Conservative management of chronic aortic dissection with underlying aortic aneurysm

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Abstract

Aortic dissection is one of the most common aortic emergencies affecting around 2000 Americans each year. It usually presents in the acute state but in a small percentage of patients aortic dissections go unnoticed and these patients survive without any adequate therapy. With recent advances in medical care and diagnostic technologies, aortic dissection can be successfully managed through surgical or medical options, consequently increasing the related survival rate. However, little is known about the optimal long-term management of patients suffering from chronic aortic dissection. The purpose of the present report is to review aortic dissection, namely its pathology and the current diagnostic tools available, and to discuss the management options for chronic aortic dissection. We report a patient in which chronic aortic dissection presented with recurring episodes of vomiting and also discuss the management plan of our patient who had a chronic aortic dissection as well as an underlying aortic aneurysm.

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Introduction

Aortic dissection is a relatively common acute emergency affecting 2-3 per 100,000 people per year.[1] It usually presents with the patient complaining of a severe chest or abdominal pain. The exact location of the pain varies with regards to which area of the aorta is affected. However, in a small number of cases, aortic dissection occurs gradually thereby not prompting the patient to seek medical care. We discuss the case of a patient who presented to our hospital with what was eventually diagnosed as a chronic aortic dissection. The medical history of the patient, the investigations requested, as well as the follow up plan devised by the medical team are also discussed.

Case Report

A 68-year old female presented to the emergency department complaining of nausea and vomiting which were getting progressively worse over the morning of her admission. She also complained of a central epigastric pain, which she described as sharp, non-stabbing and non-radiating. The pain was rated as 9 out of 10 on the intensity scale and she recalled the pain started shortly after the episodes of vomiting. Episodes of headache, dizziness, lightheadedness and slight blurring of vision were also reported but she denied any syncope, fever or chills. In terms of past medical history, she suffered from hypertension, hyperlipidemia, an abdominal aortic aneurysm and chronic renal insufficiency status post pyleography.

On examination on admission to the emergency department, the patient was found to be in hypertensive crisis with the highest blood pressure reaching 206/102 on her left arm and 243/129 on her right arm. On general observation, she was found to be appropriately orientated but in moderate distress with diaphoresis. Clinical observations recorded included a heart rate of 74 beats/min, respiratory rate of 32/min, while pulse oximetry was 97%. Abdominal examination revealed a soft, non-tender, non distended abdomen with normal bowel sounds. No carotid bruits were heard on auscultation and no palpable or pulsating mass was found. The epigastric area was tender but there was no guarding or rebound tenderness. Her cardiovascular examination revealed mild tachycardia, normal heart sounds, no murmurs and no gallop. Her respiratory examination was unremarkable apart from the presence of rales in the lungs. No abnormalities were detected in the peripheral pulses. As a first-line investigation, an echocardiogram was ordered which showed a normal sinus rhythm and non-specific ST-T wave changes. Cardiac enzymes, amylase, lipase and troponins were also ordered and were all within normal limits. Her chest X-ray was also normal showing a normally proportioned heart and no abnormality in the thoracic aorta. There was no pulmonary congestion, infiltrates or pleural effusions. Other baseline investigations included a complete blood count, liver function tests, and urea, electrolytes and creatinine levels.

Based on the location, nature of the pain and the clinical observations of the patient, a provisional diagnosis of acute aortic aneurysm was made. The first-line management was to control her excessively high blood pressure so as to reduce the risk of any possible complications. She was, therefore, started on an i.v. nitroglycerin drip in the emergency department and later switched to a nicardipine i.v. drip as there was only an initial response to the nitroglycerin. With her pain adequately managed, she was admitted to the cardiac care unit to ensure strict blood pressure control which was appropriately kept under 110/70. Computed tomography (CT) of the chest, abdomen and pelvis was performed and an aortic aneurysm was found at the base of the diaphragm. This was consistent with her history of an aneurysm that in 2006 measured 4.4 cm. Further investigation with an MRI confirmed the presence of an aneurysm still spanning 4.4 cm, but in addition, a chronic aortic dissection in the descending thoracic aorta was also seen. The dissection was located at the level of the celiac artery and measured 5.0 cm. The presence of thrombi and calcifications within the lumen created by the aortic

dissection makes the dissection consistent with a chronic presentation *versus* an acute episode.

