Emerging Concepts in Intramural Hematoma Imaging[1]

Susan E. Gutschow, MD
Christopher M. Walker, MD
Santiago Martínez-Jiménez, MD
Melissa L. Rosado-de-Christenson, MD
Justin Stowell, MD
Jeffrey R. Kunin, MD

Abbreviations: AAS = acute aortic syndrome, IMH = intramural hematoma, MIP = maximum intensity projection, PAU = penetrating aortic ulcer

RadioGraphics 2016; 36:660–674

Published online 10.1148/rg.2016150094

Content Codes: CH CT VA

[1]From the Department of Radiology, University of Missouri–Kansas City, Kansas City, Mo (S.E.G., C.M.W., S.M.J., M.L.R.d.C., J.S., J.R.K.); and Department of Radiology, Thoracic Imaging Division, Saint Luke's Hospital, 4401 Wornall Rd, Kansas City, MO 64111 (S.E.G., C.M.W., S.M.J., M.L.R.d.C., J.R.K.). Presented as an education exhibit at the 2014 RSNA Annual Meeting. Received April 5, 2015; revision requested September 30 and received October 26; accepted November 24. For this journal-based SA-CME activity, the authors C.M.W., S.M.J., and M.L.R.d.C. have provided disclosures (see end of article); all other authors, the editor, and the reviewers have disclosed no relevant relationships. **Address correspondence to** S.E.G. (e-mail: *susan.gutschow@gmail.com*).

SA-CME LEARNING OBJECTIVES

After completing this journal-based SA-CME activity, participants will be able to:

- Define IMH as a component of AAS, including its pathogenesis, natural progression, and potential complications.
- Discuss recently described imaging findings seen in IMH, particularly those associated with greatest risk for complications and mortality that should be reported by the radiologist.
- Describe surgical, endovascular, and nonsurgical treatment of IMH.

See www.rsna.org/education/search/RG.

Intramural hematoma (IMH) is included in the spectrum of acute aortic syndrome and appears as an area of hyperattenuating crescentic thickening in the aortic wall that is best seen at nonenhanced computed tomography. IMH is historically believed to originate from ruptured vasa vasorum in the aortic media without an intimal tear, but there are reports of small intimomedial tears identified prospectively at imaging or found at surgery in some cases of IMH. These reports have blurred the distinction between aortic dissection and IMH and raise questions about what truly distinguishes the entities that compose acute aortic syndrome. The pathophysiology of these subgroups and the controversies surrounding their differentiation are discussed. The natural history of IMH is highly variable; it may resolve or progress to aneurysm, dissection, or rupture. The authors review various imaging prognostic factors that should be reported by the radiologist, including Stanford classification, maximum aortic diameter, maximum IMH thickness, focal contrast enhancement (including ulcerlike projection and intramural blood pool), and pleural or pericardial effusion. Medical (nonsurgical) versus surgical treatment strategies depend primarily on the Stanford classification, although more recent studies of Asian cohorts report success of initial medical treatment in patients with Stanford type A IMH, with timed (delayed) surgery for patients who develop complications. Understanding the imaging appearance and prognostic factors of IMH helps the radiologist and surgeon identify patients at greatest risk for complications to ensure appropriate treatment and improve patient outcomes.

Introduction

Acute aortic syndrome (AAS) is classically composed of three entities: aortic dissection, intramural hematoma (IMH), and penetrating aortic ulcer (PAU) (Fig 1). Some include a fourth category, the recently described incomplete dissection. These entities share a common clinical presentation and risk factor profile, including acute chest pain and chronic hypertension (1). Despite these similarities, IMH is considered distinct from aortic dissection, PAU, and incomplete dissection. Historically, IMH has been thought to be secondary to spontaneous rupture of the vasa vasorum, often referred to as rhexis of the vasa vasorum, and is distinguished from aortic dissection in that the aortic intima remains intact in IMH. However, multidetector CT images and surgical reports have confirmed small intimomedial tears in a variable percentage of patients diagnosed with IMH (2–5). The term *intimal flap* with regard to imaging is a misnomer, as the actual flap tissue is composed of both the aortic intima and variable amounts of the delaminated media. Thus, the term *intimomedial flap* is preferred (6).

TEACHING POINTS

- IMH has historically been differentiated from aortic dissection in that the intimal layer remains intact in IMH, and IMH is believed to originate from ruptured vasa vasorum in the aortic media. However, there are increasing reports of small intimomedial tears not apparent at preoperative imaging that were found at surgery in a variable percentage of IMH cases, prompting discussion about the importance of the "microintimal tear" and whether it is the true inciting event rather than vasa vasorum rupture.
- With the advent of improved technology in the last 2 decades, more intimomedial tears are being identified in patients with IMH; thus, a better distinction may be that aortic dissection contains two intimomedial tears—an entry tear from the lumen into the media, and a reentry tear back into the aortic lumen—while IMH with intimomedial tear often has only an entry tear.
- IMH exhibits crescentic or circular aortic wall hyperattenuation that is best visualized at nonenhanced CT and may demonstrate displacement of intimal calcifications in a curvilinear distribution. This crescentic hyperattenuation is better visualized with thicker 5-mm sections than with thin sections, as there is more volume averaging and less noise with thicker slabs. Although most radiologists use thin-section acquisition for aortic protocol CT, reformatted images with a 5-mm section thickness are helpful and should be obtained in the nonenhanced portion of the examination. Use of a narrow window (width, 200 HU; level, 40 HU) will also aid in hematoma detection on nonenhanced images. Subacute IMH may be isoattenuating relative to the blood pool at nonenhanced CT. Administration of contrast agent will demonstrate a decreased diameter of the aortic lumen without diffuse enhancement of the hematoma.
- The natural history of IMH is variable. It may regress, resolve, enlarge, or progress to aneurysm or dissection. Multiple imaging prognostic factors have been identified that indicate a higher risk for complications or progression.
- Treatment strategies for IMH are similar to those for aortic dissection and depend on whether the hematoma is Stanford type A or type B.

Other entities such as PAU can result in IMH, and incomplete dissection can result in a subadventitial hemorrhage that may mimic IMH at imaging. The relationships among the entities that compose AAS are shown in Figure 2. These relationships raise questions about what differentiates the spectrum of diseases under AAS and whether the causes are similar despite their distinct imaging appearances. Regardless of the cause, it is important for the radiologist to understand pertinent imaging findings that may affect patient prognosis and treatment.

In this article, we define IMH as a component of AAS. We also describe the pathogenesis, natural progression, and potential complications of IMH, expanding on recently described imaging findings in IMH and emphasizing findings that affect prognosis. Finally, surgical, endovascular, and medical (nonsurgical) treatments are discussed.

Acute Aortic Syndrome

AAS is a life-threatening condition involving the thoracic aorta and comprises a spectrum of entities with a common clinical presentation: acute intense chest pain and hypertension. Chronic hypertension is the most significant risk factor for developing AAS. Other risk factors include smoking, atherosclerosis, diabetes, pregnancy, connective tissue disorders (Marfan, Turner, Ehlers-Danlos, and Loeys-Dietz syndromes), male sex, illicit drug use, and autoimmune conditions (1,7,8).

The subtypes of AAS historically have been thought to share a common inciting event: disruption of the aortic media (1). There is some debate on whether ruptured aortic aneurysm should be included in the spectrum of AAS. Although many authors adhere to the substantial body of literature in excluding it because of its different pathophysiologic mechanism, it is also indisputable that few differences exist clinically. Some authors include traumatic intimal tear or traumatic aortic injury as a subtype of AAS, although the clinical risk factors and traumatic inciting event are different even if the ultimate treatment may be similar (1,8). The Svensson (9) classification of aortic dissection incorporates five variants of aortic wall lesions and includes both incomplete dissection (type III) and traumatic causes (type V) (Table).

Aortic dissection and IMH are divided according to the Stanford classification system, as their location in the thoracic aorta has implications for prognosis and treatment. The thoracic aorta is classically divided into three distinct segments: *(a)* the ascending thoracic aorta, defined as proximal to the brachiocephalic artery, *(b)* the arch, which extends from the brachiocephalic artery to the left subclavian artery, and *(c)* the descending thoracic aorta, which proceeds distal to the left subclavian artery (1). Stanford type A IMH involves the ascending aorta, with or without involvement of the descending aorta. Stanford type B IMH includes all lesions that do not involve the ascending aorta (ie, lesions originating in the arch or the descending thoracic aorta) (Figs 3–5).

IMH represents 6%–20% of AAS cases, with higher incidences in Asian cohorts (1,10,11). Compared with patients with aortic dissection, patients with IMH are more likely to be older, and those with Stanford type A IMH are more likely to have a known aortic aneurysm. There is no difference in mortality in patients with IMH versus those with aortic dissection (11).

