chills, nausea, and vomiting. The patient's father had died of COPD. The patient had an 80-pack/year history of cigarette smoking, drank alcohol occasionally, and denied using illicit drugs. Medications included prednisone (2 mg a day orally), fluticasone propionate, salmeterol xinafoate, ciprofloxacin, guaifenesin, triamterene and hydrochlorothiazide, glipizide, carisoprodol, and home O₂ per nasal canula at 2 L/min.

The patient's blood pressure was 137/89 mm Hg; heart rate, 120 beats per minute; and temperature, 36.6°C (97.8°F). He was obese, alert, and oriented and was in moderate respiratory distress, using his accessory respiratory muscles. His extraocular muscles were intact, and his pupils were equal, round, and reactive. His fundi were normal. His oral pharynx contained a whitish plaque on the soft palate. His neck was supple, with no jugular venous distention, lymphopathy, or bruits. He was barrel chested with bilateral wheezing and poor air movement. He had distant heart sounds, without murmur. His abdomen was soft, nontender, and mildly distended. His bowel sounds were normal. A reducible umbilical hernia was present. Neither the liver nor the spleen was enlarged. No abdominal masses were palpated. His extremities were devoid of cyanosis, clubbing, or edema. The pulses were 2+/4+ throughout. Diffuse ecchymoses were present on all extremities. Neurological examination disclosed no abnormalities.

His blood glucose was 290 mg/dL, and his leukocyte count was $17 \times 10^3/\mu\text{L}$ with 82% segmented neutrophils, 3% bands, 8% lymphocytes, and 6% monocytes. His hematocrit was 42%. Room air arterial blood gas showed a pH of 7.31; PCO₂, 59; PO₂, 69 (93% saturation); and CO₂, 29.

Electrocardiogram showed sinus tachycardia (105 beats per minute) and low voltage. Chest radiograph showed hyperinflation and chronic changes consistent with COPD, bilateral scarring of the lung bases, some blunting of the left costophrenic angle, and no acute process.

The patient was admitted with a diagnosis of COPD exacerbation. He was started on an O₂ protocol, albuterol nebulizer therapy, intravenous levofloxacin, and intravenous methylprednisolone sodium succinate. The diabetes mellitus was treated with glipizide, a regular sliding scale insulin, and an American Diabetes Association diet. He was continued on triamterene and hydrochlorothiazide for systemic hypertension. The thrush was treated with oral fluconazole.

On hospital day 1, the patient's breathing difficulty lessened. He complained for the first time of lower back pain. It lessened with hydrocodone bitartrate and acetaminophen.

On hospital day 2, the patient's breathing difficulty further improved. He complained of an additional episode of lower back pain which lessened with hydrocodone bitartrate and acetaminophen. He also complained of a moderate midepigastriac right upper quadrant abdominal discomfort that lasted 1 hour before resolving. Methylprednisolone sodium succinate was changed to a tapering dose of prednisone.

On hospital day 3 at 3:00 AM, the patient was found on the rest room floor by a nurse. He was lethargic but responsive. His peak systolic blood pressure was 50 mm Hg, and his heart rate was 124 beats per minute. Arterial blood gas showed a pH of 6.88; PCO₂, 152; PO₂, 118; O₂ saturation, 94%; and CO₂, 27. Electrocardiogram showed sinus tachycardia (124 beats per minute), mild ST depression in the anterior leads, and an R wave in V₁ and V₂. A central line was placed; a dopamine drip was initiated. The patient was transferred to the intensive care unit and intubated. O₂ saturation initially remained >90% but later decreased. Systolic peak blood pressure remained low (30 to 80 mm Hg) despite maximal doses of dopamine and large amounts of intravenous fluids. An arterial line was placed; a levophed drip was added. The patient was given 100 mg of intravenous hydrocortisone, and the levofloxacin was changed to intravenous sterile piperacillin sodium and tazobactam sodium. Chest radiograph showed no acute changes. Serial troponin-I initially was >0.4 (normal) and later was 0.9 (borderline). D-Dimer was >6.4 (normal, <0.2). A Swan-Ganz catheter was placed and recorded the following values: cardiac index, 1.1 to 1.8 L/min; pulmonary ar-

From the Departments of Internal Medicine (David and Bornstein) and Pathology (Myers), Baylor University Medical Center, Dallas, Texas. Dr. David is now a radiology resident at the Massachusetts General Hospital in Boston.

