

An under recognized cause of chest pain

Unnikrishnan Ponnamma Kunjan Pillai,
Santosh G. John,
Aparna Narayana Kurup,
Joe Devasahayam, Alexandre Lacasse

Department of Internal Medicine, St.
Mary's Health Center, St. Louis, Missouri,
USA

Abstract

Aortic intramural hematoma (IMH) is related to but is pathologically distinct from aortic dissection. In this potentially lethal entity, there is hemorrhage into the aortic media in the absence of an intimal tear. With recent advances in imaging techniques, IMH is now increasingly recognized. The limited data available suggest that the clinical course of IMH mimics that of acute aortic dissection, and mortality rates are similar. Physicians need to be cognizant regarding this entity when they are evaluating chest pain. Here we report a case of IMH, in a 63-year-old female, which was managed conservatively.

Case Report

A 63-year-old Caucasian female presented to the emergency department with the chief complaint of intense pain between her shoulder blades, which began a few hours back. The patient reported that she was feeling well early in the day but began to experience a sharp, intense pain between her shoulder blades, which started suddenly while she was eating. She rated the pain as 8/10 and described the pain as a feeling of being *hit with a baseball bat*. The pain was unrelieved with rest and fluids. She denied any chest pain or shortness of breath, but reported some mild diaphoresis. She denied abdominal pain, but felt nauseated and vomited clear liquid mixed with food before coming to the hospital. Patient had no medical problems, was on no medications, and had not seen a physician in over five years. Patient had a 100-pack year smoking history and drinks 4-5 beers daily. She denied any illicit drug use.

On physical exam, she was noted to be a thin, frail woman in mild distress secondary to her back pain. The patient was afebrile. Her pulse was regular with a rate of 94 beats per minute. Her blood pressure was 230/134 mmHg. Her oxygen saturation was 95% on room air. The patient's lungs were clear to auscultation bilaterally with normal respiratory

effort. On cardiac examination, there was no jugular venous distension or parasternal heave. Auscultation revealed a normal S1 and S2 without murmur, rub, or gallop. Her abdomen was soft and nontender with normal bowel sounds and no bruit. The pain in her back was not reproducible with palpation and there were no signs of deformity or trauma. The peripheral arterial pulses were palpable and symmetric in all four extremities.

Laboratory data showed mild leukocytosis, with a white blood cell count of 12.400 mm³ (normal range, 4.000-10.000) and hypokalemia, with a serum potassium of 3.3 mmol/L (normal range, 3.6-5.0). Amylase and lipase were within normal limits. An electrocardiogram showed normal sinus rhythm. The patient was diagnosed with hypertensive urgency and treated with aspirin, sublingual nitroglycerin, transdermal nitroglycerin, and sublingual clonidine. Her blood pressure decreased to 102/79 and her back pain abated. The patient was to be admitted to the hospital for continued control of her blood pressure and further evaluation. A computed tomography (CT) angiography of the chest with and without contrast was obtained (Figure 1) which was followed by magnetic resonance (MR) angiogram (Figure 2). CT angiogram showed diffuse abnormal wall thickening of the descending thoracic aorta, which is peripheral to the calcified intima and extends up to the origin of the celiac and superior mesenteric arteries (Figure 1). MR angiogram of thoracic aorta showed thickening of the wall of the descending thoracic aorta starting from just beyond the origin of the left subclavian artery, with no definite wall thickening of the ascending thoracic aorta or evidence of a pericardial effusion (Figure 2). Transesophageal echocardiogram revealed an abnormal descending aorta with a thickened wall, which was eccentric and had a uniform echo dense appearance consistent with intramural hematoma, which ended at the origin of the left subclavian artery. There was no evidence of any intimal tear (Figure 3). She was admitted to the intensive care unit, her blood pressure was controlled using intravenous labetalol. Since it was a Type-B intramural hematoma, it was decided to manage conservatively with close outpatient follow-up.

Discussion

Acute aortic syndrome refers to the spectrum of aortic emergencies that include aortic dissection, intramural hematoma, penetrating atherosclerotic ulcer of the aorta, aortic aneurysm leak and rupture and traumatic aortic transection. A classic aortic dissection begins with a laceration of the aortic intima and inner layer of the aortic media forming an

Correspondence: Unnikrishnan Ponnamma Kunjan Pillai, Department of Internal Medicine, St. Mary's Health Center, St. Louis, Missouri, USA.

E-mail: unnikrishnanpk@yahoo.com

Key words: intramural hematoma, dissection.

Received for publication: 13 November 2011.

Revision received: 27 December 2011.

Accepted for publication: 2 January 2012.

This work is licensed under a Creative Commons Attribution NonCommercial 3.0 License (CC BY-NC 3.0).