On the surface, it may seem simple to separate the subtypes of AAS, but the reality is often more complicated by the spectrum of imaging findings. IMH can progress to aortic dissection because of intimal disruption (7,12,13). IMH

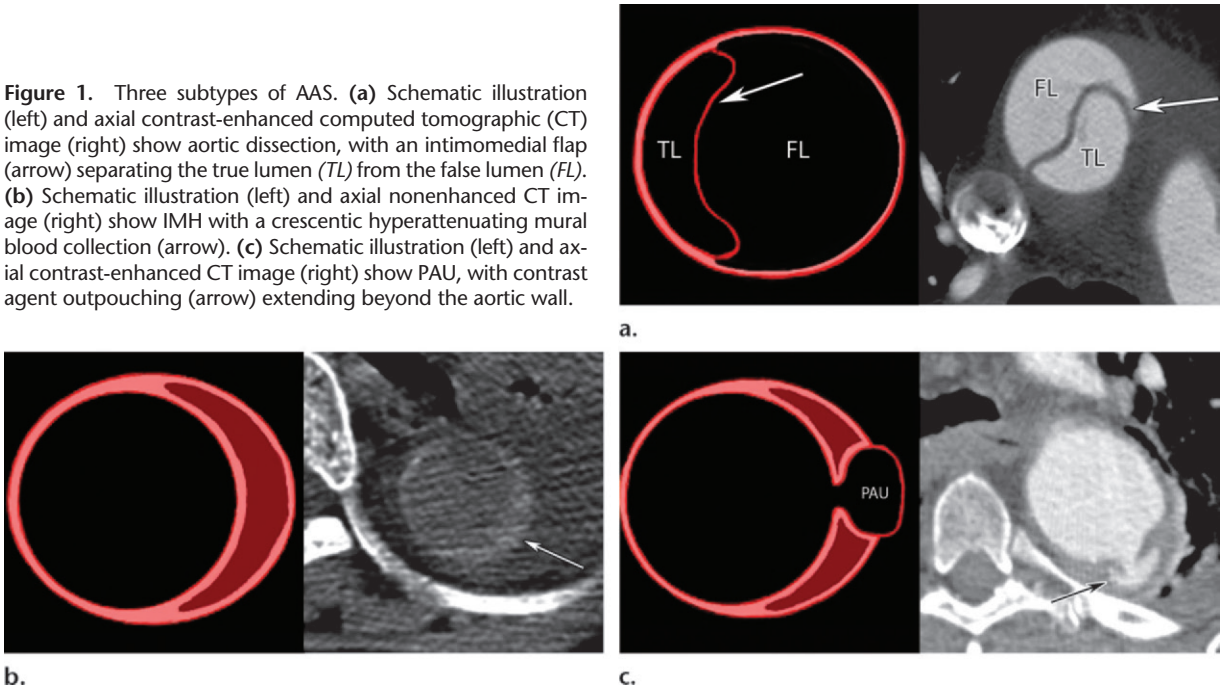


Figure 1. Three subtypes of AAS. **(a)** Schematic illustration (left) and axial contrast-enhanced computed tomographic (CT) image (right) show aortic dissection, with an intimomedial flap (arrow) separating the true lumen *(TL)* from the false lumen *(FL)*. **(b)** Schematic illustration (left) and axial nonenhanced CT image (right) show IMH with a crescentic hyperattenuating mural blood collection (arrow). **(c)** Schematic illustration (left) and axial contrast-enhanced CT image (right) show PAU, with contrast agent outpouching (arrow) extending beyond the aortic wall.

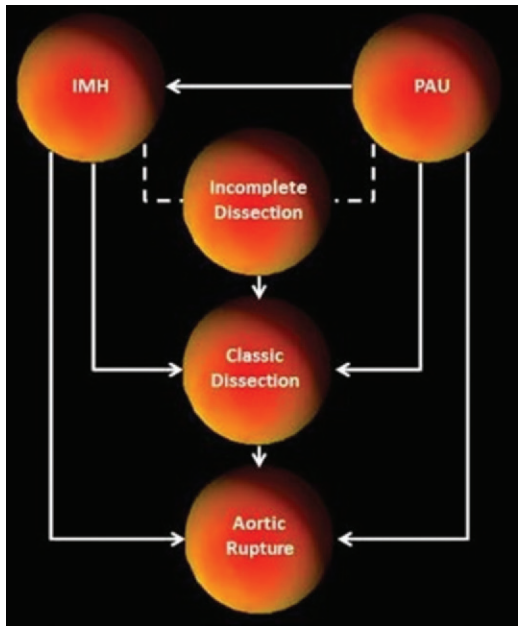


Figure 2. Diagram shows the relationships among the entities grouped under AAS. Note that although classic aortic dissection can occur as a unique entity, it may also result from progression of IMH, PAU, or incomplete dissection. All may eventually lead to aortic rupture.

may also coexist with aortic dissection in different aortic segments as a mixed-type lesion. There are several possible hypotheses to explain these mixed-type lesions: *(a)* a patient has classic aortic dissection with partial segmental false lumen thrombosis, *(b)* a patient has IMH that progresses to communicating aortic dissection in one aortic segment, or *(c)* a patient presents with findings of simultaneous IMH and classic aortic dissection in two different aortic segments (14). PAU often results in a variable amount of IMH. Areas of focal contrast enhancement can occur within the IMH, including intramural blood pool and ulcerlike projection. Intramural blood pool and ulcerlike projection have different etiologies and different appearances, factors that are useful in differentiating these two entities at imaging. However, differentiating PAU with IMH from IMH with ulcerlike projection may be difficult or impossible. An incomplete dissection consists of an intimomedial tear without substantial dissection and is often accompanied by subadventitial hemorrhage that can mimic IMH at imaging, which further complicates matters (6). The imaging features of these entities are discussed in further detail in the "Differential Diagnosis" section.

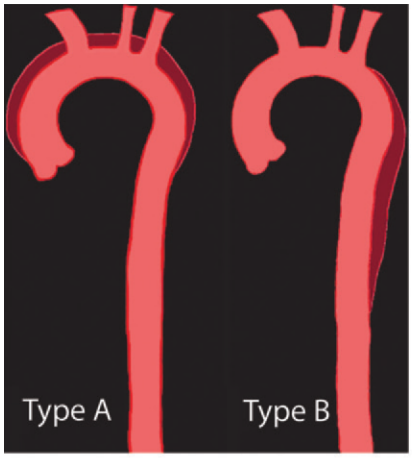
Aortic Wall Anatomy and the Vasa Vasorum

The aorta is made up of three layers: the intima, media, and adventitia. The intima is the innermost layer, composed of a single layer of endothelial cells and separated from the media by the internal elastic lamina. The adventitia is the outermost layer and contains connective tissue and perivascular nerves. The aortic media is the thickest layer, consisting of smooth muscle cells and elastic tissue. The vasa vasorum are small vessels composed of endothelial and smooth muscle cells that supply oxygenated blood to the

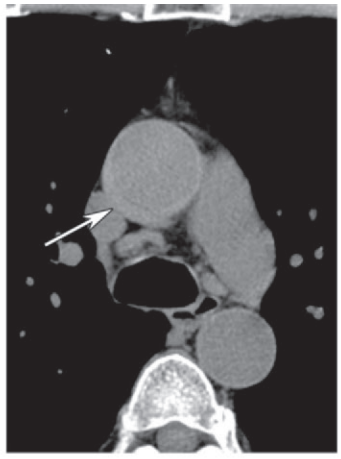
Svensson Classification of Variants of Aortic Dissection

Class	Description
I	Classic aortic dissection with dual lumina and separation of intima from media
II	IMH, no visible intimal tear
III	Limited dissection, intimal tear without hematoma, eccentric bulge
IV	Penetrating atherosclerotic ulcer
V	Iatrogenic or traumatic dissection

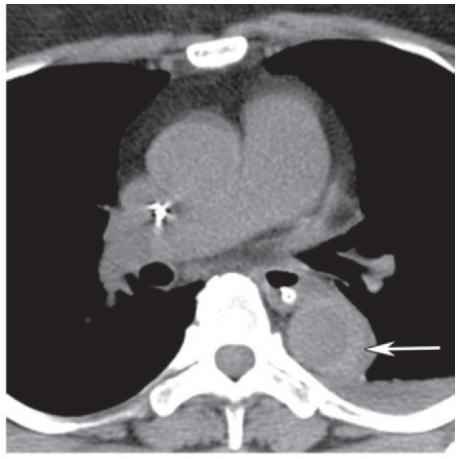
Figures 3–5. **(3)** Schematic illustration shows the Stanford classification of AAS. Stanford type A IMH involves the ascending aorta, with or without involvement of the descending aorta. Stanford type B IMH does not involve the ascending aorta and usually arises distal to the left subclavian artery, although it may also involve the arch. **(4)** CT images of Stanford type A and type B IMH. **(a)** Axial nonenhanced image shows Stanford type A IMH (arrow) involving the ascending thoracic aorta. **(b)** Axial nonenhanced image shows Stanford type B IMH (arrow) involving the descending thoracic aorta. **(5)** Progression of Stanford type A IMH in a 75-year-old woman. **(a)** Axial nonenhanced CT image shows Stanford type A acute IMH, which is associated with increased risk for progression. **(b)** Axial contrast-enhanced CT image obtained 5 days later shows progression to aortic dissection (arrow) and hemopericardium (*).