Corresponding author: Sue S. Bornstein, MD, 3801 Gaston Avenue, Suite 200, Dallas, Texas 75246.

terial wedge pressure, 29 mm Hg; and systemic vascular resistance, 1598 dynes-sec⁻¹-cm⁻⁵ (later decreasing to normal).

Transthoracic echocardiogram disclosed a left ventricular ejection fraction of 55% and no evidence of pericardial tamponade. Right ventricular systolic pressure was 49 mm Hg. Despite maximal dosing of dopamine, levophed, and neo-synephrine, the blood pressure remained low, and urine output was zero. The hematocrit decreased throughout the day from 42% to 33% to 29% after large amounts of intravenous fluids.

At 4:00 PM, heparin and thrombolytics were given because of a suspected diagnosis of pulmonary embolus. The patient died at 8:05 PM with intractable hypotension with asystole.

CASE DISCUSSION

SUE S. BORNSTEIN, MD: This patient had been discharged from the hospital 1 week before this admission for what seemed like a similar illness. On this admission, he reported a 5-day history of increasing dyspnea and a 1-day history of nonproductive cough and wheezing. He was on a tapering course of prednisone on admission. For 1 week he had mild abdominal distention without pain. He was in moderate respiratory distress, and he had thrush. He had no abdominal masses or tenderness. There were diffuse ecchymoses of the extremities.

His blood glucose was 290 mg/dL, and the white blood cell count was $17 \times 10^3/\mu\text{L}$ with a left shift. The hematocrit was 42%. One might expect his hematocrit to be a little higher because of the possible chronic hypoxemia. His platelet count was $151 \times 10^3/\mu\text{L}$. His blood gases showed acidemia with an elevated PCO₂ and an increased CO₂ with mild hypoxemia on room air. He had an increased anion gap at 18. I believe he had chronic respiratory acidosis with superimposed metabolic acidosis.

He was treated for acute exacerbation of COPD. His respiratory status improved, but he had an episode of lower back pain, which was treated with hydrocodone bitartrate and acetaminophen. On hospital day 2, he had another episode of low back pain that improved. He also had a 1-hour episode of moderate midepigastria and right upper abdominal pain. A sonogram was ordered but was not performed.

He was then found on the floor by the nurse on the morning of hospital day 3. He was hypotensive, tachycardic, and hypoxemic. On 100% non-rebreather mask, his O₂ saturation was only 77%. His blood gases showed profound respiratory acidosis with a large alveolar-arterial gradient. The electrocardiogram, which earlier had shown only sinus tachycardia and low voltage, now showed myocardial ischemia. He was resuscitated with fluids and vasopressors, but he remained hypotensive throughout the day. He was given 100 mg of hydrocortisone but to no avail. D-Dimer was >6.4, suggesting disseminated intravascular coagulation. His platelet count, fibrinogen level, and other markers of disseminated intravascular coagulation are unknown. No mention is made of any physical findings during the resuscitation.

A Swan-Ganz catheter was placed. Echocardiogram showed normal left ventricular function without tamponade physiology. The right ventricular systolic pressure was elevated, a finding consistent with pulmonary hypertension. He was a chronic cigarette smoker, and he could have had some pulmonary hypertension on that basis. He continued to deteriorate throughout the day. He was given heparin and thrombolytics in an attempt to

treat a suspected pulmonary embolus, but he died 4 hours later from intractable hypotension and asystole. His hematocrit dropped from 42% to 29% throughout the day, although he did receive massive amounts of fluid.

I will first review *extra-abdominal causes of acute abdominal pain* that are potentially catastrophic.

