An uncommon cause of chest pain – penetrating atherosclerotic aortic ulcer

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Chest pain is a very common symptom and can be of cardiac or non-cardiac origin. It accounts for approximately 5.5 million annual emergency room visits in the United States, according to 2011 CDC data. Penetrating atherosclerotic aortic ulcer (PAU), an uncommon condition, is also a potential cause of chest pain. We here report the case of a 65-year-old woman who presented with atypical chest and back pain. The pain persisted for 4 weeks necessitating two emergency room visits. Initial tests were non-significant including cardiac troponins, an electrocardiogram (EKG), and a chest X-ray on her first visit. Upon her second visit, she underwent a computed tomography angiogram of chest with contrast which revealed a PAU with an intramural hematoma in descending aorta. The PAU was finally diagnosed with an exclusion of other chest pain causes. She was treated non-surgically with a blood pressure control strategy and pain management. After a 2-month period of smoking cessation and following the achievement of a controlled blood pressure, she felt well without chest pain.

Keywords: chest pain; ischemic heart disease; penetrating atherosclerotic aortic ulcers; hypertension; smoking and tobacco

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Penetrating atherosclerotic aortic ulcer (PAU) can be defined as ‘an ulceration of atherosclerotic plaque that penetrates the intima then progresses into tunica media layer with or without associated pseudoaneurysm or intramural hematoma (IMH)’ (1, 2). PAU was initially reported by Shennan Schumacher in 1934 then followed by Stanson et al. in 1986 (3, 4), but it was not a common disease until 1980 when advances were made in the non-invasive imaging era such as computed tomography (CT) and magnetic resonance imaging (MRI). It presents mostly with chest pain or chronic back pain. Due to its rarity and vague clinical presentation, it is an uncommon differential diagnosis for chest pain which often results as a ‘late diagnosis’. We report this case as a reminder that PAU may present with a very common symptom (chest pain) and clinicians should be aware of uncommon chest pain etiologies.

Case presentation

A 66-year-old black woman presented with chest pain for 6 days. Three weeks prior to this presentation, she was evaluated in the emergency room for similar complaint and was discharged after having normal blood and imaging tests including cardiac enzymes, electrocardiogram (EKG), and chest X-ray. Her chest pain was described as sharp, constant, 6/10 in intensity, located in the substernal area, radiating to the back, no exacerbating factors, and improved by non-steroidal anti-inflammatory drugs (NSAIDs). She denied any shortness of breath, cough, or hemoptyis. She had no previous history of rash, prior shingles, or lower back pain. She had a past medical history of hypertension and was taking amlodipine and metoprolol at home. Her family history included hypertension in both parents. She was a chronic smoker, smoking an average of five cigarettes a day for more than 20 years, but denied any alcohol or illicit drug use. Her vital signs were significant for mildly elevated blood pressure of 160/98 mmHg with a pulse rate of 75/min.

On examination, she had normal pulses, a normal heart rate, and rhythm without any murmurs or gallops. There was no chest wall tenderness, abdominal discomfort or costovertebral angle tenderness, and pedal edema. Initial EKG showed normal sinus rhythm at 86 bpm, normal axis with no evidence of ST elevation or depression (Fig. 1). A chest X-ray was unremarkable. Routine blood tests were done, which showed normal values of complete blood count, basic metabolic panel, liver function tests,
TSH, coagulation profile, lipid profile, and serum cardiac troponin I (cTnI). Other labs revealed ESR 25 mm/h (normal 0–20 mm/h), CRP 6 mg/L (normal <5 mg/L), and D-dimer 1.51 mg/L (normal <0.5 mg/L).

A list of differential diagnoses including acute myocardial infarction, acute pulmonary embolism, aortic dissection, myocarditis, pericarditis, and musculoskeletal pain was made. Acute myocardial infarction and pericarditis were ruled out with the presence of normal EKG, normal cTnI, ESR, and high normal CRP. Myocarditis was less likely with the findings of serology tests. CT angiogram of the chest with contrast (CTA) was obtained, which showed a protrusion of the medial wall of the descending thoracic aorta into a thickened aortic wall indicative of a penetrating atherosclerotic ulcer with IMH (Fig. 2a, b). Transthoracic echocardiogram (TTE) showed normal left ventricular size, wall thickness, and systolic function (ejection fraction – 59.85%). Exercise stress test did not reveal any signs of myocardial ischemia. With the normal findings of cardiac workup and CT angiogram, PAU with IMH was diagnosed as a cause of recurrent chest pain.

Her blood pressure was high around 150–160/90–100 mmHg at the time of admission. She was treated with oral labetalol for hypertension and intravenous

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**Fig. 1.** ECG demonstrating normal sinus rhythm at 86 bpm. No other significant abnormalities were noted.

**Fig. 2.** CT angiogram of chest with contrast (a) transaxial and (b) coronal planes demonstrating a penetrating atherosclerotic aortic ulcer (blue arrow) with a mural hematoma approximately 2.3 cm in the largest transverse diameter and 1.25 cm in the largest thickness (blue arrow).
morphine for pain control. She was symptomatically improved within a few days of hospitalization. As her condition did not require immediate surgical intervention, she was scheduled for outpatient follow-up and was counseled about medication compliance for anti-hypertensive and smoking cessation. After 2 weeks, she followed up in the clinic where she stated that her chest and back pain had resolved. Her blood pressure was controlled with BP of 130/80 mmHg and also she had successfully quit smoking.