Once stabilized, the patient was reassessed and it was concluded that her epigastric pain was most likely due to hypertensive emergency with an underlying aortic aneurysm and chronic aortic dissection. Based on the location of the dissection, it was classified as Stanford B, and given the aneurysm was less that 5.5 cm, it was decided that it would be best managed conservatively with strict blood pressure control. The patient was, therefore, started on labetolol. Strict control of the patient's blood pressure and hyperlipidemia was also the conservative choice in treatment of the coexisting aneurysm. The patient has now been monitored for the last two years.

Discussion

Aortic dissection classically results from blood separating the layers of the aortic media forming a false lumen. In over 90% of cases, the aortic intima is the one which initiates the aortic dissection exposing a usually compromised aortic medial layer to high pressure arterial pressure resulting in a further and more serious tear through the aorta.[2,3] Degeneration of the aortic media and loss of muscle tissue cause the aorta to dilate even further, increasing the wall tension as well as the false lumen diameter. The transition between the dilated and undilated portion of the aorta is subject to high wall tension and is also the region with the strongest pulsatile force.[4] The cause of the initial tear varies but frequent predisposing factors include advanced age, atherosclerosis and connective tissue disorders.[5]

In patients below the age of 40 years, deficiencies in the collagen and elastin content of the wall in cases such as Marfan's syndrome are the likely cause of an aortic dissection.[1] Vascular conditions that damage the elastic and muscular component of the aorta also increase the likelihood of an aortic dissection, but the most common predisposing factor for an aortic dissection is hypertension.[6]

Typically, aortic dissections can be classified using various parameters, one of them being the anatomical extent of the dissection.[7] Proximal dissections involving the ascending aorta are termed Stanford type A or DeBakey type I or II, whereas Stanford type B or DeBakey type III affect the distal and descending aorta.[8,9]

Recent studies suggested that intramural hemorrhage, intramural hematoma and aortic ulcers may also be suggestive of an evolving dissection and, for this reason, an additional form of classification has been proposed:[10] Class 1 describes a classic aortic dissection with an intimal flap between true and false lumen; Class 2, a medial disruption with formation of intramural hemorrhage; Class 3 involves a subtle dissection without hematoma; Class 4 is characterized by a plaque rupture leading to aortic ulceration surrounding hematoma; Class 5 describes an Iatrogenic and traumatic dissection. In addition to classification based on location, aortic dissection can either be acute or chronic. Conventionally, an acute aortic dissection is any dissection presenting within 14 days whereas in patients who present after this 2-week window it is termed as chronic.[11-13] In around 80% of cases, aortic dissection presents with an acute onset of chest pain that radiates to the back prompting the patient to seek medical care.[6] If the dissection is present in the thoracic portion of the aorta, the patient may also experience a worsening chest pain, a non-productive cough due to bronchial irritation or dysphasia due to the compression of the esophagus.[10] However, in about 30% of patients, aortic dissection is diagnosed in its chronic state. This is because in a small portion of patients symptoms of a chronic aortic dissection can be vague and non-specific, thereby not prompting the patient to seek medical help.[10,14] In around 2% of cases, chronic thoracic aortic dissection can cause paralysis or paraplegia due to distal embolus of the thrombus, and in cases in which the chronic aortic dissection involves the descending or abdominal aorta, patients may experience intermittent claudication due to the involvement of the iliac arteries. In the event in which the renal arteries are compromised, an increased blood creatinine and a reduction in urine output will follow.[15] Because of the various symptoms that can be associated with a chronic aortic dissection, the latter is often not suspected and is, therefore, missed. As a consequence, these patients are left undiagnosed and the aortic dissection is picked up in its chronic state either incidentally or as part of an investigation for another complaint.

Other reasons for the late diagnosis of AD include: i) patients surviving surgery or medical therapy for a persistent aortic dissection; and ii) acute patients being treated medically for any aortic conditions.[13,16,17]

The most commonly used imaging techniques used in establishing an aortic dissection diagnosis are chest X-rays, computerized axial tomography, magnetic resonance imaging (MRI) and trans-esophageal echography (TEE). On a chest X-ray, positive findings include an abnormal curvature of the aortic arch, superior mediastinal widening with pleural effusion, whereas on a CT scan, aortic wall thickening and mediastinal displacement are usually diagnostical.[6,18,19]

A chronic aortic dissection tends to be associated with a weakened aortic wall the degeneration of which can eventually lead to the rupture of the aorta. Another important complication of a chronic dissection is the gradual impairment of distal perfusion which can eventually lead to ischemic syndromes. In order to detect any unstable aortic weakness, it is advisable to order serial imaging of the aorta either in the form of MRI or CT. Measuring the diameters of the aorta at various levels and evaluating the state of the false lumen will provide a thorough and continuous assessment of the aortic dissection. Any complication should then be detected and addressed before it causes any life-threatening complication.[20]

