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Clinical Manifestations of Aortic Dissection

I. A. Khan

Aortic dissection remains undiagnosed in a substantial number of patients in spite of many remarkable improvements in diagnostic modalities. Although, the most common initial clinical manifestation of aortic dissection is pain, as many as 5 to 15 % of aortic dissections remain painless and result in delay in diagnosis and treatment. The successful outcome in aortic dissection depends on a combination of early diagnosis and prompt treatment, which is based on awareness of the whole spectrum of presenting symptoms. High index of clinical suspicion is the crux of early diagnosis in aortic dissection. Diagnosis of aortic dissection should be considered in patients who present with unexplained symptoms that could be secondary to aortic dissection or its complications, especially so in presence of high-risk predisposing factors. In addition to pain, aortic dissection may manifest with various features of different organ systems involvement. The various clinical manifestations of aortic dissection are elaborated in this article. *J Clin Basic Cardiol 2001; 4: 265–7.*

Key words: aortic dissection, clinical manifestations, clinical presentations, clinical features, painless, asymptomatic

Delay in the accurate diagnosis is a major contributory factor in consistently high mortality of aortic dissection in spite of significant progress in diagnostic and treatment skills [1, 2]. Incidence of aortic dissection is 27–32/million/year, and is related to the prevalence of risk factors, such as hypertension, aortic wall diseases, and advanced age [3, 4]. The mortality of untreated aortic dissection increases by 1 to 2 % per hour after presentation, with 40 % surviving 24 hours and less than 10 % surviving 72 hours [5, 6]. Most of the untreated patients with proximal aortic dissection and half of those with distal dissection die within one year, usually within the first two weeks. Death is usually caused by acute aortic regurgitation, branch vessel obstruction, or aortic rupture, and the risk of fatal rupture of untreated ascending aorta dissection is around 90 % [7]. Long-term survival of aortic dissection in patients who undergo surgical repair and survive long enough to leave the hospital is currently 80 % and 40 % at 5 and 10 years, respectively [8].

The crux of successful outcome in aortic dissection is an early diagnosis and prompt administration of the appropriate treatment. The most common initial clinical presentation of aortic dissection is pain but as many as 5 to 15 % of aortic dissections remain painless, and therefore, cause diagnostic dilemma and delay in treatment [1, 2, 9]. The delay in diagnosis not only delays the treatment but also may result in wrong diagnosis and inadvertent administration of inappropriate medications [10–11]. According to an autopsy study [12], aortic dissection was the most common missed diagnosis, and according to a recently published analysis [13], physicians suspected diagnosis of aortic dissection on initial presentation in 43 % of patients in whom diagnosis was confirmed subsequently. The index of suspicion was highest (86 %) in patients who presented with chest and back pain, followed by chest pain (45 %) and abdominal pain (8 %). Due to a tremendous tragedy associated with late or missed diagnosis of aortic dissection, it is imperative to be aware of all the clinical manifestations of aortic dissection. Aortic dissection has a diversity of presentations, therefore, a high index of suspicion should be used, especially in population at high risk, such as patients with chronic hypertension, Marfan's syndrome, Turner syndrome, Ehlers-Danlos syndrome, aortic aneurysm, anuloaortic ectasia, aortic arch hypoplasia, coar-

tation of aorta, bicuspid aortic valve, connective tissue disorders, pregnancy, and cocaine abuse [14–16].

Pain in Aortic Dissection

Pain is the most common presenting symptom of aortic dissection. It is usually midline pain experienced in front and back of trunk depending on the location of dissection. According to a recently published report from the International Registry of Acute Aortic Dissection, 95 % of patients reported any pain, which was of abrupt onset in 85 % of cases [1]. According to this report, sharp pain was reported by 64 % of patients, whereas the classical tearing or ripping type was reported by 51 % of patients. The most common site of pain was chest (73 %): anterior chest pain more common than the posterior (61 vs. 36 %). The back pain was experienced by 53 % and abdominal pain by 30 % of patients. Patients with dissection of ascending aorta and arch more frequently experience chest pain, whereas, patients with dissection of descending aorta more frequently experience posterior chest, back, and abdominal pain, which may extend down the back to the hips and legs in cases where dissection process extends distally.

