CASE REPORT

Pleuritic chest pain and fever: An unusual presentation of aortic dissection

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Abstract

It remains a challenge to diagnose aortic dissection in primary care, as classic clinical features are not always present. This case describes an atypical presentation of aortic dissection, in which the patient walked in with pleuritic central chest pain associated with a fever and elevated C-reactive protein. Classic features of tearing pain, pulse differentials, and a widened mediastinum on chest X-ray were absent. This unusual presentation highlights the need for a heightened level of clinical suspicion for aortic dissection in the absence of classic features. The case is discussed with reference to the literature on the sensitivity and specificity of the classic signs and symptoms of aortic dissection. A combination of the aortic dissection detection risk score (ADD-RS) and D-dimer test is helpful in ruling out this frequently lethal condition.

Introduction

Aortic dissection (AD) is an uncommon but potentially fatal condition. In the undergraduate medical curriculum as well as the postgraduate primary care curriculum, this condition is frequently emphasised as a differential diagnosis to be considered in patients presenting with chest pain. Its classic clinical features include sudden onset, tearing upper back pain, pulse or blood pressure differentials between bilateral limbs, and a widened mediastinum on chest X-ray. However, recent literature indicates that patients more often describe the pain of aortic dissection (AD) as severe and sharp or knife-like (68%) rather than with the classic description of tearing or ripping pain (50%).[1,2] This case report describes an atypical presentation of aortic dissection in which a patient presented with sharp, pleuritic, central chest pain, which was associated with a fever and significant C-reactive protein (CRP) elevation.

Case

A 63-year old gentleman presented to our primary care clinic with a two-day history of sudden-onset central chest pain at rest which radiated up to his neck. It was sharp in nature and worse on inspiration. He felt feverish but had neither a cough nor dyspnoea. His pain had been constant, with a score of 7 out of 10. Paracetamol relieved his fever but not the pain. There was no haemoptysis, calf pain, history of immobility, or trauma to his chest. He was on perindopril for hypertension and atorvastatin for dyslipidaemia. He quit smoking seven years ago and did not use any recreational drugs.

Clinically, he looked well and was not tachypnoeic. His oral temperature was 38.3ºC, oxygen saturation was 97% on air, respiratory rate was 14 breaths per minute, blood pressure was 135/78 mmHg, and pulse was regular at 95 beats per minute, with no radio-radial delay. On auscultation, his heart sounds were normal. There were equal vesicular breath sounds in the bilateral lung fields. There was no calf swelling or tenderness bilaterally. Examination of other systems, including neurological examinations, were normal.

His electrocardiogram (ECG) did not show any abnormality. A chest X-ray was performed, and it showed no significant abnormality (**Figure 1**). The following blood tests were ordered: full blood count (FBC), C-reactive protein (CRP), renal profile (RP), liver function tests (LFT), and Troponin T (Trop T).

At this point in time, the differential diagnosis considered was early atypical pneumonia given the pleuritic nature of the pain and fever. An atypical presentation of acute coronary syndrome (ACS) was also considered in view of his cardiovascular risk factors.

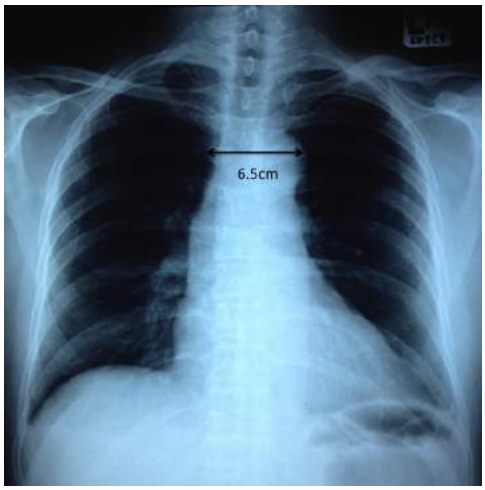

Figure 1. Chest radiograph (PA erect) is normal with no widened mediastinum (normal <7.5cm) or double aortic contour.

He was given paracetamol and kept for observation while waiting for his investigation results. An hour later, the laboratory investigations showed an elevated Trop T of 22 (Normal<14 ng/L), white cell count (WCC) of 13.11 x 10^3 (4.0-10.0 x 10^3/μL), neutrophilia of 10.37 x 10^3 (1.9-8 x 10^3/μL), and a CRP of 112.9 (<5.0 mg/L).

Upon discussion with the cardiologist on duty, the patient was admitted for observation with a plan to repeat an ECG and troponin in six hours. Oral antibiotics with atypical pneumonia coverage were given. Upon review, his symptoms settled, and the repeat ECG and troponin were normal. He was discharged with a course of oral antibiotics, a review appointment in three days, and appropriate safety netting.