Myocardial ischemia must always be considered. This man was at risk for coronary artery disease. The electrocardiographic changes and the elevated troponin suggest myocardial ischemia. There was possible ischemia in the anterolateral leads plus R waves in V₁ and V₂, which suggest posterior wall myocardial infarction. These changes also are consistent with acute right ventricular strain from a pulmonary embolus. The decreased cardiac output suggests acute myocardial infarction. Although not particularly elevated initially, the pulmonary arterial wedge pressure did rise later. The central venous pressure was only modestly elevated. Initially, the systemic vascular resistance was elevated, but it decreased later. In cardiogenic shock, the systemic vascular resistance should be elevated (perhaps ≥ 2000 dynes). Also, the patient was on maximal doses of vasopressors, including dopamine, which in high doses can increase the wedge pressure.

Another important potential cause of this patient's death is *massive pulmonary embolus*. This is what he was ultimately treated for. In patients with COPD, the diagnosis can be difficult. Our patient had a few of the signs of pulmonary embolism. He had dyspnea, cough, tachypnea, tachycardia, and finally shock. A widened alveolar-arterial gradient is present in most patients with pulmonary embolus. Our patient had an elevated right ventricular pressure, a finding consistent with a pulmonary embolus. However, the mildly elevated central venous pressure does not support this diagnosis. Also, in massive pulmonary embolus the pulmonary wedge pressure should either be normal or low (1). A study in the May 1999 issue of the *American Journal of Critical and Respiratory Care Medicine* concluded that a negative D-dimer assay is useful in excluding the presence of pulmonary embolus in patients who have symptoms present for <1 week with normal liver function and no malignancy (2). The value of D-dimer in the diagnosis of pulmonary embolus is for its negative predictive value in patients with low to intermediate predictability.

Cardiac tamponade is something to consider. Neither the echocardiogram nor the Swan-Ganz catheter numbers showed findings indicative of tamponade.

Acute adrenal insufficiency was considered. He was given hydrocortisone because he had been on steroids. We don't know if the steroid use was chronic or acute, but that treatment was not successful.

Now to life-threatening *intra-abdominal causes of acute abdominal pain*. One diagnostic possibility is a *perforated duodenal ulcer*. The pain is sudden, sharp, and severe. It is initially located in the epigastrium but quickly spreads throughout the abdomen. Hypotension and fever develop within 4 to 6 hours. Examination would be expected to reveal diffuse peritonitis and a typical board-like abdomen. A history of ulcer disease was not mentioned in our patient, and I do not believe that this is a correct diagnosis. *Ruptured diverticulum* could present a similar picture, but there was no abdominal tenderness or mass. *Mesenteric ischemia* due to acute mesenteric artery occlusion presents with pain out of proportion to the physical findings, but shock is late (3).

Figure 1. Photographs of (a) the AAA and (b) cross sections of the aneurysm disclosing in the intra-aneurysmal thrombus. The maximal diameter of the aneurysm was 6.5 cm, right to left, and 7 cm, cephalad to caudad.

I believe that the cause of death in our patient was a vascular catastrophe, specifically a *ruptured abdominal aortic aneurysm* (AAA). This is diagnosed most commonly in the seventh decade of life, and affected men outnumber women 4 to 1. It accounts for at least 15,000 deaths per year in the USA. Rupture of AAA is the tenth leading cause of death in men >55 years old (4). These patients have risk factors, particularly cigarette smoking and systemic hypertension, associated with occlusive vascular disease. There is recent evidence of a genetic disposition toward AAA (4).

Of 231 patients with ruptured AAA who presented to the Mayo Clinic, vague abdominal pain was the most common complaint (4). AAA should be considered in any elderly patient with abdominal, back, or flank pain. I believe that the episode of abdominal pain and back pain on hospital day 2 may have been the initial rupture of the AAA. If he bled retroperitoneally, which is the most common route, the bleed could have been contained for hours or days. The fundamental *clinical triad of a ruptured AAA* is abdominal or back pain of sudden onset, hypotension, and a palpable abdominal mass. Unfortunately, the complete triad is present in only about 50% to 75% of patients. I think that the ecchymoses may have been from emboli from the aorta. The abdominal distention almost certainly was due to the increasing size of the AAA.

