Really bad pizza or something serious?

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Wilard's presentation

- A 63-year-old male presents to the ED via ambulance
- He was alone at his cottage eating pizza when he experienced sudden epigastric pain and back pain. He was incontinent of stool and lost the use of his legs. He lay on the floor until it improved and then drove to the local service station to call 9-1-1
- At the scene, he still complained of residual, bilateral leg weakness and left arm weakness. He noted pressure in his upper abdomen, radiating into his back
- His past medical history is significant for subarachnoid hemorrhage on two occasions. Both times, an aneurysm was identified and surgically clipped
- His medications include enteric coated acetylsalicylic acid, metoprolol and atorvastatin
- His vitals are as follows:
  - Temperature = 37.2 C
  - Heart rate = 42 bpm
  - Respiratory rate = 18 breaths per minute
  - BP = 96/60 mmHg
- The initial examination shows mild weakness in both legs and the left arm
- After transfer to a tertiary care center for further investigation, his physical examination is normal. His chest X-ray is shown in Figure 1

Wilard's treatment

- an ultrasound of the aorta revealed a suspected "flap." Subsequent CT scans showed an extensive aortic dissection from the aortic arch down into the iliac arteries (Figure 2)
- He was taken to the OR for emergency repair and was intubated for one week post-operatively, with copious lung secretions and delirium
- He was discharged home two weeks post-operatively

Questions & Answers

1. What is this patient's differential diagnosis?

The number of possible unifying diagnoses for this presentation are limited to:

- Aortic dissection (AD)
- Cerebrovascular event
- Spinovascular event
- Intoxication and pancreatitis
- Psychogenic event

A viable central nervous system event is almost unfathomable as it would need to involve both hemispheres in order to affect both legs. The one differential diagnosis that accounts for the whole story is AD.

2. What is the pathophysiology of AD?

The key pathophysiologic feature of AD is medial degeneration. Patients with hypertension have increased hemodynamic forces that accelerate medial degeneration. This is similarly true for those with bicuspid aortic valves. Patients with connective tissue diseases, like Marfan's and Ehlers-Danlos syndromes, have an intrinsically weaker aortic wall.

An intimal tear allows blood to gain access to the media and dissects the aorta along that plane. AD often begins at the aortic arch, aortic root or near the take-off of the subclavian artery. The overall result may be the formation of a true and false lumen, dissection through the adventitia (rupture), dissection into the coronaries or pericardium, or aortic insufficiency. Type A dissections involve the proximal aorta and Type B dissections are restricted to the distal aorta (Stanford classification).

3. What are the clinical features of AD?

According to the International Registry of Aortic Dissection (IRAD), so-called classic findings of AD-like pulse deficits and the murmur of aortic insufficiency are not common, occurring in only 15% and 32% of patients, respectively. Chest pain is a frequent complaint, but back pain also occurs in more than half of patients. The pain is usually described as abrupt in onset and severe. Other descriptors include "tearing" (51%), "sharp" (64%) and radiating (28%). A history of hypertension is common and consistent with the pathophysiology of dissection and half of patients have a systolic BP higher than 150 mmHg at the time of presentation.

Type A dissections have a higher mortality than type Bs. Without surgical repair, mortality is 20% on day one and 50% by one month. With surgery, mortality improves to 10% on day one and 20% by one month.

4. What investigations are useful in diagnosing AD?

History and physical examination should raise the index of suspicion, but is of poor diagnostic quality in itself. While AD has some well-described chest X-ray

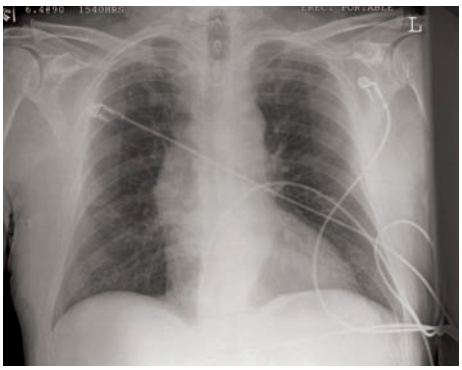

Figure 1. Chest X-ray showing a widened mediastinum.