He returned to the emergency department two days later with a similar persistent pain, lethargy, and feelings of feverishness. His temperature and other vital signs were normal, and there were right basal crackles on auscultation of his lungs. This time, his ECG (**Figure 2**) showed non-specific changes (widespread ST elevation on leads I, II, V2-V6; ST depression on leads aVR and V1). Repeat blood tests, including for troponin, and blood cultures were normal except for an elevated CRP (334 mg/L). A transthoracic echocardiogram was performed and showed mild global pericardial effusion with a left ventricular ejection fraction of 48%.

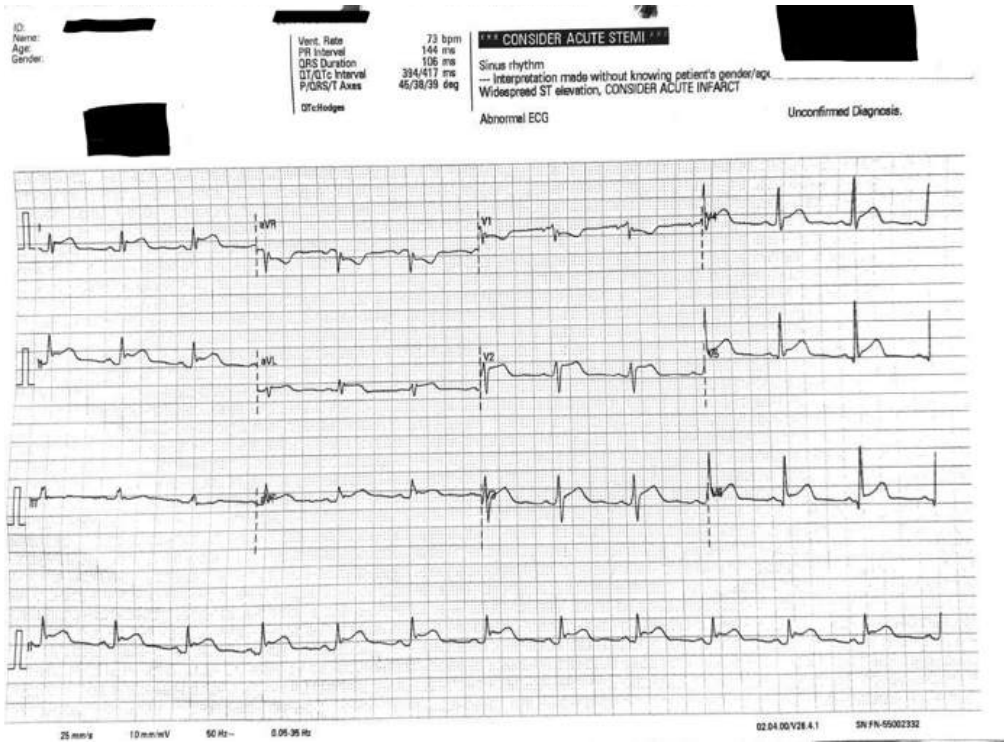

Figure 2. ECG on second presentation shows widespread ST elevation on leads I, II, V2-V6; ST depression on leads aVR and V1.

The working diagnosis at this point was community-acquired pneumonia with acute pericarditis. He was started on intravenous ceftriaxone and oral ibuprofen. His symptoms did not improve after two days of treatment; hence, a CT thorax was performed to investigate further.

The CT thorax showed a dilated ascending aorta with possible dissection. Thus, a CT aortogram was subsequently performed. This aortogram showed a Stanford A ascending AD (**Figure 3**). He was admitted to intensive care for strict pulse and blood pressure control.[3,4] The cardiothoracic surgical team repaired the ascending aorta surgically using an open approach, while the descending aorta was managed using a stent-graft.[5] The patient recovered well post-operatively and was discharged after five days. He was reviewed in the outpatient clinic after 6 months and has continued to do well.

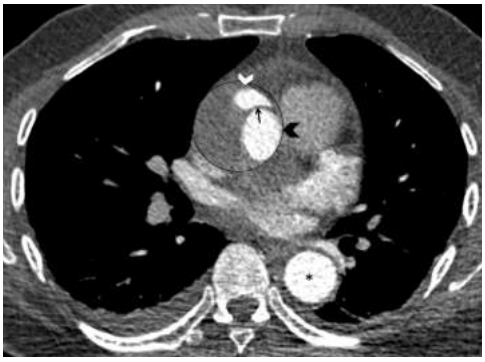

Figure 3. Axial CT aortogram. The ascending aorta (circled) is dilated and measures 4.6cm in diameter (normal 3.5cm). An intimal flap (black arrow) divides the aortic lumen into a true lumen (black arrowhead) and a false lumen which is partially thrombosed (white arrowhead). A normal descending thoracic aorta is seen posteriorly (*).

