An Unusual Presentation of Penetrating Aortic Ulcers, A Case Report

Samantha Ward  
*Henry Ford Health System*

Abby Waterfield  
*Henry Ford Health System*

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Penetrating aortic ulcers (PAUs) were recognized as a separate disease process from aortic dissection in 1986 and affects 0.6 - 2.1 in 100,000 people per year [1]. It is estimated that 1 in 8 patients with penetrating aortic ulcers will progress to aortic dissections [2]. Patients with PAUs often present with severe chest pain, severe back pain, shortness of breath and unstable vital signs, often mimicking aortic dissection or pulmonary embolism. This case report describes an unusual presentation of PAU and highlights the importance of maintaining a high index of suspicion for aortic syndromes.

Case Presentation

The patient, a 74-year-old female, with past medical history of anxiety, depression, hyperlipidemia and hypertension, presented to the Emergency Department due to a burning pain on her left shoulder. The pain started three days prior and did not radiate. Nothing made the pain better or worse. She had tried Motrin and Tylenol which did not help. She denied trauma to the area. It was not associated with fevers, chest pain, shortness of breath, weakness or rash. As a side note, the patient noted that she was seeing a therapist outpatient for anxiety. She stated that “my family and doctors think I’m crazy and this is all in my head”. She admitted to weaning herself off of her psychiatric medications two weeks prior. She stated that she was stressed and anxious about the shoulder pain which, she felt, was making her symptoms worse. The patient denied a history of blood clots, smoking, recent surgery, and a family history of heart disease.

On exam, the patient was bradycardic (HR 59), normotensive (136/74) and afebrile. She was not tachypnic or hypoxic. The lungs were clear to auscultation and the chest was nontender. His heart rate was bradycardic, but in a regular rhythm without murmur, rub or gallop. Her abdomen was soft, nontender and no hepatosplenomegaly was appreciated. The patient was neurologically intact. She did not have edema or tenderness of the calves. She had full range of motion of her left shoulder and no tenderness to palpation.

A cardiac workup was then performed. Her EKG showed sinus bradycardia without ST elevations or T-wave inversions. Her lab work including CBC, BMP, and Troponin, revealed no major abnormalities. Chest X-ray was within normal limits without infiltrate or widened mediastinum [Image 1].

Given the normal physical exam, stable vital signs and benign laboratory results, it was thought that the patient may have been presenting with symptoms of a shingles rash that had not yet erupted or nerve pain.

At this time, the patient was re-evaluated. Her vitals were stable and her exam remained unchanged. The patient also noted that her pain had improved with pain medication. However, in the course of our discussion, the patient stated that she “could not get comfortable”. This symptom did not seem to fit with our initial diagnosis of shingles or nerve pain. The decision was then made hold the patient’s discharge and to obtain a CTA of her chest to rule out aortic dissection. No aortic dissection or aneurysm was seen. However, the patient was found to have numerous penetrating aortic ulcers along the aortic arch [Image 2]. They ranged in size from small to large. The patient was also found to have two, small bilateral pulmonary embolisms.

Cardiothoracic surgery was immediately consulted. The patient was then placed on an Esmolol drip with a systolic blood pressure goal of less than 110 and heart rate in the 60s. The patient was also placed on a heparin drip due to the pulmonary embolisms. Ultimately, the patient was admitted to the ICU for further monitoring. After being monitored for five days, with repeat imaging showing stable ulcerations, the patient was discharged home on Eliquis and anti-hypertensive medications. She did not require emergent surgical intervention and continues to follow regularly with cardiothoracic surgery.

Imaging and Diagrams

Figure 1: CXR obtained in ED showing no acute process

Figure 2: CTA obtained in ED showing multiple penetrating aortic ulcers in the aortic arch

Introduction

Penetrating aortic ulcer (PAU) is an ulcerative aortic disease involving the media and lead to an intramural hematoma. PAU is the least common disease process of aortic syndromes, accounting for about 2-7% [3]. The majority, 85-95%, of PAUs are found in the descending aorta [3].

Patients who present with PAUs, typically present with signs and symptoms similar to an aortic dissection including, severe chest pain, severe back pain, shortness of breath and unstable vital signs. Risk factors for PAUs are the same risk factors as you would see with atherosclerotic disease. It is believed that PAUs are associated with atherosclerosis of the aorta.

While PAUs are poorly understood, even less is known about how to treat them. There is debate in the literature, particularly among patients with PAUs who are asymptomatic, whether all patients with PAUs require surgery, which type of surgery is best and how emergently these patients should be taken to the OR. However, while in the ED, the treatment remains the same across the spectrum of disease. The mainstay of treatment is blood pressure and heart rate control. Typically, medications such as Esmolol and Nitroprusside are used. Beta blockers, like esmolol, have been shown to decrease mortality [4]. Alternatively, calcium channel blockers may be used. In addition to medical interventions, these patients should be monitored closely with repeated imaging in an ICU setting as they can be rapidly progressing and fatal.

Case Presentation Continued

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Discussion

Acute aortic syndrome is a term for a spectrum of diseases involving the aorta that present with similar symptoms. These diseases include the lethal triad of aortic dissection, intramural hematoma and penetrating aortic ulcer in descending order of severity. They may coexist in the same patient and evolve from one end of the spectrum to the next. It is estimated that 1 in 8 patients with penetrating aortic ulcers will progress to aortic dissections [2]. However, only 5% of patients with aortic dissection, are thought to have had a preceding aortic ulcer.

Penetrating aortic ulcers (PAUs) are ulcerative lesions that erode the internal elastic lamina of the aorta. Eventually, that erosion may progress into the media and lead to an intramural hematoma. PAU is the least common disease process of aortic syndromes, accounting for about 2-7% [3]. The majority, 85-95%, of PAUs are found in the descending aorta [3].

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While PAUs are poorly understood, even less is known about how to treat them. There is debate in the literature, particularly among patients with PAUs who are asymptomatic, whether all patients with PAUs require surgery, which type of surgery is best and how emergently these patients should be taken to the OR. However, while in the ED, the treatment remains the same across the spectrum of disease. The mainstay of treatment is blood pressure and heart rate control. Typically, medications such as Esmolol and Nitroprusside are used. Beta blockers, like esmolol, have been shown to decrease mortality [4]. Alternatively, calcium channel blockers may be used. In addition to medical interventions, these patients should be monitored closely with repeated imaging in an ICU setting as they can be rapidly progressing and fatal.

Conclusion

This case demonstrates that a seemingly benign and common symptom, such as shoulder pain, can ultimately be the only presenting sign of a life-threatening diagnosis. All physicians must maintain a high degree of suspicion, particularly in the ED setting. This case also demonstrates the dangers of anchoring bias. Had we anchored ourselves on the shingles diagnosis and discharged the patient upon a normal reassessment, we would have missed two life threatening diseases.

References