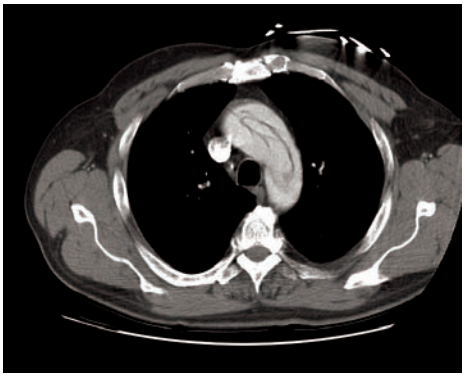

Figure 2. CT scan of Wilard's chest, showing an intimal flap in the aortic arch.

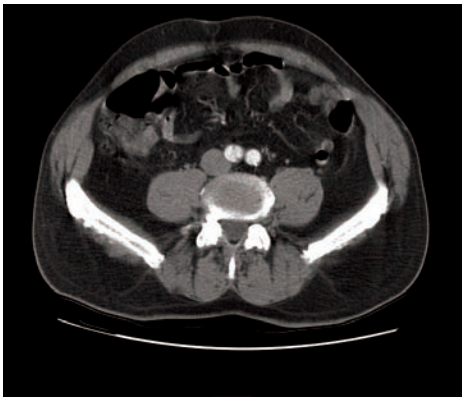

Figure 3. CT scan of Wilard's abdomen, showing the dissection flap extending into the right iliac artery.

findings, the sensitivity is poor. Notably, in IRAD, 12% of patients had an entirely normal chest X-ray. A widened mediastinum is defined as one that exceeds a quarter of the width of the chest and is noted in less than two-thirds of patients. ECGs are normal in about a third of patients, with nonspecific ST-T wave changes occurring in 41%. A minority of patients have an ischemic ECG (15%), presumably due to dissection into the coronaries or other mismatch of myocardial oxygen supply and demand.

Transesophageal echocardiography (TEE) is a very specific test, with sensitivities that improve with increased operator skill. Its major benefit is the fact that it is a bedside test, making it the fist choice in the hemodynamically unstable patient. However, the aortic arch remains in a blind spot of sorts because of the interposed trachea/left bronchus.

Aortography has been considered the gold standard test in the past, but the sensitivity and specificity are not as high as initially thought and it does have a small mortality rate (0.3%). The test is also long in duration and involves use of contrast.

Helical CT scan has a sensitivity and specificity approaching 100%. While the scan itself is fast, it is still measurably unsafe in an unstable patient and again involves the use of contrast.

MRI, on the other hand, also has excellent sensitivity and specificity for the detection of aortic dissection and does not require contrast. The test can be very difficult to obtain in most institutions and still involves a long trip out of the ED.

So, there are a number of diagnostic options. A decision analysis was published in the *Annals of Emergency Medicine* in 1996 by Sarasin *et al*. The bottom line is that low probability patients should be investigated with CT, MRI, TEE, or aortography and a negative result is sufficient to rule out AD. Low probability (< 15%) patients were those who had "severe, acute chest pain with no other suggestive features." A single negative test necessitates a second diagnostic test if the probability of disease is > 15%, keeping in mind the fact that time delays may affect mortality.

5. *How is AD managed?*

Analgesia is important to decrease sympathetic tone. Sodium nitroprusside should be used to control BP, maintaining systolic BP at 100 mmHg to 120 mmHg. A ß-blocker should be used in conjunction with this, as vasodilators may increase the heart rate and shearing forces on the aorta. Esmolol is very short-acting and readily titrated. Lebetalol has mixed α and ß activity and can be used alone (without sodium nitroprusside).

Patients with Type A AD benefit from prompt surgical intervention; whereas, those with Type B dissection are often managed medically. Dx