

Dissection of the aorta, a rare combination of coarctation, a bicuspid aortic valve, and hypertension in a young patient

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Aortic dissection is an uncommon but very lethal disease. Majority of cases are found in elderly patients with a history of hypertension. Young patients usually have other risk factors such as vasculitis, Marfan syndrome, unrecognized coarctation of the aorta, and a bicuspid aortic valve. We present a case of a young patient who presented with epigastric pain mimicking peptic ulcer disease that was later on proved to be type B dissection of the aorta. The patient had a unique combination of hypertension, a bicuspid aortic valve, and postductal coarctation.

Keywords:

bicuspid aortic valve, coarctation of the aorta, hypertension, type B dissection

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Introduction

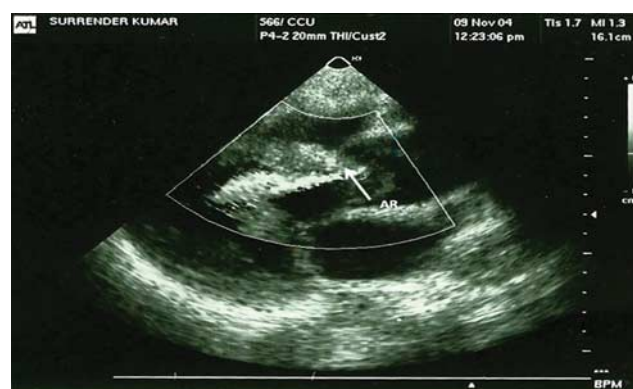
Aortic dissection is a relatively uncommon, although fatal condition that may result in adverse outcomes. Patients usually present with severe chest or abdominal pain and acute hemodynamic compromise. Presentation is highly variable and diagnosis is easily missed if suspicion is not very high. Prompt diagnosis and management is key to reducing morbidity and mortality. The primary event is a tear in the intima on the background of cystic median necrosis [1]. The most important risk factor is hypertension along with atherosclerosis, but in young patients, other factors may be more important such as vasculitis, rheumatoid arthritis, syphilitic aortitis, Marfan syndrome, Ehler–Danlos syndrome, preexisting aneurysm, coarctation of the aorta, and a bicuspid aortic valve [2]. Descending abdominal aortic dissections are usually managed successfully medically, although recently, there has been a trend toward endovascular stent management.

A 47-year-old man, smoker, known to have bronchial asthma, hypertension, and peptic ulcer disease was admitted with a history of upper abdominal pain, mainly epigastric, associated with vomiting. Pain was moderate in intensity and sometimes radiating to both flanks, and was continuous for the last 3–4 h, with no chest pain or shortness of breath. On examination, the patient was conscious and alert; his blood pressure was 160/100 and heart rate was 100/min, with no discrepancies in pulses. There was mild epigastric tenderness, mild bilateral expiratory wheeze, and short systolic murmur along the left sternal edge.

Initially, a provisional diagnosis of peptic ulcer disease versus acute coronary syndrome was made and he was started on treatment. Serial ECGs and cardiac markers were normal. The patient continued to have epigastric pain. Repeated ECG showed new changes in inferior leads. An echocardiogram indicated a bicuspid aortic valve with significant aortic regurgitation (Fig. 1). On the fourth day of admission, the patient developed severe

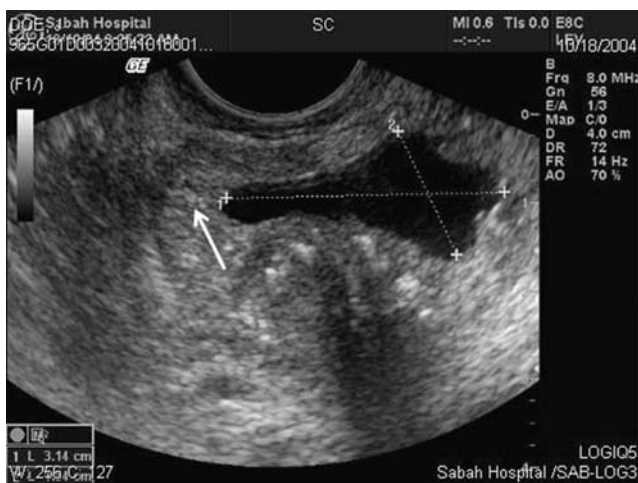
epigastric pain, with repeated vomiting associated with abdominal guarding and rigidity. A diagnosis of acute pancreatitis versus a perforated ulcer was considered differential diagnosis of acute pancreatitis versus a perforated peptic ulcer was considered and serum amylase, radiograph, and ultrasonography of abdomen were carried out. Amylase and radiograph were normal, but ultrasonography showed a large thrombus in the aorta at the level of the second lumbar vertebra (Fig. 2). Acute dissection of the aorta was suspected and an urgent computed tomography (CT) angiogram was carried out, which showed postductal coarctation with an aberrant right subclavian artery, Stanford-B dissection of the aorta extending into common iliac arteries, and a thrombosed false lumen with patent vital abdominal arteries (Figs 3 and 4). The patient was seen by a vascular surgeon and advised medical management, with good control of blood pressure and pain. The patient improved within a few days without any complications. Finally, the patient was discharged to follow-up with a vascular surgeon. At the 6-month follow-up, he was doing well and did not require surgery.