There is a well-known association between AAA and disseminated intravascular coagulopathy (DIC). A 1983 article in the *Archives of Surgery* reported on 76 patients

who had ruptured AAA (5). In 22 patients with an infrarenal AAA, 8 had elevated fibrin split products with normal platelets and fibrinogen levels. An almost chronic type of DIC seems to exist in these patients. I think the blood gas and the profound respiratory acidosis in our patient was due in part to the decreased blood flow to the diaphragm from the hemorrhagic shock. In shock, the demand for blood flow to the diaphragm increases from 2% to 20%, and with the patient's low cardiac output and index, he simply could not supply the increased demand.

The patient had a low cardiac index throughout. Initially this was in the face of an adequate pulmonary wedge pressure. At 3:20 PM, the wedge pressure went up. He may have had transient left ventricular dysfunction on the basis of low blood flow, and he was being aggressively volume resuscitated and was receiving vasopressors. At 4:00 PM, heparin and thrombolytics were given, and at 4:30 PM, his hemodynamics began to deteriorate. I think this represents the hemodynamic effects of exsanguination due to the ruptured AAA accelerated by the thrombolytics.

AUTOPSY FINDINGS

LYNDAY G. MYERS, MD: At autopsy, neither a saddle pulmonary embolus nor smaller pulmonary emboli were present. The abdomen was distended but soft. Clotted blood was found in the retroperitoneal area; however, the peritoneal cavity was free of bloody fluid. An estimated 2000 mL of blood clot was observed within the mesenteric root and retroperitoneum. A 7-cm AAA was present caudal to the superior mesenteric artery (Figure 1). No additional aneurysms were present in the aorta. A 1.5- to 2-cm rupture site was identified on the right posterior aspect of the aorta, 7 cm cephalad to the bifurcation. A thrombus was present within the aneurysm. The cause of death was ruptured AAA in the presence of atherosclerosis.

The heart weighed 410 g and floated in water because of the excessive fat (Figure 2). No myocardial lesions were present. The 4 cavities were of normal size. The lumens of the major epicardial coronary arteries were narrowed <75% in cross-sectional area.

Figure 2. (a) The heart contained so much subepicardial adipose tissue that it floated in water. Only a small portion of myocardium was visible on the exterior of the heart. (b) Transverse sections of the cardiac ventricles are shown. The ventricular cavity is small and is shown as it would be in peak systole. No lesions were noted in the myocardial walls, although the coronary arteries contained diffuse atherosclerotic plaques without severe coronary narrowing. The heart weighed 410 g, which was heavier than it should have been. The cephalad portion of the atrial septum was massively infiltrated with fat (lipomatous hypertrophy of the atrial septum).

FOLLOW-UP DISCUSSION

JAMES K. DAVID, MD: An AAA is a dilation of the abdominal aorta >1.5 to 2.0 times normal. In most people this translates into a diameter >3.0 cm. It is a true aneurysm, involving all 3 layers. Most are fusiform in shape, involving the artery's entire circumference. Ninety-five percent are located below the level of the renal arteries, and many extend to or beyond the aortic bifurcation (6).

The pathogenesis of AAA is multifactorial. Ninety-five percent are atherosclerotic (7). Atherosclerotic causality is inferred because atherosclerosis is seen in other arteries in patients with an AAA (8). Familial clustering is seen in 20% of patients (8). The familial variety of AAA can be either X-linked or autosomally inherited. When comparing first-degree relatives with non-first-degree relatives, there is a relative risk of 11.6. Therefore, it is important to screen family members. Women are more frequently affected with the familial variety than men (9). Some investigators have suggested that a mutation in the gene encoding type III procollagen is involved. Others have reported abnormally high levels of collagenases, elastases, and other proteolytic enzymes in these patients (10). The remaining 5% of cases are due to inflammatory (autoimmune) processes, collagen disorders, infections, or rarely trauma (10, 11).