©Copyright U.P.K. Pillai et al., 2012

Licensee PAGEPress, Italy

Clinics and Practice 2012; 2:e12

doi:10.4081/cp.2012.e12

entrance tear that allows entering blood to split the aortic media.¹

Aortic intramural hematoma (IMH), first described in pathological literature in 1920, is related to but is pathologically distinct from aortic dissection. It is characterized by the absence of an entry tear, absence of an intimal flap, and absence of evidence of communication between the medial hemorrhage and the aortic lumen.² In a review of 505 autopsy cases of aortic dissection from 1933 to 1954, Hirst *et al.* found a 4% incidence of IMH.³ These intramural hematomas like dissections, involve the ascending aorta, arch or both (Type A) or the descending aorta (Type B). Though deemed as pathologically distinct entities, intramural hematoma may lead into acute aortic dissection, aneurysm or aortic rupture.⁴

Aortic intramural hematoma may be a primary event in hypertensive patients due to spontaneous bleeding from vasa vasorum into the media or may be caused by a penetrating atherosclerotic ulcer.⁴ Vaso vasorum *vessels of vessels*, forms network of vessels in adventitia, which also supply the outer media plays a significant role in pathogenesis of all aortic pathologies including intramural hematoma, aortic dissection and aortic aneurysm.⁴ Animal studies have shown that aortic distensibility decreases when vaso vasorum are removed from the vessel wall. Penetrating atherosclerotic ulcer develops, when an atheromatous plaque, ulcerates through the internal elastic lamina and exposes the media to high-pressure aortic blood flow leading to intra medial hematoma.⁵

The risk factors for IMH are very much similar to cardiovascular diseases, with hypertension being the most common among them. Pregnancy and some congenital disorders such as Marfan's syndrome, Ehlers-Danlos syndrome, annuloaortic ectasia and bicuspid aortic valve are the other possible predisposing



Figure 1. Computed tomography angiography of the chest showed diffuse abnormal wall thickening of the descending thoracic aorta.



Figure 2. Magnetic resonance angiogram of thoracic aorta showed thickening of the wall of the descending thoracic aorta.

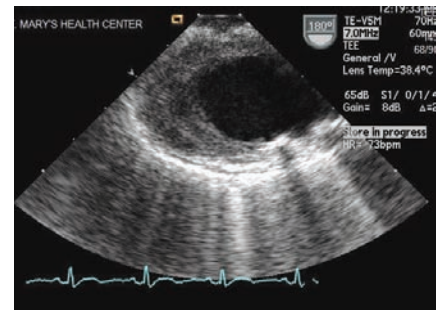


Figure 3. Transesophageal echocardiogram revealed an abnormal descending aorta with a thickened wall.

factors for IMH.⁶ Aortic intramural hematoma can occur as a primary event or as a result of blunt chest trauma, in which there is spontaneous rupture of the nutrient vasa vasorum, with circumferential or longitudinal spread of the hematoma over a variable distance along the media layer of the aorta. Cases of intramural hematoma following cardiac resuscitation have been reported. IMH is difficult to distinguish from classic dissection on purely clinical grounds. In clinical series 13 to 27% of patients with a diagnosis of aortic dissection in fact had IMH.⁷ Patients with IMH are typically elderly with history of hypertension. Unlike classic aortic dissection, ratio of men to women appears equal. Risk factors and clinical features at presentation at presentation are similar to aortic dissection. Clinically IMH most commonly occurs in the descending aorta and in older patients. Chest pain and back pain are the most frequent symptoms.

Diagnostic studies should reveal fresh thrombus within the aortic wall, which should manifest in TEE as either crescentic or circular thickening of the aortic wall with maximal thickness greater than or equal to 7 mm without intimal flap or tear or any longitudinal flow in the false lumen.⁸ On unenhanced CT, intramural hematoma is hyper dense. MRI identifies slow flow in the false lumen in dissection and no flow in an intramural hematoma. Dynamic phase-contrast MRI is more sensitive than gradient refocused echo sequences in differentiating aortic dissection from intramural hematoma.⁹ Intramural hematoma may be distinguished from mural thrombus by the identification of the intima; mural thrombus lies on top of the intima, which is frequently calcified, whereas intramural hematoma is sub intimal. Some patients with intramural hematoma have limited hemorrhage and responds well to medical therapy. Intramural hematoma leads to weakening of the aorta and may progress either to outward rupture of the aortic wall or to inward disruption of the intima which leads to communicating aortic dissection.⁶ Rate of conversion of IMH to dissection varies according to the site of IMH, with various studies

reporting 3% to 14% conversion rates in IAH involving the descending aorta and in 11% to 88% of IMH involving the ascending aorta.⁹ Although a rare phenomenon, spontaneous resorption of IMH has also been reported.⁶