Discussion

This case demonstrated the diagnostic challenges often faced by frontline doctors when dealing with atypical chest pain. In all cases of chest pain, a bilateral pulse or blood pressure differential can be useful signs to rule in thoracic AD when they are present, with a specificity of 99% (95% CI= 96-100%).[6] However, with a low sensitivity of 38% (95% CI= 30-47%), these are less useful signs to rule out AD when they are absent.[6] Neurological deficit and hypotension are two other examination findings that have a high specificities of 95% (95% CIs of 93-97% and 93-96%, respectively).[6] However, both have very low sensitivities of 18% (95% CI= 11-30%) and 15% (95% CI= 10-23%), respectively.[6] The presence of any of these three features increases the likelihood of AD. However, there was no radio-radial delay, hypotension, or neurological deficit in this patient, and their absence could not exclude the diagnosis.

In this case, atypical pneumonia with acute pericarditis was considered to be the most likely diagnosis, as the patient presented with pleuritic chest pain, fever, and an elevated CRP level. A review of the literature shows that fever is a common accompanying feature for AD[7-10] and that it has been found in one-third of all patients with AD.[11] The characteristic fever in AD is distinct from that with an infective aetiology.[12] It has been reported that the fever, which is secondary to AD, is seen mostly in type B dissections and that it begins between 48 to 72 hours after the dissection occurs.[12] The variability in body temperature is significantly less when compared with a fever that is secondary to an infection.[12] This retrospective study also showed that in patients with a fever that is secondary to AD, microbiological investigations produced negative results.[12] Patients also showed no response to antibiotic therapy, and their fever subsided in 24 hours when treated with indomethacin.[12] In our patient, the blood culture was negative, and his fever resolved with ibuprofen. Elevated WCC[13,14] and CRP[13-16] have also been found to be associated with AD. CRP elevation is thought to be secondary to the systemic acute- phase reaction and the underlying vascular inflammation process.[16]

As he was not responding to treatment, another possible differential diagnosis that should have been considered at this stage was pulmonary embolism (PE). His calculated Wells score would have put him into the "PE unlikely" group, necessitating a D-dimer test as the next step in the investigation.[17] In this case, the D-dimer test was likely to have been positive. Therefore, referral for CT angiography would have been the next recommended course of action and would have shown the dissection.

Elevated D-dimer (≥500 ng/mL) has also been found to be highly sensitive (96.7% sensitivity, 95% CI= 93.6-98.6%) for AD[18] but is inadequate as a stand-alone test.[19] The European Society of Cardiology recommends utilizing the Aortic Dissection Detection Risk Score (ADD-RS) and performing the D-dimer test as a secondary filter when the ADD-RS score is ≤ 1.[3] The ADD-RS is a pre-test probability assessment tool that can help

with AD risk stratification.[4] It incorporates the presence of high risk conditions, pain, and examination features.[4] The score represents the total number of groups out of three (see **Figure 4** for group details) with the presence of any single risk factor. An ADD-RS score of > 1 necessitates referral for a CT Angiography.[18] A recent multicentre prospective study examined the diagnostic accuracy of utilizing the combination of the ADD-RS and the D-dimer test for detection of AD. This study concluded that an ADD-RS score ≤1 plus a negative D-dimer had a sensitivity of 98.8% (95% CI= 96.4-99.7%), negative predictive value of 99.7% (95% CI= 99.1-99.9%), and negative likelihood ratio of 0.02 (95% CI= 0.01-0.07).[18] Therefore, this combination is useful in ruling out AD (**Figure 4**). This new clinical strategy, which combines a pre-test probability assessment tool, ADD-RS, with the D-dimer test, supports doctors in terms of reducing misdiagnoses and over-testing for AD.[18] However, further validation studies in the primary care setting should be done before integrating this tool into clinical practice.