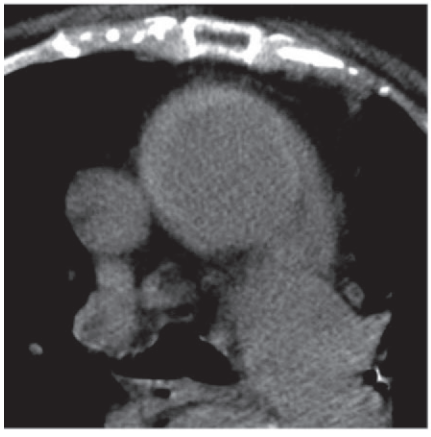
3.



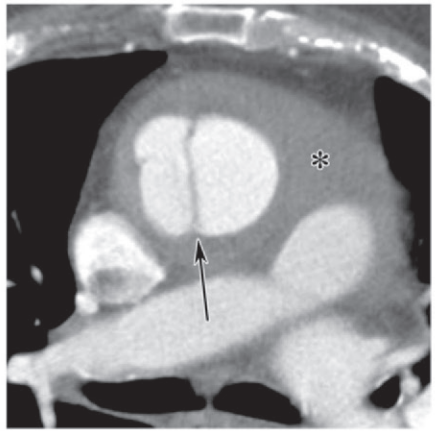
4a.



4b.



5a.



5b.

outer walls of large arteries and veins. The vasa vasorum externa penetrate the aortic wall from the outer adventitia to supply the outer media (Fig 6). The vasa vasorum externa arise from the coronary and brachiocephalic arteries in the ascending aorta, the intercostal arteries in the descending aorta, and the lumbar and mesenteric arteries in the abdominal aorta (15).

IMH: Pathophysiology and Controversies

Long-standing hypertension incites a sequence of histologic alterations in the aortic wall: smooth muscle hypertrophy and hyperplasia, vasa vasorum constriction and occlusion, and, consequently, an ischemic and stiffened outer media (15,16). The inner media continues to receive diffused nutrients

from the aortic lumen and maintains normal elasticity. This elasticity differential results in increased shear stress at the interface of the inner and outer media, which may lead to a medial tear and IMH or aortic dissection (16). This has been confirmed by analysis of pathologic specimens from patients with aortic dissection, in which tears were observed in the outer third of the media (17).

IMH has historically been differentiated from aortic dissection in that the intimal layer remains intact in IMH, and IMH is believed to originate from ruptured vasa vasorum in the aortic media. However, there are increasing reports of small intimomedial tears not apparent at preoperative imaging that were found at surgery in a variable percentage of IMH cases, prompting discussion about the importance of the "microintimal tear" and whether it is the true inciting event rather than vasa vasorum rupture (2,3,5). An alternative explanation is that these cases labeled IMH were actually cases of thrombosed or noncommunicating aortic dissection that were erroneously diagnosed as IMH at imaging because of lack of a visible intimomedial tear. The improved spatial resolution of thin-section multidetector CT allows prospective identification of some intimomedial tears in patients with IMH. Investigations of type A IMH have identified intimomedial defects in 58%–73% of patients, although one study included patients with coexistent type B aortic dissection, and not all studies differentiated small intimomedial defects from a larger ulcerlike projection or PAU (2,3,5). It is possible that even more of these patients may have had intimomedial defects that were not discovered surgically because there was not enough clinical suspicion for distal intimomedial defects to prompt further exploration at surgery, particularly in cases of IMH isolated to the ascending aorta (2).

Perhaps a better question is which comes first—vasa vasorum rupture or intimomedial tear. Pathologic specimens from a series of patients with aortic dissection and IMH have demonstrated extravasation of erythrocytes from the vasa vasorum, even in cases where no intimomedial tear was identified (17). Patients with aortic dissection and those with IMH in this series had media separation that occurred alongside the pathway of the vasa vasorum; thus, it seems likely that vasa vasorum dysfunction from long-standing ischemia is the common denominator in both entities and that vasa vasorum rupture and intimomedial tears represent a secondary phenomenon.

With the advent of improved technology in the last 2 decades, more intimomedial tears are being identified in patients with IMH; thus, a better distinction may be that aortic dissection contains two intimomedial tears—an entry tear from the lumen into the media, and a reentry tear back into the aortic lumen—while IMH with intimomedial tear often has an entry tear only (Fig 7). There are probably anatomic and mechanical reasons for this difference. The location of the media dissection is closer to the adventitial side in IMH when compared to its location in aortic dissection. This may explain why aortic dissection has a reentry tear and why IMH has no reentry tear and is more likely to rupture (18).

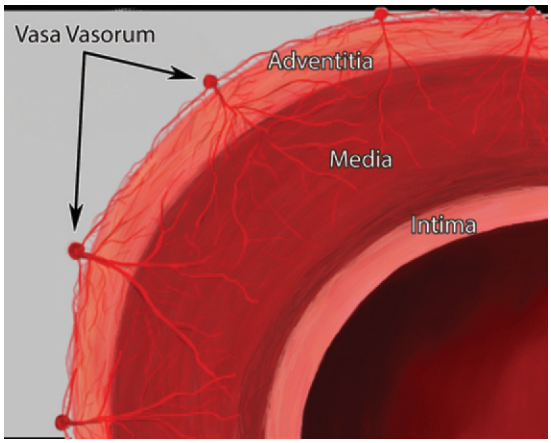


Figure 6. Schematic illustration shows the three layers of the aortic wall: the intima, media, and adventitia. The vasa vasorum (double-headed arrow) are small vessels that supply nutrients to the aorta and penetrate the aortic wall from the outer adventitia.

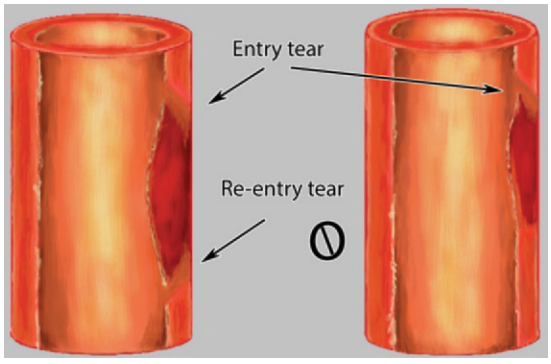


Figure 7. Schematic illustration shows the distinction between aortic dissection and IMH. Aortic dissection (left) usually contains two intimal tears—an entry tear from the aortic lumen into the media, and a reentry tear back into the lumen. IMH (right) may exhibit only an entry tear into the media.

The mean intimomedial tear size is also significantly smaller in patients with IMH (1.8 cm ± 1.0) compared with those with aortic dissection (2.9 cm ± 1.2) (19). On the basis of documentation of intimomedial tears in a number of cases, some authors now consider IMH a subtype of aortic dissection and propose use of the term *thrombosed-type aortic dissection* or *aortic dissection with a closed and thrombosed false lumen* (4,19,20).