Treatment for aortic dissection varies greatly according to the location and severity. Surgery is generally indicated for dissection involving the ascending aorta, mainly because of the higher risk of complications associated with it. Medical management, on the other hand, is generally reserved for descending aortic dissections.[10]

The general prognosis of chronic aortic dissection varies between patients according to the location of the dissection and the extent to which corresponding vessels are involved. Chronic Stanford Type A aortic dissection is often associated with a higher risk of complications with surgical intervention during which the area of the aorta with the intimal tear is resected and replaced with a Dacron graft.[10] The operative mortality rate is usually less than 10%, and serious complications are rare with ascending aortic dissections.

The development of more impermeable grafts, such as woven Dacron, collagen-impregnated Hemashield (Meadox Medicals, Oakland, NJ, USA), aortic grafts, and gel-coated Carbo-Seal Ascending Aortic Prothesis (Sulzer CarboMedics, Austin, TX, USA) has greatly enhanced the surgical repair of thoracic aortic dissections, and with the introduction of profound hypothermic circulatory arrest and retrograde cerebral perfusion, the morbidity and mortality rates associated with this highly invasive surgery have decreased.

Dissections involving the arch are more complicated than those involving only the ascending aorta because the innominate, carotid, and subclavian vessels branch from the arch; deep hypothermic arrest is, therefore, usually required. An arrest time of less than 45 min is usually associated with a 10% incidence of central nervous system complications. Retrograde cerebral perfusion may also increase the protection of the central nervous system during the arrest period.

Aortic stent grafting is another form of surgical intervention and may prove feasible. Recent studies have shown that it has offered good results in a small series of patients. It

may be a reasonable alternative in high-risk patients in the near future

Studies have shown that patients with Stanford type A who undergo surgical treatment have a 30% mortality rate whereas patients who receive medical treatment have a mortality rate of 60%. Co-morbidities and advanced aged can pose a contraindication to surgery in selected patients. In Chronic Stanford Type B aortic dissection, medical management is generally advised with research showing that medically treated patients with type B dissection have a 10% mortality rate whereas surgically treated patients with type B dissection have a 30% mortality rate.[21] As part of the general management of chronic aortic dissection, it is also advisable for clinicians to treat the chronic aortic dissection as well as the underlying cause and any risk factors that the patient may have. Such risk factors include increasing age, increased aortic size and the presence of a patent false lumen.

The presence of a persistent hypertension, which was the case with our patient, should also be appropriately controlled.

Another factor which should be appropriately controlled in the management of chronic aortic dissections is the presence of any connections between the true and false lumen which can further increase the risk of an aortic dissection. It has previously been suggested that a thrombosed false lumen slows the rate of aortic growth. However, recent studies by Tsai *et al*. which investigated the presence of fenestrations in patients with Stanford type B ortic dissections have concluded that the prognosis of patients is not improved whether a false lumen is thrombosed or not.[21] Nevertheless, the ultimate goal of the management of chronic aortic dissection is to detect any weakness in the aortic wall and to manage it appropriately so as to completely restore blood flow in the true aortic lumen. Various measures have been suggested for the long-term management of aortic dissection and they all have the end goal of reducing the pressure on the dissection. Long-term medical therapy usually involves a beta-adrenergic blocker combined with other anti-hypertensive medications. Anti-hypertensives (*e.g*. hydralazine, minoxidil) that produce a hyperdynamic response that would increase dP/dt (*i.e*. alter the duration of P or T waves) should, however, be avoided.

A chronic aortic dissection tends to be associated with complications which weaken the aortic wall. Degeneration of the aortic vessel can eventually lead to the rupture of the aorta. Another important complication of a chronic dissection is the gradual impairment of distal perfusion which can eventually lead to ischemic syndromes

Studies have shown that appropriate control of blood pressure reduces the rate at which the aortic dissection progresses and decreases the risk of complications. All patients with a chronic aortic dissection should, therefore, have their blood pressure strictly controlled whether they have undergone surgical repair or not. In fact, current guidelines recommend that the blood pressure of patients at risk of developing an aortic dissection should be kept below 130/80 mmHg.[17,22,23] Anti-hypertensive agents such as beta blockers are recommended since they have the additional advantage of reducing aortic wall stress, thereby improving the prognosis of aortic dissection patients. In cases in whom the patient is unresponsive to beta blockers, calcium channel antagonists are a reasonable alternative.[11]