Cardiovascular Manifestations

Aortic dissection may present with symptoms secondary to acute aortic regurgitation, which is the second most common cause of death in aortic dissection [4, 17]. Acute aortic regurgitation results in acute congestive heart failure. Moderate to severe aortic regurgitation has been reported in 18 to 45 % cases of proximal aortic dissection [18–19]. Mechanisms underlying aortic regurgitation include dilatation of aortic root and aortic annulus, tearing of annulus or aortic valve leaflets, and downward displacement of one leaflet below the line of valve closure due to pressure from an asymmetric false lumen [17]. Patients with aortic dissection may present with congestive heart failure secondary to the regional wall motion abnormalities, which have been seen in 10 to 15 % of the cases [18, 19]. The regional wall motion abnormalities are primarily due to myocardial ischaemia caused by low coronary perfusion, which could be secondary to compression of a

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coronary artery by the expanding false lumen, extension of the dissection process into a coronary artery, low blood pressure, or a combination of these. Involvement of the right coronary artery is more common than the left sided coronary arteries. Aortic dissection may cause acute myocardial infarction secondary to involvement of the coronary ostium in dissection process [20].

Another cardiovascular presentation of aortic dissection is hypotension/cardiogenic shock [4]. Factors contributing to the precipitation of shock include acute severe aortic regurgitation, acute myocardial ischaemia or infarction, aortic rupture, and pericardial effusion [19–21]. Aortic rupture is the most common cause of cardiogenic shock and death in aortic dissection. Rupture most frequently occurs into the mediastinum. Rupture or leak into pericardial cavity usually results in acute pericardial effusion/cardiac tamponade [22]. Pericardial effusion in aortic dissection in most cases is due to transudate of fluid through intact wall of the false lumen. In a transoesophageal echocardiographic study, pericardial effusion was seen in 48 % of patients with aortic dissection but in most cases it was small, and only 2 out of 19 patients had clinically significant moderate size effusion [18]. Therefore, the presence of pericardial effusion in patients with aortic dissection is not always secondary to rupture of the dissection process into the pericardial cavity, but larger pericardial effusions, even transudative, may result in hypotension and haemodynamic compromise.

False lumen of aortic dissection may compress on or rupture into a surrounding cardiac chamber or great vessel resulting in a different subset of symptoms. Compression of right atrium, superior vena cava, or right pulmonary artery may cause haemodynamic compromise [23–25]. An aorto-right atrial fistula has presented with atypical clinical findings such as a continuous murmur, low-output cardiac failure, or a complete atrioventricular conduction block that was due to haemorrhage at atrioventricular junction [26–28]. About 15 to 20 % of patients with aortic dissection manifest symptoms related to extremity ischaemia, which could be the initial presenting feature [29–32].

Neurological Manifestations

Neurological deficits involving brain, spinal cord, or peripheral nerves are seen in 18 to 30 % cases of aortic dissection [33, 34]. Cerebral ischaemia is the most frequent neurological manifestation with proximal aortic dissection, whereas spinal cord ischaemia and ischaemic neuropathies are more common with distal aortic dissection [33]. Stroke affects 5 to 10 % of cases of aortic dissection, and is generally massive [33–36]. It is caused by dissection and occlusion of the carotid arteries. The low cerebral perfusion may cause symptoms of transient cerebral hypoperfusion ranging from altered mental status to syncope [37, 38]. Stroke and syncope have been reported as initial presenting symptoms of aortic dissection [35–37].