It is easier to separate IMH from PAU on the basis of pathophysiology. PAU involves ulceration of

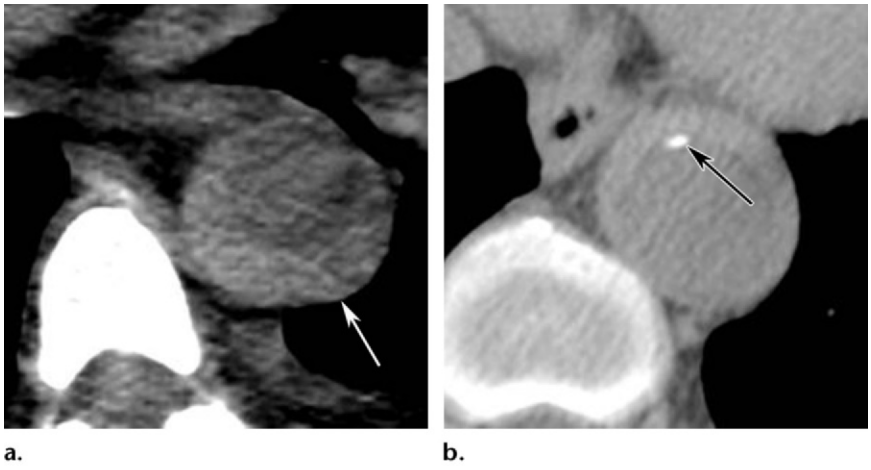


Figure 8. CT features of acute IMH in three patients. **(a)** Axial nonenhanced CT image in a 57-year-old man shows hyperattenuating crescentic thickening of the aortic wall (arrow). **(b)** Axial nonenhanced CT image in a 73-year-old woman shows displacement of intimal calcifications (arrow). **(c)** Sagittal contrast-enhanced CT image in a 77-year-old man shows decreased diameter of the aortic lumen.

an aortic plaque through the internal elastic lamina into the media, with secondary variable amounts of medial hematoma. The pathophysiology of incomplete dissection is also distinct and involves an intimomedial tear without intramural separation. IMH is absent in an incomplete dissection, which is frequently associated with subadventitial hemorrhage (hemorrhage between the adventitia and the media) (6). This is a histologic difference that is difficult to distinguish at imaging because both intramural and subadventitial hemorrhage will appear as mural thickening surrounding the aorta. The intimomedial tears in incomplete dissection are typically located along the posterior ascending aorta just above the left coronary artery ostium, which likely relates to aortic wall stress patterns (21).

Imaging Protocol

Although magnetic resonance (MR) imaging, transesophageal echocardiography, or conventional angiography can be performed, CT is the preferred modality for evaluating AAS in emergency departments and hospital inpatient settings because CT is easily accessible, has a rapid scan time, and is noninvasive. A typical protocol begins with nonenhanced CT through the thorax. This is followed by contrast-enhanced CT of the entire aorta, from the proximal arch vessels to the common iliac arteries. Thin-section image acquisition (<1.25 mm) is performed with 80–100 mL of higher iodine concentrations of contrast material (300–350 mg/mL) injected at rapid infusion rates (2–5 mL/sec), with target opacification of the aorta greater than 150–250 HU achieved with bolus tracking or test bolus techniques (22–24). Prospective electrocardiographic gating is helpful in eliminating pulsation artifact in the ascending aorta, although it is not always necessary or available in practice.

IMH exhibits crescentic or circular aortic wall hyperattenuation that is best visualized at nonenhanced CT and may demonstrate displacement of intimal calcifications in a curvilinear distribution (Fig 8). This crescentic hyperattenuation is better visualized with thicker 5-mm sections than with thin sections, as there is more volume averaging and less noise with thicker slabs. Although most radiologists use thin-section acquisition for aortic protocol CT, reformatted images with a 5-mm section thickness are helpful and should be obtained in the nonenhanced portion of the examination. Use of a narrow window (width, 200 HU; level, 40 HU) will also aid in hematoma detection on nonenhanced images. Subacute IMH may be isoattenuating relative to the blood pool at nonenhanced CT. Administration of contrast agent will demonstrate a decreased diameter of the aortic lumen without diffuse enhancement of the hematoma. This decreased luminal diameter is often better appreciated on sagittal or oblique reformatted images and can be useful in differentiating IMH from other entities such as contained aortic rupture.

Natural History and Prognostic Factors

The natural history of IMH is variable. It may regress, resolve, enlarge, or progress to aneurysm or dissection. Multiple imaging prognostic factors

have been identified that indicate a higher risk for complications or progression. Alerting the primary physician to the presence or absence of these prognostic factors can help ensure that patients are treated appropriately, and identification of these factors becomes more important in type B IMH or at institutions where type A IMH may be initially managed nonsurgically. Whenever IMH is imaged, the following items should be included in the radiology report: Stanford classification; maximum aortic diameter; maximum hematoma thickness; presence or absence of focal contrast enhancement (ie, intramural blood pool or ulcerlike projection); ulcerlike projection diameter and depth; and presence or absence of pleural effusion, pericardial effusion, and periaortic hematoma.

Stanford Classification

The differentiation of IMH into Stanford type A or B has implications for prognosis. Stanford type A IMH results in increased risk for pericardial and/or pleural effusion, aortic dissection, aneurysm formation, and death (Fig 7) (7,25). About 40% of cases of IMH are type A, compared with 72% of aortic dissection cases (11,18).

Maximum Aortic Diameter

Aneurysmal dilatation of the aorta in combination with IMH carries an increased independent risk for adverse events, including expansion, progression to aortic dissection, rupture, incomplete resolution, need for surgery, and death (10,12,26,27). This is related to aortic wall stress and the Laplace law, which states that the stress on a cylinder is directly proportional to its diameter (28). Suggested diameter cutoff values for identifying patients at increased risk can be stratified into Stanford type A or type B IMH. Those with Stanford type A IMH are at higher risk for adverse outcomes when the maximum aortic diameter exceeds 48–55 mm (3,10), while those with type B IMH are at higher risk for adverse outcomes when the maximum aortic diameter exceeds 40–41 mm (2,28,29).

Maximum IMH Thickness

Maximum hematoma thickness predicts adverse outcomes, with increased thickness decreasing the likelihood of complete resorption, as well as increasing the risk for progression, aortic dissection, need for surgery, and death (3,10,12,23,27,30,31). Hematoma thickness is typically based on axial measurements or measurements obtained perpendicular to the longitudinal axis of the aorta lumen, with suggested thickness cutoff values of 10–11 mm (Fig 9) (3,23,31).

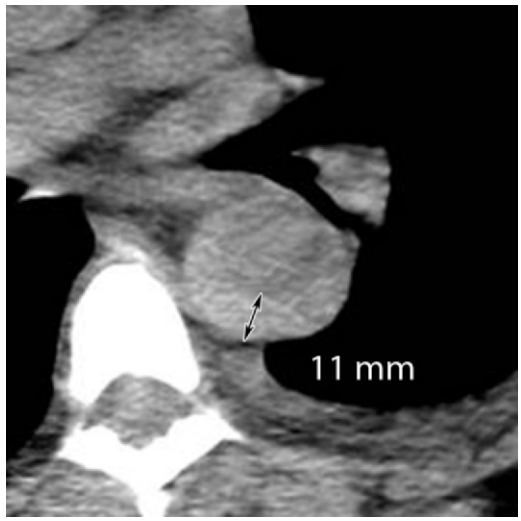


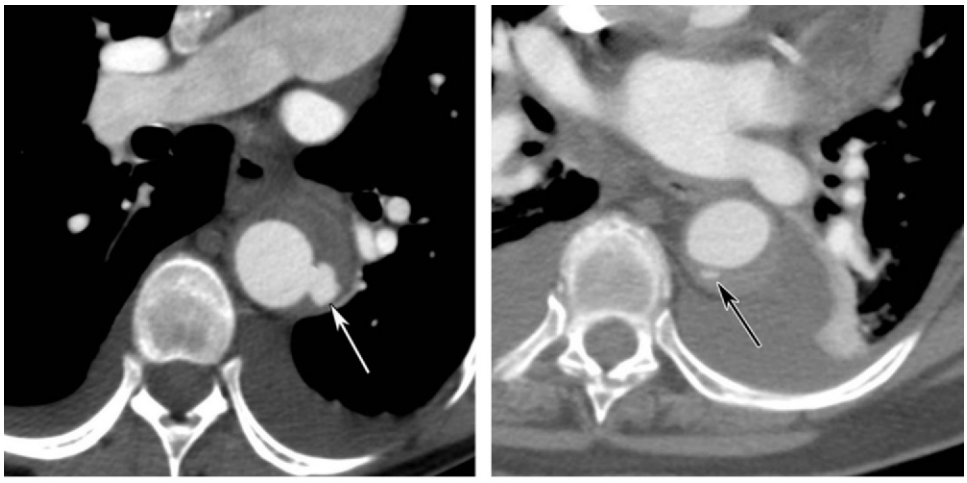
Figure 9. Prognostic factor of maximal IMH thickness in a 57-year-old man. Axial nonenhanced CT image shows an IMH thickness of 11 mm (double-headed arrow). The patient is at increased risk for progression and mortality.

Focal Contrast Enhancement

Focal contrast enhancement within an IMH can be subdivided into two types: ulcerlike projection and intramural blood pool (Fig 10). An ulcerlike projection is a small localized area of contrast enhancement extending from the aortic lumen into the IMH, with a visible communication (ie, broad neck >3 mm) (Fig 11). An intramural blood pool is a similar small localized area of contrast enhancement within the IMH, but it has a very small (<2-mm) or imperceptible communication to the aortic lumen (2,23,25). Patients with a thicker IMH are more likely to develop focal contrast enhancement (2).