Other anti-hypertensive medications such as angiotensin converting enzyme-inhibitors are believed to lower the risk of complications from aortic dissections such as aneurysm rupture by suppressing any inflammation occurring within the aortic walls. Lipid lowering agents such as statins are also believed to reduce any inflammation and are, therefore, beneficial in controlling blood pressure.[24]

In the majority of cases, the first line of management of a chronic aortic dissection is medical therapy. However, in up to 40% of cases, a surgical intervention is often recommended.[25,26]

Operative repair of the aneurysm and insertion of a prosthetic graft are indicated for abdominal aortic aneurysms of any size that are expanding rapidly or are associated with symptoms. For asymptomatic aneurysms, surgery is indicated if the diameter is over 5.5 cm. In randomized trials of patients with abdominal aortic aneurysms below 5.5 cm, there was no difference in the long-term (5-8 year) mortality rate between those followed with ultrasound surveillance and those undergoing elective aneurysm repair. Therefore, serial non-invasive follow up of smaller aneurysms (<5 cm) is an alternative to immediate surgery.

Propagation of the dissection resulting in an aortic diameter over 60 mm and a growth rate greater than 5 mm annually is also a major surgical indication. Others include an enlarging hematoma compromising major aortic branches, bleeding into the pleural cavity, and development of saccular aneurysm.

There are two possible surgical options. In an open aortic aneurysm repair, the aneurysm sac is opened and a prosthetic graft is used to reconstruct the aorta. If the aneurysm only involves the abdominal aorta, a tube graft can be used to replace the aorta. If the aneurysm extends distally to the iliac arteries, a prosthetic bifurcated graft is used for either an aorto-bi-iliac or aorto-bi-femoral bypass reconstruction.

The other surgical intervention is the endovascular approach, which involves the reconstruction of the aortic arch segment using a Dacron-covered stent. The principle of endovascular repair involves the implantation of an aortic stent graft that is fixed proximally and distally to the non-aneurysmal aortoiliac segments thereby excluding the aneurysm from the aortic circulation through the endolumen. Unlike open surgical repair, endovascular treatment does not remove or eliminate the aneurysm sac which is, therefore, subjected to potential aneurysm expansion or even rupture as persistent aneurysm sac pressurization may occur following endograft implantation. Anatomic eligibility for endovascular repair is mainly based on three areas: the proximal aortic neck, common iliac arteries, and the external iliac and common femoral arteries, which relate to the proximal and distal landing zones or fixation sites and the access vessels, respectively.

Endovascular stenting remains an option for treatment of some type B dissections. Some studies recommend that patients with complicated acute type B dissections undergo endovascular stenting with the aim of covering the primary intimal tear. Repair of the descending aorta is associated with a higher incidence of paraplegia than repair of other types of dissections because of interruption of segmental blood supply to the spinal cord.

The primary success rate after endovascular repair has been reported to be as high as 95%. The less invasive nature of this procedure is attractive to many physicians and patients. In addition, virtually all reports indicate a decrease in blood loss, transfusion requirements, and length of intensive care and hospital stay for endovascular repair of abdominal aortic aneurysm when compared to the standard surgical approach. With the advent of bifurcated grafts and improved delivery systems in the future, the only real limitation will be cost.[27]

The DREAM trial is a multicenter randomized trial that compared open *versus* endovascular repair among a group of 345 patients at 28 European centers using multiple different devices including: Gore, AneuRx, and Zenith. Patients were included only if they were considered to be candidates for both types of repairs. The operative mortality rate was 4.6% in the operative group *versus* 1.2% in the endoluminal group at 30 days. When looking at the combined rate of operative mortality and severe complications, there was an incidence of 9.8% in the open-repair *versus* 4.7% in the endoluminal group. The difference here was largely due to the higher frequency of pulmonary complications seen in the open group. There was a higher incidence of graft-related complications in the endoluminal group. There was no difference in the rate of non-vascular local complications between the two groups.[27,28]

Conclusions

Aortic dissection is a common condition which typically presents in the acute phase. In a small number of patients it can present with atypical symptoms thereby delaying its diagnosis and progressing into a chronic condition. Clinicians should, therefore, maintain a high degree of clinical suspicion. With advanced medical technology and diagnostic techniques, the prognosis of patients suffering from aortic dissections has improved drastically but with a progressively ageing population, there is an increasing risk of complications and a high risk of mortality and morbidity associated. Careful follow up of patients with chronic aortic dissection is important and routine investigations such as serial imaging should be carried out to detect any early complication. It is recommended that all patients with an aortic dissection have a physical examination, a chest X-ray and imaging of the aortal every six months.

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