Type A IMH may advance to complete dissection and can rupture through adventitia causing pericardial effusion, hemothorax, and mediastinal hemorrhage. Mortality from proximal lesions is greater than distal IMH and mortality is highest within the first 24 to 72 hours after hospital admission. The maximum thickness of the hematoma on the initial CT is the most significant factor for predicting the development of aortic dissection and aortic aneurysm. Patients with Type A IMH and ulcer like projections, as revealed by initial and short term follow up CT examinations, should be followed up with subsequent CT examinations to monitor for the development of an aortic aneurysm, which is a relatively common chronic complication of IMH.¹⁰ If IMH develops at the convexity of the distal arch, supra-aortic branches prevent retrograde extension toward the ascending aorta. If an aortic IMH develops at the free lateral wall or at the concavity, it may affect the entire thoracic aorta, due to the lack of the natural barrier of the supra-aortic branches.

Definite guidelines for the management of intramural hematoma are yet to be derived. Initial medical treatment, endovascular surgery or classic open surgery is the common treatment of IMH. A study by Ledbetter *et al.*, on morbidity and mortality for 168 patients with IMH, 30-day mortality was 18% with surgical repair of proximal IMH, and 33% with surgery to distal IMH compared to 60% and 8% with medical treatment of type A and type B IMH, respectively.¹¹ Most authorities suggest treatment of intramural hematoma similar to aortic dissection with early surgical intervention in Type A and medical management for Type B lesions.¹² Surgery is usually indicated when there are signs of expansion of the IMH, rupture into the pleural or pericardial cavity, uncontrollable symptoms like chest pain, if the patient becomes hemodynamically unstable or

dilated aorta more than 5 cm.^{6,13} Other possible alternative is endovascular repair if there are no other comorbidities like uncontrolled hypertension or persistent pain.⁶ Oral β -blocker therapy helps in control of heart rate and blood pressure and may improve long-term prognosis of IMH independent of anatomical location. Intramural hematomas have a more favorable outcome compared to aortic dissection, as the hematoma is non-communicating with the aortic lumen.

References

1. Coady MA, Rizzo JA, Elefteriades JA. Pathologic variants of thoracic aortic dissections: penetrating atherosclerotic ulcers and intramural hematomas. *Cardiol Clin* 1999;17:637-57.
2. Schappert T, Sadony V, Schoen F, et al. Diagnosis and therapeutic consequences of intramural aortic hematoma. *J Card Surg* 1994;9:508-15.
3. Hirst AE, Johns VJ, Kime SW. Dissecting aneurysm of the aorta: a review of 505 cases. *Medicine* 1958;37:217-9.
4. Baikoussis NG, Apostolakis EE, Papakonstantinou NA, et al. The implication of vasa vasorum in surgical diseases of the aorta. *Eur J Cardiothorac Surg* 2011;40:412-7.
5. Baikoussis NG, Apostolakis EE. Penetrating atherosclerotic ulcer of the thoracic aorta: diagnosis and treatment. *Hellenic J Cardiol* 2010;51:153-7.
6. Baikoussis NG, Apostolakis EE, Siminelakis SN, et al. Intramural haematoma of the thoracic aorta: who's to be alerted the cardiologist or the cardiac surgeon? *J Cardio-thorac Surg* 2009;4:54.
7. Alfonso F, Goicolea J, Aragoncillo P, et al. Diagnosis of intramural hematoma by intravascular ultrasound imaging. *Am J Cardiol* 1995;76:735-38.
8. Hiratzka LF, Bakris GL, Beckman JA, et al. Guidelines for the diagnosis and management of patients with Thoracic Aortic

- Disease. *Circulation* 2010;121:e266-369.
9. Katarzyna J. Macura, Frank M, et al. Pathogenesis in Acute Aortic Syndromes: Aortic Dissection, Intramural Hematoma, and Penetrating Atherosclerotic Aortic Ulcer. *AJR* 2003;189:309-16.
 10. Lee YK, Seo JB, Jang YM, et al. Acute and chronic complications of aortic intramural hematoma on follow-up computed tomography: incidence and predictor analysis. *J Comput Assist Tomogr* 2007;31:435-40.
 11. Ledbetter S, Stuk JL, Kaufman JA. Helical (spiral) CT in the evaluation of emergent thoracic aortic syndromes: Traumatic aortic rupture, aortic aneurysm, aortic dissection, intramural hematoma, and penetrating atherosclerotic ulcer. *Radiol Clin North Am* 1999;37:575-89.
 12. Sawhney NS, DeMaria AN, Blanchard DG. Aortic intramural hematoma: an increasingly recognized and potentially fatal entity. *Chest* 2001;120:1340-6.
 13. Baikoussis NG, Apostolakis EE. Penetrating atherosclerotic ulcer of the thoracic aorta: diagnosis and treatment. *Hellenic J Cardiol* 2010;51:153-7.