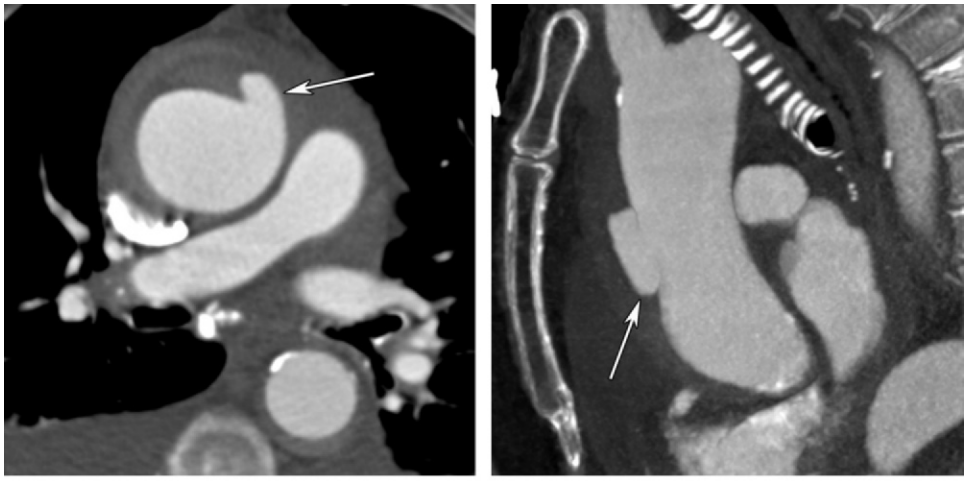
Ulcerlike Projection

Ulcerlike projection is distinguished from PAU in that it typically is not present at initial CT but is identified at follow-up imaging. In addition, an ulcerlike projection may occur in patients with no evidence of atherosclerotic disease. Although many believe that an ulcerlike projection represents a new intimal disruption, others postulate that affected patients may have had an existing intimomedial defect that was not visible at initial imaging because of thrombosis of the false lumen and lack of flow communication between the true and false lumens (4,32). The mean time of development after the acute event can range from 2.4 to 17.8 months (32,33). Ulcerlike projection has a poor prognosis, particularly when it is located in the ascending aorta or aortic arch, and frequently progresses to dissection, aneurysm, or rupture (4,27,32). A larger ulcerlike projection diameter and depth correlate with a higher rate of complications (4,30,34). Suggested threshold



a. b.

Figure 10. Focal contrast enhancement. **(a)** Axial contrast-enhanced CT image in a 66-year-old woman shows an ulcerlike projection (arrow) with localized contrast enhancement extending from the aortic lumen into the IMH and a visible communication. **(b)** Axial contrast-enhanced CT image in an 81-year-old man shows an intramural blood pool (arrow), defined as a small focal area of contrast enhancement in the IMH without a visible communication to the aortic lumen or with a small (<2-mm) connection.



a. b.

Figure 11. CT images in an 89-year-old woman with Stanford type A IMH complicated by an ulcerlike projection. Axial contrast-enhanced **(a)** and sagittal oblique maximum intensity projection (MIP) **(b)** images show an ulcerlike projection (arrow), a poor prognostic indicator with increased risk for complications.

values for identifying patients at greatest risk include an ulcerlike projection diameter of 10–20 mm and depth of 5–10 mm.

Intramural Blood Pool

Intramural blood pool is differentiated from ulcerlike projection in that there is either no visible communication with the aortic lumen or a very small connection. An intramural blood pool is more likely to occur in the descending aorta and is sometimes referred to as an aortic branch artery tear or an aortic branch artery pseudoaneurysm because there often is a visible connection with an intercostal, lumbar, or bronchial artery (Fig 12) (23,35,36). An intramural blood pool may be visible as a string of contrast agent poolings on coronal or sagittal reconstructed images, a finding referred to as the Chinese ring-sword sign (36). Continued improvements in thin-section CT may allow increased visibility of the connection to the branch artery as well as to the aortic lumen. Studies of intramural blood pool are limited, and the prognostic significance is uncertain. On the basis of current available literature, intramural blood pool does not appear to carry increased risk for IMH progression, need for surgery, or mortality but does have higher risk for incomplete hematoma resorption (23,30). Larger intramural blood pools and those with a visible connection to a

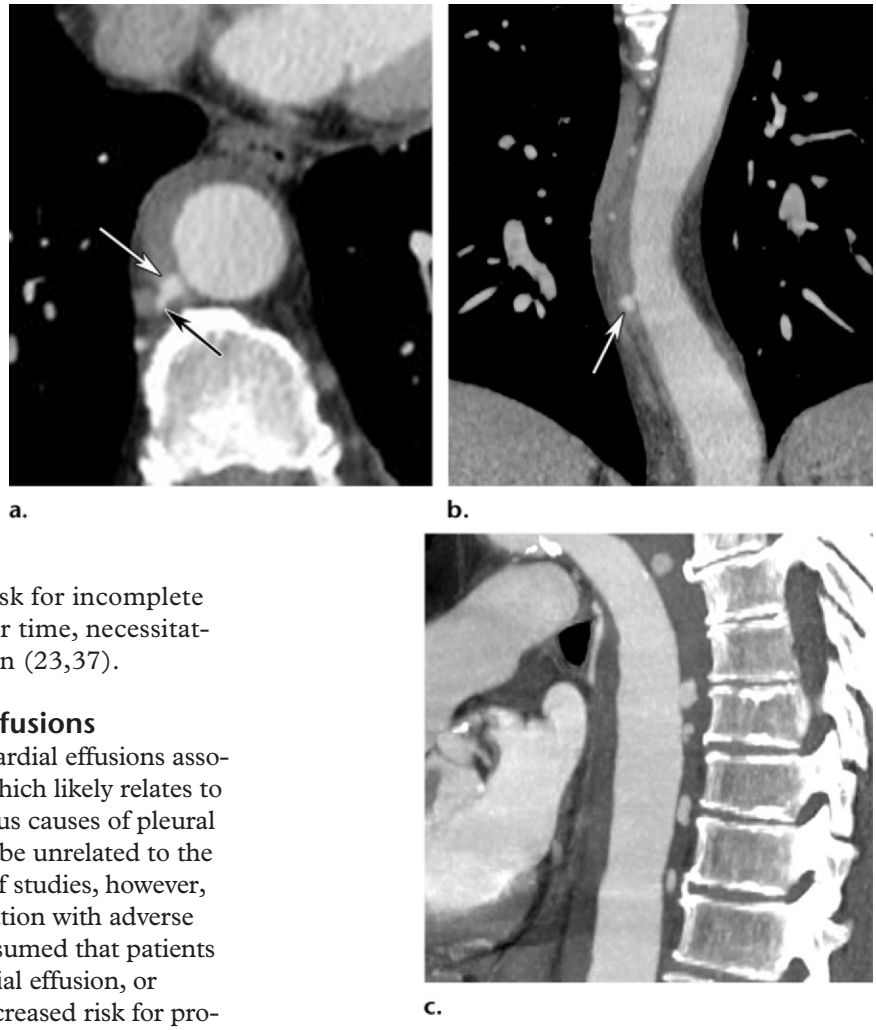


Figure 12. Intramural blood pool in a 62-year-old man with IMH. **(a, b)** Axial **(a)** and coronal **(b)** contrast-enhanced CT images show an intramural blood pool (white arrow) with absent or a tiny communication with the true aortic lumen. Note the connection with a right posterior intercostal artery (black arrow in **a**). A limited number of studies have shown no known adverse risk factors associated with intramural blood pool other than a higher rate of incomplete hematoma resorption. **(c)** Sagittal contrast-enhanced MIP CT image shows a string of contrast material poolings, a finding referred to as the "Chinese ring-sword sign."

branch artery are at higher risk for incomplete resorption and may grow over time, necessitating endovascular embolization (23,37).