Figure 1



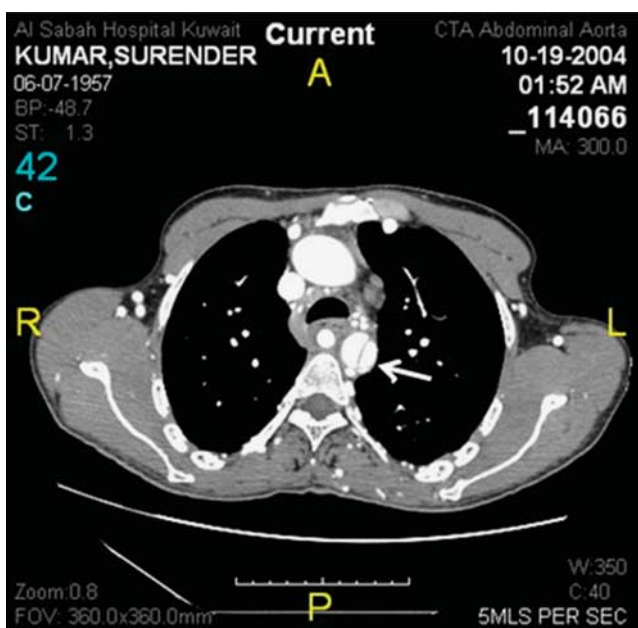
Echocardiogram showing AR.

Figure 2



Ultrasonography abdomen showing thrombus in the aorta (arrow).

Figure 3

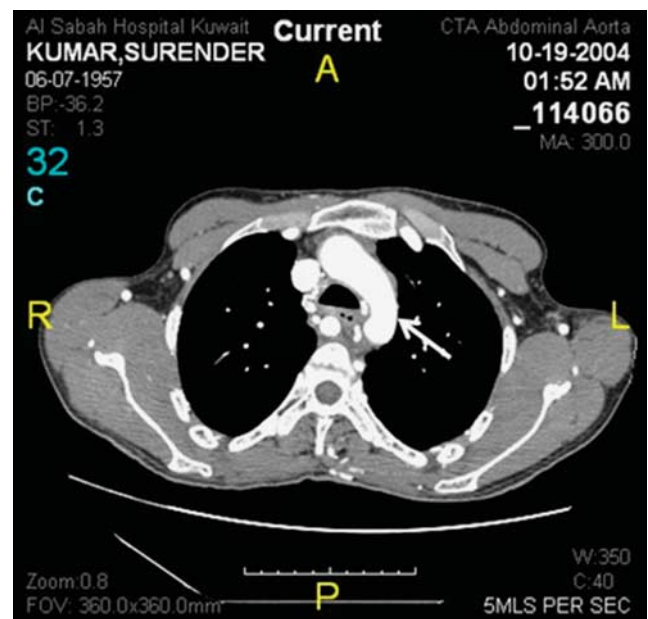


Computed tomography angiogram showing dissection of the descending aorta (arrow).

Discussion

Diseases of the aorta are among the most feared conditions encountered in the emergency department. Acute aortic dissection is not an infrequent clinical entity. The estimated incidence of aortic dissection is 5–30 cases/million individuals/year and is related to the prevalence of risk factors in different study populations [3,4]. If untreated, mortality from type A dissection can be as high as 50% at 72 h and 90% at 3 months [5]. The overall in-hospital mortality for type B dissection is

Figure 4



Computed tomography angiogram showing postductal coarctation of the aorta (arrow).

reported to be 11% but for the patients in the high-risk group, the mortality can be as high as 71%.

Two different anatomic systems, the De Bakey and Daily (Stanford), have been used to classify aortic dissections [6]. The Daily system is used more widely. It classifies dissections that involve the ascending as type A, irrespective of the site of the primary intimal tear, and all other dissections as type B.

Ascending aortic dissections are almost twice as common as descending dissections. The right lateral wall of the ascending aorta is the most common site of the aortic dissection and arch involvement is observed only in up to 30% of cases [7]. There are several variants of aortic dissections including intramural hematoma and intimal tear without hematoma [8].

Patients with aortic dissection typically present with severe, sharp, or tearing/ripping posterior chest or back pain (dissection distal to the left subclavian artery) or anterior chest pain (ascending aortic dissection). In addition to pain, type B dissection can lead to splanchnic ischemia, renal insufficiency, lower extremity ischemia, and focal neurological deficit because of spinal artery involvement, leading to spinal cord ischemia [9].

About 96% of dissections can be identified on the basis of the following three clinical features: abrupt onset of thoracic or abdominal pain that is sharp, tearing, or ripping, mediastinal/aortic widening on chest radiograph, variations in pulse (absence of a proximal extremity or a carotid pulse), and blood pressure difference of more than 20 mmHg between the right and the left arm [10]. Dissection of the descending aorta can masquerade as acute abdominal emergencies such as a peptic ulcer with perforation, acute cholecystitis or pancreatitis, and renal colic [11].

Imaging modalities are the main diagnostic tools and, recently, there has been a shift from an invasive to a noninvasive strategy. Selection of a diagnostic test requires considerations of information, local experience, and expertise. Thoracic CT, MRI, and multiplane transesophageal echocardiography (TEE) are the preferred methods. Bedside TEE is preferred for patients with acute chest pain and those in an unstable condition [12], whereas MRI is a better choice in patients with chronic chest pain and hemodynamically stable patients. CT with contrast is reserved for situations in which both TEE and MRI are unavailable or contraindicated, although CT is still carried out in certain centers as the first choice. Aortography and coronary angiography are recommended only in specific situations.

Medical management remains the standard treatment in uncomplicated type B dissections because of the availability of potent β -blockers and lower mortality compared with a surgical approach, which is reserved only for complicated cases [13]. The short-term prognosis for patients with type B dissections is better than that in patients with type A dissections. Overall, 89% of patients with type B aortic dissections survive to hospital discharge, but the long-term prognosis is poor [14].

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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