AAA is primarily a disease of older people, affecting 2% to 5% of the population >60 years of age. They are more common in men, with a male to female ratio of 4:1. Mortality following rupture is the tenth leading cause of death in men >55 years old (10). It is the 13th leading cause of death overall in the USA, with an estimated 15,000 fatalities each year (11).

At presentation, 75% are asymptomatic (4). An AAA is most commonly discovered as a pulsatile abdominal mass on routine examination. It also may be discovered incidentally during abdominal plain film, sonography, or computed tomography (CT).

Routine screening is controversial. Consider a screening abdominal sonogram for men >60 years old; first-degree relatives; and those with systemic hypertension, a history of smoking, peripheral vascular disease, and other peripheral aneurysms (12).

Diagnostic imaging methods include *abdominal ultrasound*, which is highly accurate, obtainable, and safe (7). It is also useful in following the size of the aneurysm. *Contrast CT* also is excellent and is the most accurate method for determining aneurysmal size. In addition, it provides useful information on aortic wall thickness. Although more costly, *magnetic resonance imaging* provides an excellent level of detail and is useful in providing a 3-dimensional presentation. *Aortic arteriogram* is not accurate in determining size because true size is obscured by mural thrombi, but it can be useful in evaluating vascular anatomy preoperatively.

Rupture is the most lethal complication of AAA. The overall mortality is 90% (9). Cronenwett et al analyzed 30 potential risk factors for rupture in patients with AAA and found that only diastolic hypertension, initial aneurysmal size, and COPD were significant (13).

Why is aneurysmal size so important? Laplace's law states that wall tension is directly proportional to pressure and diameter. The average growth rate of an AAA is 2 to 4 mm per year (6). Because of Laplace's law, larger aneurysms grow more rapidly. Larger aneurysms are clearly at greater risk of rupture (*Table*).

Table. Risk of AAA rupture based on size at time of detection and years after initial diagnosis*

Aneurysm size	Risk of rupture	
	1 year	5 years
<5 cm	4%	20%
5–7 cm	7%	33%
>7 cm	20%	>90%

*Compiled from Blackburne LH (6).

One year before the first successful AAA repair by Dubost in 1951, Estes published 5- and 10-year survival rates of 102 untreated patients (14). The results were 19% and 0%, respectively. Szilagyi et al in 1966 published a study of both surgical and nonsurgical groups (15). They found that surgical repair of AAA nearly doubled life expectancy. Crawford et al in 1981 showed that elective surgical mortality decreased from 18% to 1% over a 25-year period due to improvements in surgical technique, anesthesia, and postoperative management (16).

Surgical repair of AAA is indicated for aneurysms >5 cm in diameter without major contraindications. Following aneurysmal size with serial ultrasound every 3 to 6 months is indicated for those with a diameter of 3 to 4 cm. AAAs with diameters between 4 and 5 cm can be either followed or repaired. Surgical repair is also indicated for those with a growth rate of >5 mm over a 6-month period (7) and for AAAs that are symptomatic or complicated (11). A complicated AAA is defined as one with associated thrombus, adjacent organ ischemia (e.g., ischemic bowel), or peripheral emboli. Patients must be taken directly to the operating room for emergent repair if they present with the complete clinical triad or have a known AAA and associated abdominal pain or hypotension (7).

Appropriate preoperative evaluation is important in elective repair. There is a 5% to 10% risk of perioperative acute myocardial infarction for patients with uncorrected coronary artery disease (9). Preoperative cardiac evaluation usually includes a cardiac stress test or cardiac catheterization. Other important aspects of the preoperative evaluation include pulmonary, hepatic, renal, and hematological work-ups.

The standard operative management is prosthetic graft replacement with adventitial rewrapping (6). A new approach, endovascular stent placement, is currently in a clinical research stage. Common intraoperative complications include atheroembolism, declamping hypotension, acute renal failure (most commonly acute tubular necrosis), ureteral injury, and hemorrhage. Important postoperative complications include acute myocardial infarction, congestive heart failure, colonic ischemia, aortoenteric fistula, aortocaval fistula, graft infection, and anterior spinal syndrome.

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