Pleural and Pericardial Effusions

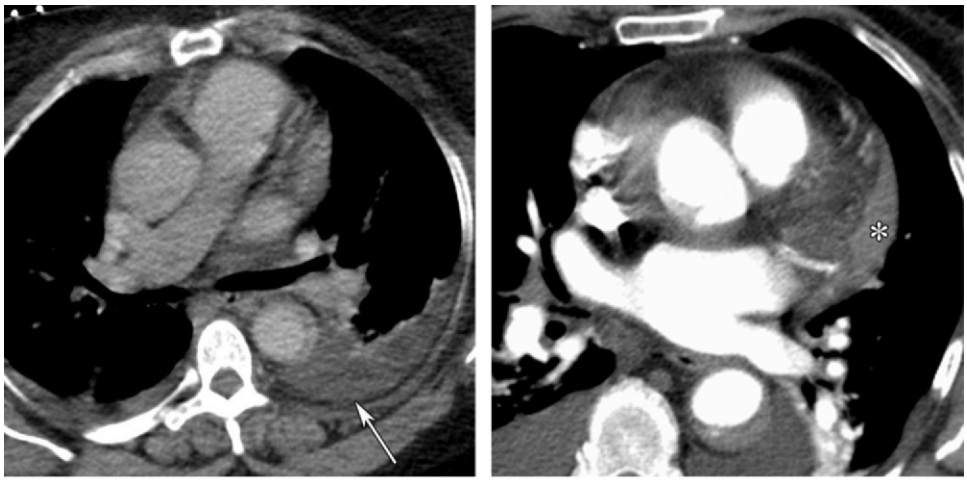
The data on pleural and pericardial effusions associated with IMH are mixed, which likely relates to the fact that there are numerous causes of pleural and pericardial fluid that may be unrelated to the patient's IMH. The majority of studies, however, tend to show a positive correlation with adverse outcomes, and it should be assumed that patients with pleural effusion, pericardial effusion, or periaortic hematoma are at increased risk for progression and mortality (Fig 13) (2,23,34,38,39). Only a few investigations failed to demonstrate a statistically significant association of pleural and pericardial effusions with poor outcomes (3,31).

Treatment and Imaging Follow-up

Treatment strategies for IMH are similar to those for aortic dissection and depend on whether the hematoma is Stanford type A or type B. Type B IMH is initially treated nonsurgically. Treatment is aimed at reducing aortic wall stress, primarily through the use of β blockers such as labetalol that affect heart rate, ventricle contractility, and systemic blood pressure (8,40). Surveillance imaging in patients with nonsurgically treated type B IMH or surgically treated IMH is similar to that in patients with classic aortic dissection: CT or MR imaging performed before discharge and at 1, 3, 6, and 12 months after the acute event and, if stable, annually thereafter to detect potential complications (40). Surgical or endovascular intervention may be required in the event of complications (eg, enlargement of aortic diameter, development of an ulcerlike projection, or hematoma progression to frank dissection) (Fig 14) (41). Patients with large (>10-mm) or enlarging intramural blood pools may also require endovascular embolization with coils and/or an Amplatzer vascular plug (St Jude Medical, St Paul, Minn) (37).

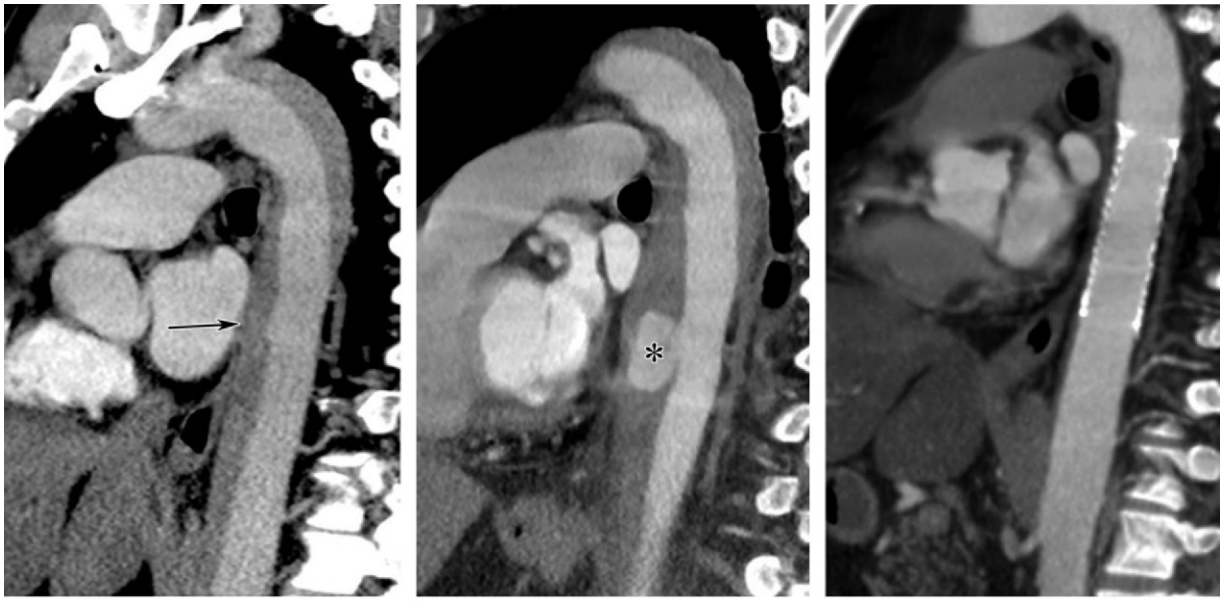
Historically, type A IMH was considered a surgical emergency, according to evidence that showed a low mortality rate with early surgical treatment as opposed to mortality rates of 40%–80% with nonsurgical treatment (7,11). Surgical options for patients with IMH (including those with complications such as an ulcerlike projection) include open repair and hybrid combinations of open repair and thoracic endovascular aortic repair (TEVAR) (1,41). Suggested surveillance imaging after stent-graft placement includes CT or MR imaging before discharge and at 1, 2, 6, and 12 months after the acute event and, if stable, annually thereafter to detect possible complications (40).

Increasingly, there are reports of Asian cohorts (eg, Japan, South Korea) having successful initial nonsurgical treatment, with timed (delayed) surgery reserved for patients who develop



a. b.

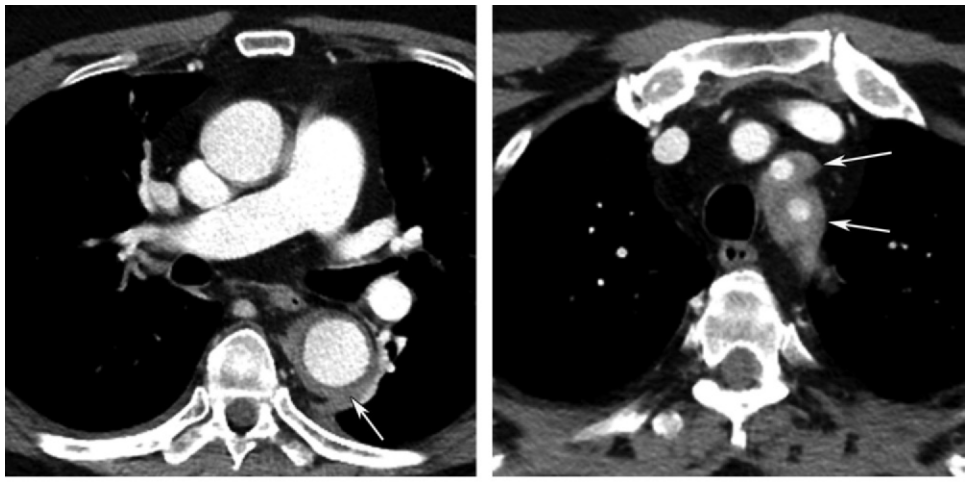
Figure 13. Pleural and pericardial effusions. **(a)** Axial contrast-enhanced CT image in a 77-year-old woman shows Stanford type B IMH, with greater pleural effusions on the left (arrow) than on the right. **(b)** Axial contrast-enhanced CT image in an 81-year-old man shows Stanford type A IMH with pericardial hemorrhage (*) and bilateral pleural effusions. Because there are many causes of pleural and pericardial fluid that may be unrelated to IMH, the value of this particular finding is indeterminate. Most studies show a trend toward adverse outcome, with patients at increased risk for progression and mortality.



a. b. c.

Figure 14. Stanford type B IMH with development of an ulcerlike projection in a 49-year-old man. **(a)** Sagittal contrast-enhanced CT image obtained at presentation shows Stanford type B acute IMH (arrow). **(b)** Sagittal contrast-enhanced follow-up CT image shows development of an ulcerlike projection (*), a complication that portends a poor prognosis. **(c)** Sagittal contrast-enhanced CT image shows the endovascular stent-graft used for treatment.

complications. This strategy requires aggressive radiologic surveillance, including multiple imaging studies performed within the 1st week and then typically weekly for 2–4 weeks, monthly during the next 3–6 months, and every 6–12 months or sooner if complications are suspected clinically (27). A recent study of 101 patients with type A IMH that used the approach of initial nonsurgical treatment for stable patients (with the possibility of later surgery) found no difference in mortality when compared with patients with IMH who were initially treated surgically, and no difference in mortality when compared with patients with surgically treated type A aortic dissection. Emergent surgery was performed in 16% of patients with IMH, and 29% ultimately required surgery (10). Similar smaller studies have also shown good survival with initial nonsurgical treatment of type A IMH. However, 19%–45% of affected patients



a. b.

