

Case Report

A Blind Spot in the Eye of Imaging Technology: Penetrating Atheromatous Ulcer

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Acute aortic syndrome (AAS) is a term that is used to describe a similar clinical profile that may have different underlying pathophysiological mechanisms. It includes classic aortic dissection, intramural aortic hematoma, and penetrating atherosclerotic ulcer. We describe the case of a 77-year-old female who presented with syncope of unknown duration. The chest X-ray was suggestive of a widened mediastinum. The initial work-up with a computed tomography scan and transesophageal echocardiogram failed to diagnose a penetrating atherosclerotic ulcer. We discuss the importance of a high degree of clinical suspicion for AAS and the utility of different imaging technologies in making the diagnosis.

Acute aortic syndrome (AAS), is a term that is used to describe a similar clinical profile that may have different underlying pathophysiological mechanisms.¹ It includes classic aortic dissection, intramural aortic hematoma, and penetrating atherosclerotic ulcer; and, more recently, incomplete dissection. There is a possibility of an underlying link, considering that these conditions coexist and frequently precede each other.² Moreover, the likelihood of a downward clinical course is high, making early diagnosis and management of paramount importance.

Case presentation

We present the case of a 77-year-old female of Japanese origin, with a past medical history significant only for essential hypertension, who presented with syncope of unknown duration in the field. Her presenting vitals in the emergency department were: blood pressure 100/80 mmHg, heart rate 120 beats per minute, respiratory rate 32 per minute, temperature 38°C.

At presentation she was alert, awake and oriented, but appeared tachypneic. The physical examination was unremarkable. Her initial chest X-ray (CXR) showed left upper lobe infiltrate, and evidence of left ventricular hypertrophy was present on the electrocardiogram (Figures 1-2). She was treated empirically with vancomycin and cefepime, along with 2.5 L normal saline.

During her stay in the emergency department, her blood pressure rose to 218/117 mmHg and she received intravenous labetalol 55 mg total (15 mg + 20 mg + 20 mg). On hospital day 2, the previous CXR, although rotated, was suggestive of a widened mediastinum. A computed tomography (CT) scan of the chest with contrast was performed. This study revealed an intramural hematoma within the ascending aorta, extending to the proximal anterior arch, with hemopericardium anteriorly and inferiorly, without any evidence of intimal flap, dissection, or ulcerated plaque (Figure 3). Transesophageal echocardiography (TEE) was also performed, and failed to show evidence of

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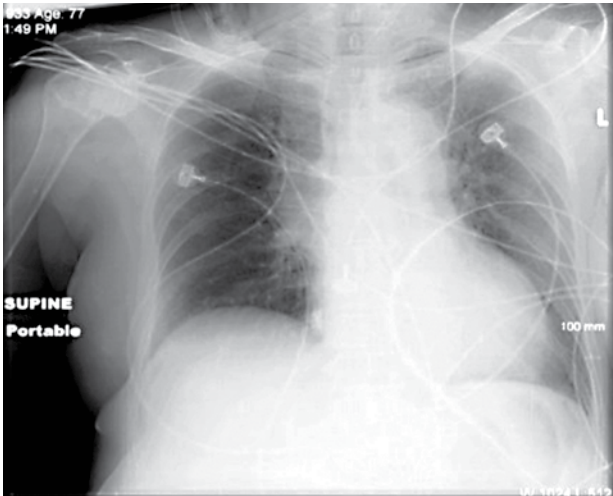


Figure 1. Chest X-ray on admission.

any intramural hematoma or aortic dissection in the ascending aorta, thoracic aorta, or descending aorta. The patient was transferred to the coronary care unit for medical management.

On hospital day 3, a decision was made to operate. Intraoperative findings included a penetrating atherosclerotic ulcer (PAU) along the inferior portion of the arch, which was a full-thickness ulceration

that appeared to bleed and had some fresh thrombus present at the base; a small intramural hematoma of the ascending aorta, extending down to the aortic root and up to the base of the innominate and carotid arteries; and 150 ml of hemopericardium, mostly inferiorly and laterally. Replacement of the ascending aorta and inferior arch was done using a #28 intervascular Dacron graft under circulatory arrest. The patient tolerated the surgery well with no complications. She continued to do well postoperatively.