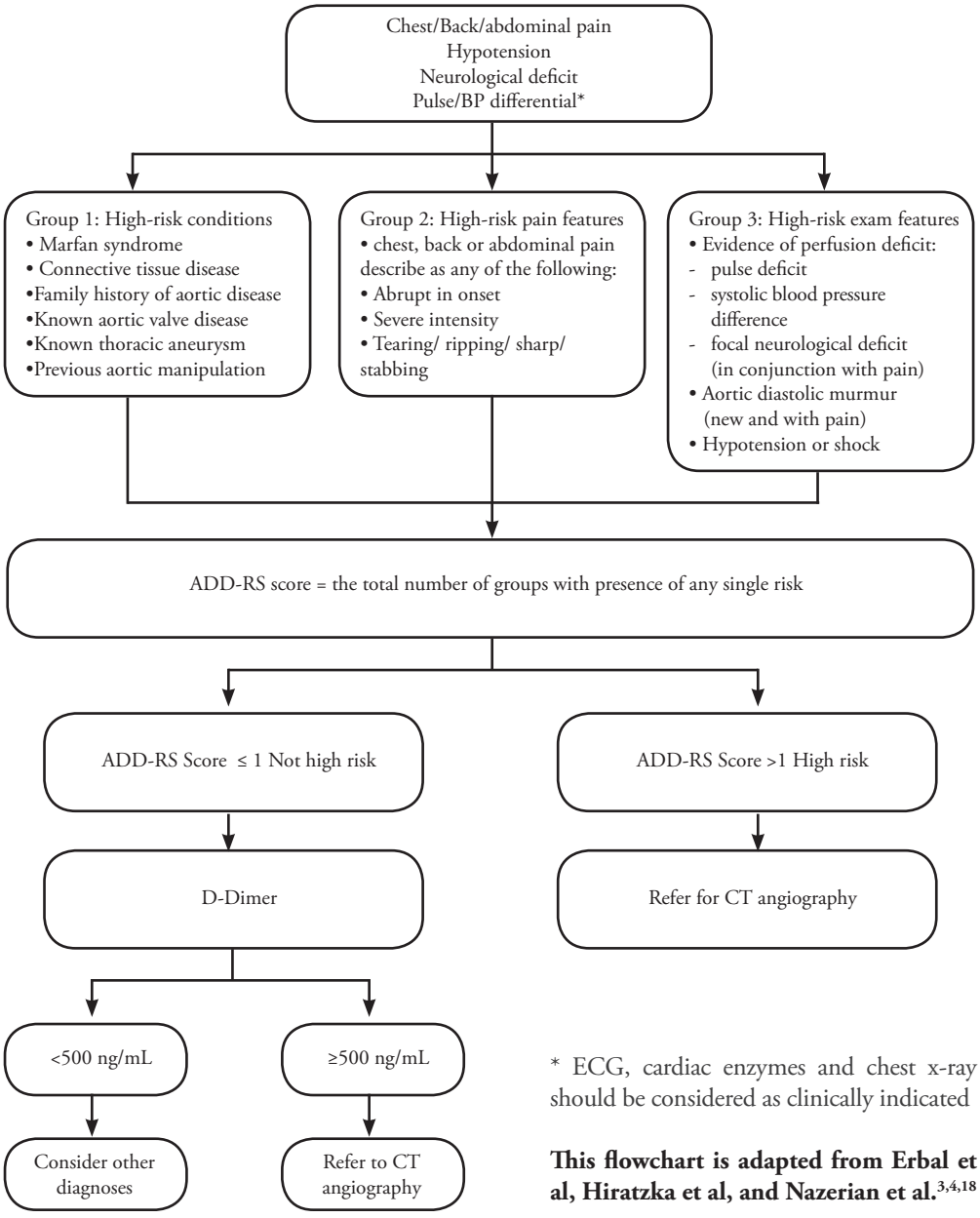

Figure 4: Flowchart for AD evaluation incorporating the ADD-RS and the D-dimer test.

Summary

AD should be considered in all patients with atypical chest pain. It may present with fever and elevated CRP. Pulse or blood pressure differentials, hypotension, and neurologic deficits can be helpful in terms of suggesting aortic dissection, but their absence cannot rule out the diagnosis. The ADD-RS is a useful risk stratification tool for AD, and the combination of the ADD-RS and the D-dimer test is helpful in ruling out AD. However, the D-dimer test is not widely available in the Malaysian primary care setting. Therefore, patients with an ADD-RS score ≤ 1 should be referred for D-dimer testing in the secondary care setting.

"We declare that there are no competing interests and that no funding was received for this case study.

Acknowledgement

We would like to acknowledge Prof. Dr. Mohd Zaki Salleh for his valuable input into this case report."

How does this paper make a difference to general practice?

- The unusual presentation highlights the need for a heightened level of clinical suspicion of AD in the absence of classic features.
- It highlights that:
  - fever and elevated CRP are possible associated features of AD.
  - the absence of classic features, such as pulse or blood pressure differentials, hypotension, and neurological deficits, does not rule out AD due the low sensitivity exhibited by these features.
  - the ADD-RS is a useful tool for AD risk stratification.
  - the combination of the ADD-RS and the D-dimer test is useful in ruling out AD.

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