Figure 15. Takayasu arteritis in a 50-year-old man. Axial contrast-enhanced CT images (**b** obtained at a higher level than **a**) show uniform thickening of the vessel walls (arrows). Enhancement after contrast agent administration is an imaging feature that helps distinguish Takayasu arteritis from IMH.

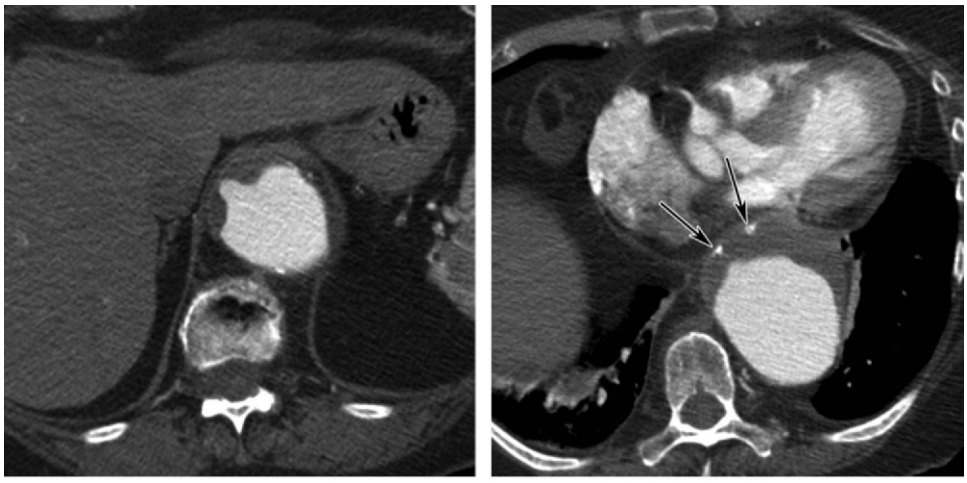
may eventually require surgery because of progression or complications (27,42). A larger clinical review of 328 patients with type A IMH showed an almost 43% higher mortality rate for those who underwent nonsurgical treatment compared with those who underwent surgery (14.4% vs 10.1%), although the finding was not statistically significant (18).

Ultimately, it may be inappropriate to extrapolate these strategies for use in other patient populations because the prevalence of type A IMH in Asian populations is much higher than what is reported in the International Registry of Aortic Dissection (28.3% versus 3.6%, respectively) (10). Some U.S. surgeons advocate a middle-ground approach of urgent rather than emergent surgery, with the hypothesis that the delay allows inflammation to subside and makes surgery easier. This approach involves initial nonsurgical treatment of hemodynamically stable patients with type A IMH, with delayed expectant surgical repair within 3–4 days of presentation, and has been shown to have no difference in outcomes between those undergoing immediate versus delayed surgery (5). This approach also allows stable patients to undergo initial nonsurgical treatment followed by transfer to a larger hospital center with more experienced surgeons (42).

Differential Diagnosis

The main differential diagnosis of IMH is aortitis (eg, Takayasu arteritis or giant cell arteritis), which can also cause uniform thickening of the aortic wall. Aortitis may result in hyperattenuating wall thickening at nonenhanced CT; the wall typically shows enhancement after contrast agent administration and may be associated with transmural calcification (Fig 15) (43,44). In addition, fluorodeoxyglucose (FDG) positron emission tomography (PET) will show diffuse FDG accumulation in areas of active inflammation in the aortic wall in the setting of arteritis and can be used to monitor treatment response (45,46). Intraluminal thrombus can also simulate IMH but will not appear hyperattenuating at nonenhanced CT. When calcification does occur with thrombus, it tends to be irregular and thick, versus the thin curvilinear intimal calcification displacement that may be seen with IMH. Intraluminal thrombus typically has an irregular internal contour that is unlike the smooth contour seen with IMH, and it usually occurs in aneurysmal segments of the aorta (Fig 16). In addition, the aortic lumen is mildly narrowed in IMH, with a clear transition seen between the involved and normal aorta, a feature not typically seen in aortitis and thrombus. The clinical presentation should help distinguish both entities from IMH. Typically, neither patients with aortitis nor those with intraluminal thrombus should present with acute chest pain. In cases where the clinical presentation is not helpful, other ancillary findings, including patient demographics, laboratory evaluation (eg, erythrocyte sedimentation rate, C-reactive protein level), and rheumatologic workup may be necessary to differentiate the possible causes of chest pain.

Contained rupture of an abdominal or thoracic aortic aneurysm may mimic acute IMH because both entities demonstrate crescentic high attenuation. Whereas IMH begins in the aortic media, the high attenuation in contained rupture begins in the intraluminal thrombus and may



a. b.

Figure 16. Intraluminal thrombus in an 85-year-old woman. Axial contrast-enhanced CT images obtained at different levels show the irregular contour of an intraluminal thrombus in **a** and absence of displacement of intimal calcification (arrows in **b**), findings that help distinguish intraluminal thrombus from IMH. Intraluminal thrombus typically occurs in association with a thoracic or abdominal aortic aneurysm.

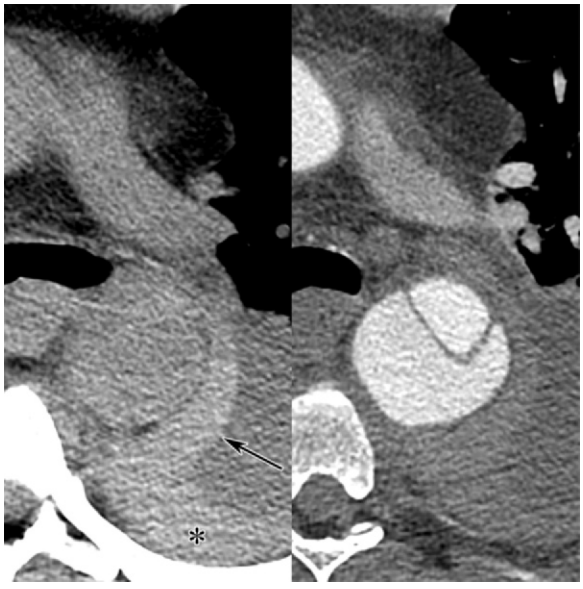


Figure 17. Contained aortic rupture in a 63-year-old man. Axial nonenhanced (left) and contrast-enhanced (right) CT images show the hyperattenuating crescent sign (arrow) of periaortic hematoma, a finding seen in a contained aortic rupture in the setting of aortic dissection. Note extension of the hematoma into the adjacent pleural space (*).

later penetrate the wall (47). Contained rupture is focal and often occurs as a complication of a preexisting aortic aneurysm (Fig 17). This contrasts with IMH, which is usually elongated with a decreased luminal diameter. Although aneurysm can be seen with IMH, it tends to occur in the subacute or chronic stage, which should not demonstrate a hyperattenuating hematoma. Patients with impending rupture are usually hemodynamically unstable.

Aortic intimal sarcoma is another differential consideration, although this entity tends to exhibit a lobulated contour, may extend beyond the confines of the aortic wall, and should enhance after administration of intravenous contrast material, a finding that is often better appreciated at MR imaging. Patients with aortic intimal sarcoma have a different clinical presentation than those with IMH, most commonly organ or extremity ischemia resulting from tumor embolization (48).

Incomplete dissection is characterized by an intimomedial tear along the posterior ascending aorta above the left coronary ostium and is accompanied by an adjacent protrusion of contrast material without extravasation. There may be mural thickening surrounding the adjacent aorta that mimics IMH, but the hemorrhage is located between the adventitia and the media (subadventitial) rather than between the inner and outer media as with IMH (Fig 18). Differentiation from IMH may be difficult because the clinical symptoms and risk factors are similar, with key features of incomplete dissection including the characteristic location near the left coronary ostium, an eccentric bulge near the tear, and frequent concomitant ascending aorta dilatation (6,21). Thus, if something resembling IMH is visualized directly above the left coronary artery, incomplete dissection must be suspected or considered.