Discussion

The term AAS, as described above, was first given by Vilacosta et al and associates around 1998 to bring under one roof three different pathophysiological mechanisms presenting in a similar clinical manner.^{1,3} Clinically and classically, the description of “aortic pain” as severely intense, acute, tearing or ripping, pulsating and migratory chest pain may suggest that the patient has an underlying AAS.⁴ However, according to the International Registry of Acute Aortic Dissection (IRAD), in contrast to the “aortic pain” mentioned above, the majority of people described a sudden onset of severe sharp pain. Moreover, involvement of the ascending aorta is suggested by pain

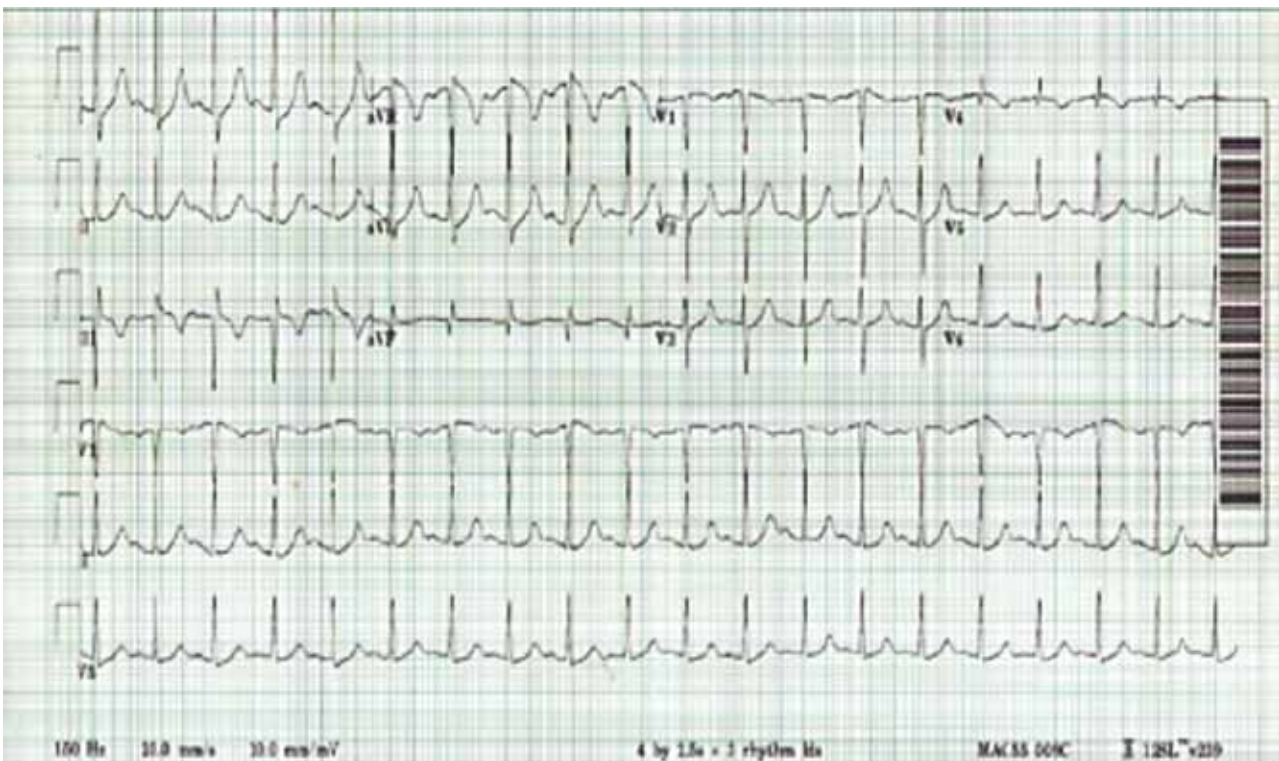


Figure 2. Electrocardiogram on admission.