**Discussion**

Acute aortic syndrome (AAS) is a constellation of aortic dissection, IMH, and penetrating atherosclerotic disease. With recent developments in diagnostic modalities, PAU has been frequently identified over the past few years with 2.3–7.6% estimated prevalence in symptomatic patients with AAS, but its background and actual incidence remains unknown (5, 6). PAU usually develops in old age, atherosclerotic disease, coronary artery disease, hypertension, hyperlipidemia, a strong smoking history, and renal disease (7, 8). Chest and back pain are the most common symptoms of PAU. It cannot be differentiated from aortic dissection by symptoms alone at initial emergency room visit, where exclusion of classic aortic dissection is crucial (5, 8). PAU can be found in the ascending and descending thoracic aorta while the latter was being more common.

PAU, known as a disease of the intima, can start as a benign atherosclerotic plaque in the aorta, which can gradually progress to deep media layer associated with IMH due to disruption of vasa vasoform that may be a nidus of aortic dissection (9). It can be associated with complications such as acute aortic dissection, aortic rupture, aortogastric fistula, and false aneurysm (4, 10, 11). Moreover, the risk of aortic rupture was higher in patients with PAU (40%) compared with patients with type-A (7%) and type-B dissections (3.6%) (9). It is worth noting that the understanding of PAU in the literature came from imaging studies rather than histopathologic findings (8, 9). Because of its vague presentation, it is often challenging to diagnose PAU in the first medical encounter. Therefore, PAU is often found while the patient is undergoing diagnostic tests to rule out other pathologies.

Traditionally, PAU was diagnosed by aortography; however, since the early 1980s, CTA, MRI, and TTE have replaced aortography to diagnose aortic pathology. Table 1 summarizes the radiologic findings of PAU (6, 8, 12, 13). However, CTA is the best diagnostic choice in PAU (8). Management of PAU depends on several factors including the presence of symptoms, location of ulcer, extent and size of the ulcer, and presence of IMH (8, 14). The presence of PAU in ascending aorta with persistent symptoms, hemodynamic instability, IMH, and progression of ulcer size are major predictors of adverse outcome which require early surgical intervention (14).

We here report the case of a 65-year-old woman with recurrent chest and back pain of unknown origin, later found to have PAU with IMH in the descending aorta as a cause of her chest pain. There was a similar case reported in which a patient presenting with persistent chest pain was determined to have a PAU in the descending aorta. CT showed varying degrees of sub-adventitial hemorrhage in the thoracic aorta; therefore, this patient underwent surgery successfully (15). Singhal et al. reported a case of PAU, which presented with chest and back pain of uncertain origin. EKG and cTnI findings were normal. CTA chest showed a PAU in the ascending aorta associated with aneurysmal dilatation, which required immediate surgical intervention (16). The above cases highlight the importance of early detection and surgical treatment of PAU to prevent adverse outcomes if CT scan findings were of concern.

Interdisciplinary expert consensus from the *European Journal of Cardio-Thoracic Surgery* made the statement that medical treatment was indicated in patients with uncomplicated courses whereas thoracic endovascular aortic repair (TEVAR), or surgical therapy, was considered in complicated cases including persistently symptomatic patients despite medical treatment, asymptomatic patients with large pleural effusions, IMH, and large initial PAU depth (>10 mm) and diameter (>20 mm) (14). In asymptomatic patients who do not require surgical intervention, follow-up is recommended every 6 months for the first 3 years and every year thereafter with imaging techniques (14). Our patient did not have other aortic complications throughout the hospital stay. Since there was no surgical indication for our patient, she was conservatively managed with tight blood pressure control and smoking cessation. She was symptomatically better and discharged to outpatient follow-up with further imaging studies.

In conclusion, PAU is an underestimated aortic disease that can present with chest and back pain. Until now, there is no sufficient data in the literature to suggest which chest pain patient should undergo an extensive workup. Individualized clinical judgment with a detailed history and

**Table 1. Radiologic findings of PAU**

| Imaging                  | Findings                                                                 |
|-------------------------|--------------------------------------------------------------------------|
| CT angiogram with IV contrast | Localized, crater-like, contrast-filled out-pouching of the aorta through intima layer |
| MRI                     | High intensity in the aortic wall in both T1- and T2-weighted images (useful in determination of hematoma age) |
| TEE                     | Crater-like or focal out-pouching with rough edges in an atherosclerotic aortic wall |
thorough physical examination is a valuable tool in the evaluation of chest pain, and it is paramount to think of PAU if chronic chest and back pain in elderly patients are unrelated to cardiac or musculoskeletal origin, where further diagnostic imaging might be necessary. This case presents an essential message to all physicians. We mostly focus on the assessment of acute coronary syndrome and pulmonary pathologies in patients with chest pain but unfortunately we forget to consider other etiologies that may be equally dangerous. Since PAU is one of the AAS with a high risk of complications, we may miss this life-threatening diagnosis if we do not have a hunch and perform appropriate tests.

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