Spinal cord is involved in up to 10 % of the cases of aortic dissection. It is associated with the dissection of the descending aorta. Occlusion of the intercostal arteries, artery of Adamkiewicz, or thoracic radicular artery may cause various spinal cord syndromes, including transverse myelitis, progressive myelopathy, spinal cord infarction, anterior spinal cord syndrome, paraplegia, and quadriplegia [38–41]. A watershed area found between the territories of the artery of Adamkiewicz and the thoracic radicular artery is more prone to ischaemic damage from aortic dissection [42]. There are reports where aortic dissection presented with paraplegia, transient reversible paraplegia, and transverse myelopathy secondary to the spinal cord infarction [36, 38, 39, 43].

Peripheral nerve involvement in aortic dissection is rare but may result in protean neurological symptoms. It could be secondary to neuronal ischaemia or by the direct compression of a nerve by the false lumen of dissection. Patients with aortic dissection could present with paresthesia due to acute peripheral nerve ischaemia and ischaemic lumbosacral plexopathy, and Horner's syndrome due to injury to the superior cervical sympathetic ganglion [43, 44]. Similarly, they may present as hoarseness of voice due to compressive involvement of the recurrent laryngeal nerve by an expanding false lumen at level of the aortic arch [45]. The lengthy course of the recurrent laryngeal nerve in thoracic cavity and around the aortic arch may make it more vulnerable to compression or traction by the dilated false lumen of dissection.

Pulmonary Manifestations

Pulmonary manifestations of aortic dissection are rare. Left pleural space is the third most common space after mediastinum and pericardial cavity where aortic dissection ruptures. Dissection of the descending thoracic aorta may present with left-sided haemorrhagic pleural effusion [46, 47]. Aortic dissection should be considered in the differential diagnosis of the unexplained, nontraumatic left haemorrhagic pleural effusion. Major pulmonary arteries may become involved in the process of dissection [24, 48]. The expanding false lumen can rupture into the pulmonary artery resulting in acute aorto-pulmonary fistula and severe haemodynamic compromise [48]. Aortic dissection has been misdiagnosed as acute pulmonary embolism because of the acute occlusion of right pulmonary artery due to extrinsic compression by dissecting aneurysm [24]. External compression of a branch of pulmonary artery by the expanding false lumen may result in unilateral pulmonary oedema [49], and compression of lung parenchyma may result in haemoptysis [50].

Gastrointestinal Manifestations

Acute gastrointestinal haemorrhage is a rare presentation of aortic dissection [51, 52]. Anatomic proximity of esophagus makes it more vulnerable to be involved in the dissection process [53, 54]. Dissecting aortic haematoma may perforate esophagus resulting in an acute gastrointestinal bleeding [54]. Direct compression of oesophagus by the false lumen of aortic dissection may result in oesophageal ischaemia, dysphagia, and odynophagia [53, 55], and involvement of the mesenteric arteries may result in bowel infarction and intestinal haemorrhage [52, 56]. A false aneurysm formation in retroperitoneum secondary to aortic dissection may rupture into duodenum resulting in massive gastrointestinal bleeding [51].

Other Manifestations

There are reports where patients with aortic dissection have presented with fever of unknown origin, which typically has been accompanied by anaemia and high erythrocyte sedimentation rate [57]. Aortic dissection has been diagnosed occasionally on an imaging study, such as transoesophageal echocardiography, computerized tomography, or magnetic resonance imaging, which was done apparently for some other reason [58–60].

Conclusion

The diagnosis of aortic dissection should be considered in patients who present with unexplained symptoms that could be secondary to aortic dissection or its complications, espe-

cially so in presence of the high-risk predisposing factors for aortic dissection. Variations in the clinical manifestations and lack of clinical suspicion may delay diagnosis, especially in cases of painless aortic dissection. Awareness of the whole spectrum of the clinical manifestations of aortic dissection is an important element in its early diagnosis and management. Clinical features of aortic dissection should be appreciated more broadly because it remains undiagnosed in a substantial number of patients in spite of the remarkable improvement in the diagnostic modalities.

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