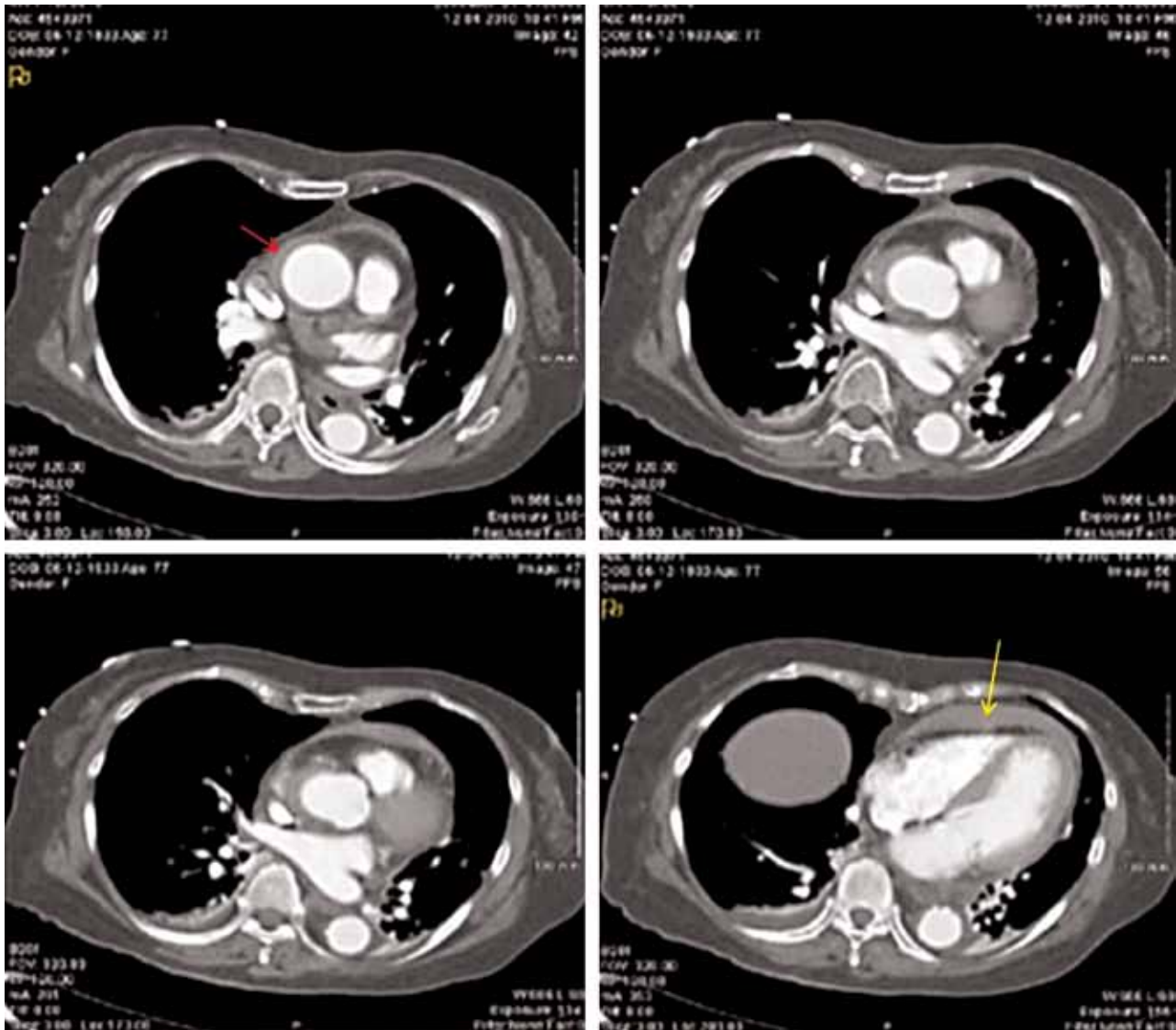


Figure 3. Computed tomography of the chest with contrast, showing intramural hematoma of the ascending aorta (red arrow) and hemo-pericardium (yellow arrow).

radiating to the neck, jaw or throat, whereas pain radiating to the back or abdomen suggests involvement of the descending aorta.⁴ Notably, the initial presentation is syncope in only 13% of proximal and 4% of distal cases, according to IRAD, indicating that a high degree of suspicion of AAS in the setting of syncope is strongly recommended.

In the case reported here, a patient who initially presented with syncope was eventually found to have PAU. PAU was first described by Shennan et al in 1934, however it was Stanson et al in 1986 who showed its presence as a separate clinical and pathological entity.⁵ PAU is a lesion that penetrates the internal elastic lamina through the media.^{1,5} Data from

specialized centers and IRAD have shown that its incidence ranges from 2.3–11%. PAU usually tends to occur in elderly men with hypertension, tobacco use, and coronary artery disease, as reported in the Mayo Clinic Series,⁶ and is usually located in the descending aorta,⁶⁻⁸ whereas PAU in the ascending aorta, as in our case, is extremely rare. In one review article, only 6 of 134 patients with PAU had a location in the ascending aorta.⁷ Differentiating PAU from the other AAS entities, as well as from acute pulmonary embolism (PE) and acute myocardial infarction (AMI) is of paramount importance. To differentiate the above conditions on the basis of history, aided by physical, electrocardiographic, and cardiac biomarkers, of-

ten poses a diagnostic challenge. Recent studies have proposed D-dimers as a screening tool to help triage AAS/PE and AMI.⁹

This leads us to a consideration of the role of imaging in the diagnosis of PAU. The modalities available are CXR, aortography, CT scan, magnetic resonance imaging and TEE. CXR has a very limited role, with 64% sensitivity for AAS, which drops to 47% if the pathology is located in the ascending aorta.¹⁰ Despite these limitations, the clue on the CXR in our case was still an aid to our diagnosis. CT scan is the best test for diagnosing PAU, which appears as a focal, contrast-filled outpouching, with jagged margins, and extending beyond the expected aortic wall boundaries, usually with severe underlying atheromatous disease.¹¹⁻¹³ The average sensitivity exceeds 95% and specificity ranges from 87-100%.^{14,15} TEE has been shown to reach a sensitivity of 99% and a specificity of 89%, with a positive and negative predictive values of 89% and 99%, respectively. However, in the present case the chest CT scan and TEE both failed to show the PAU in the ascending aorta. The CT scan was able to demonstrate the intramural hematoma in the ascending aorta, but the TEE, read by two experienced echocardiographers, failed to show any aortic pathology.¹⁶

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