It may be more difficult to distinguish IMH from PAU because *(a)* the clinical presentation

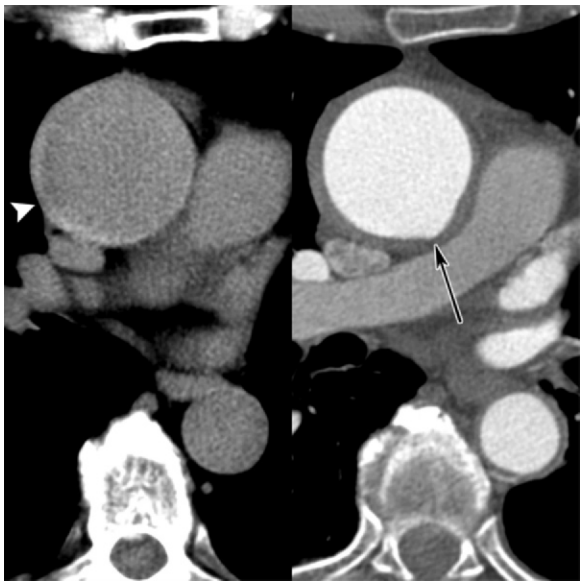


Figure 18. Incomplete aortic dissection in a 75-year-old woman. Axial nonenhanced (left) and contrast-enhanced (right) CT images show the hyperattenuating crescent sign of subadventitial hemorrhage (arrowhead) and a small protrusion of contrast material (arrow) along the posterior ascending aorta above the left coronary ostium, findings seen in incomplete aortic dissection. The ascending aorta is also dilated.

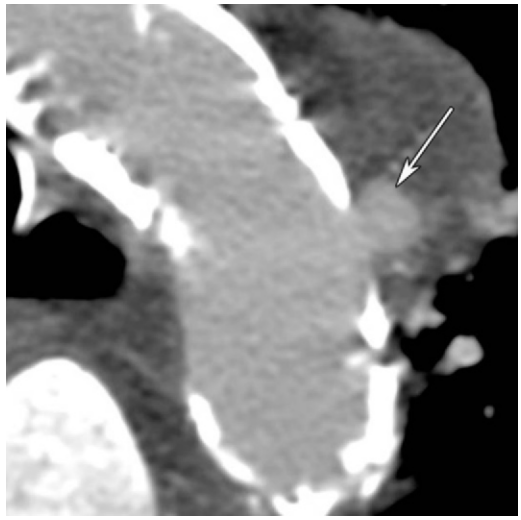


Figure 19. PAU in a 78-year-old man. Axial contrast-enhanced CT image shows PAU (arrow), which is ulceration of an aortic plaque through the internal elastic lamina into the media. Unlike an ulcerlike projection, there is usually adjacent calcified atherosclerotic disease. PAU is frequently associated with IMH, but in this example no IMH is seen.

of affected patients may be identical, *(b)* PAU can cause IMH, and *(c)* IMH can develop an ulcerlike projection. PAUs involve disruption of the internal elastic lamina with extension into the media and a collection of contrast material extending beyond the expected wall of the aorta, originate within atherosclerotic segments of the aorta, and are found in the descending thoracic aorta in more than 90% of cases (Fig 19) (1,22). When PAU occurs with intramural hemorrhage, the hematoma is usually localized, although it may involve a long length of aorta. PAUs are seen at initial CT, while an ulcerlike projection is thought to represent a new intimal disruption seen at follow-up imaging, is typically not associated with underlying atherosclerotic plaque, and is more commonly located in the ascending aorta (23,25). Ultimately, PAU may be indistinguishable from an ulcerlike projection without pathologic evaluation, which leads some to lump both conditions together under the broad term *ulcer-like projection*. Pragmatically, treatment is similar for both entities and typically involves surgical or endovascular treatment. Given that the pathophysiology of IMH and PAU is different, it is possible that as more research is performed, their treatments may evolve differently.

STRUCTURED CT REPORT TEMPLATE FOR
AORTIC INTRAMURAL HEMATOMA

Date of Examination: _____
Indication: _____
Comparison: _____

FINDINGS:
Intramural hematoma present: Y / N
Chronicity: acute / subacute / chronic
Stanford classification: A / B
Maximum aortic diameter:
    If Stanford A (> 50 mm / < 50 mm): _____
    If Stanford B (> 40 mm / < 40 mm): _____
Maximal hematoma thickness (> 11 mm): _____
Focal contrast enhancement:
    Intramural blood pool: present / absent
    Ulcer-like projection: present / absent
        Max depth (> 5-10 mm): _____
        Max diameter (> 10-20 mm):_____
Pleural effusion:
    Right: none / small / moderate / large
    Left: none / small / moderate / large
Pericardial effusion: none / small / moderate / large
Periaortic hematoma: present / absent
IMPRESSION: _____

Figure 20. Example of a standard CT report for IMH.

Conclusion

The pathogenesis of IMH remains controversial, with increasing evidence suggesting that many IMHs are associated with microintimal tears. Imaging findings that affect prognosis should be reported and include Stanford type, maximal aortic diameter, IMH thickness, and presence of an ulcerlike projection. Figure 20 provides an ex-

ample of a suggested standard imaging report for IMH. Conventional treatment of IMH is similar to that of aortic dissection: surgery for Stanford type A IMH and nonsurgical treatment for Stanford type B IMH.

***Disclosures of Conflicts of Interest.*—C.M.W.** *Activities related to the present article:* disclosed no relevant relationships. *Activities not related to the present article:* royalties from Elsevier and Amirsys. *Other activities:* disclosed no relevant relationships. **M.L.R.d.C.** *Activities related to the present article:* disclosed no relevant relationships. *Activities not related to the present article:* royalties from Amirsys, Thieme, and the American Registry of Pathology. *Other activities:* disclosed no relevant relationships. **S.M.J.** *Activities related to the present article:* disclosed no relevant relationships. *Activities not related to the present article:* royalties from Amirsys and Oxford University Press. *Other activities:* disclosed no relevant relationships.

References

1. Nienaber CA, Powell JT. Management of acute aortic syndromes. Eur Heart J 2012;33(1):26–35.
2. Park KH, Lim C, Choi JH, et al. Prevalence of aortic intimal defect in surgically treated acute type A intramural hematoma. Ann Thorac Surg 2008;86(5):1494–1500.
3. Sawaki S, Hirate Y, Ashida S, Takanohashi A, Yagami K, Usui M. Clinical outcomes of medical treatment of acute type A intramural hematoma. Asian Cardiovasc Thorac Ann 2010;18(4):354–359.
4. Kitai T, Kaji S, Yamamuro A, et al. Detection of intimal defect by 64-row multidetector computed tomography in patients with acute aortic intramural hematoma. Circulation 2011;124(11 suppl):S174–S178.
5. Estrera AL, Sandhu HK, Leake SS, et al. Early and late outcomes of acute type A aortic dissection with intramural hematoma. J Thorac Cardiovasc Surg 2015;149(1):137–142.
6. Vilacosta I, Aragoncillo P, Cañadas V, Román JA, Ferreirós J, Rodríguez E. Acute aortic syndrome: a new look at an old conundrum. Postgrad Med J 2010;86(1011):52–61.
7. Nienaber CA, von Kodolitsch Y, Petersen B, et al. Intramural hemorrhage of the thoracic aorta: diagnostic and therapeutic implications. Circulation 1995;92(6):1465–1472.
8. Carpenter SW, Kodolitsch YV, Debus ES, et al. Acute aortic syndromes: definition, prognosis and treatment options. J Cardiovasc Surg (Torino) 2014;55(2 suppl 1):133–144.
9. Svensson LG, Labib SB, Eisenhauer AC, Butterly JR. Intimal tear without hematoma: an important variant of aortic dissection that can elude current imaging techniques. Circulation 1999;99(10):1331–1336.
10. Song JK, Yim JH, Ahn JM, et al. Outcomes of patients with acute type A aortic intramural hematoma. Circulation 2009;120(21):2046–2052.
11. Harris KM, Braverman AC, Eagle KA, et al. Acute aortic intramural hematoma: an analysis from the International Registry of Acute Aortic Dissection. Circulation 2012;126(11 suppl 1):S91–S96.
12. Evangelista A, Dominguez R, Sebastia C, et al. Long-term follow-up of aortic intramural hematoma: predictors of outcome. Circulation 2003;108(5):583–589.
13. von Kodolitsch Y, Csösz SK, Koschyk DH, et al. Intramural hematoma of the aorta: predictors of progression to dissection and rupture. Circulation 2003;107(8):1158–1163.
14. Vilacosta I, Ferreirós J, Bustos A, Román JA, Aragoncillo P. Intramural aortic hematoma and aortic ulcers, physiopathology and natural history. In: Rousseau H, Verhoye JP, Heautot JF, eds. Thoracic aortic diseases. Berlin, Germany: Springer, 2006; 277–288.
15. Tanaka H, Zaima N, Sasaki T, et al. Adventitial vasa vasorum arteriosclerosis in abdominal aortic aneurysm. PLoS One 2013;8(2):e57398. doi:10.1371/journal.pone.0057398.
16. Pereira AH. Rupture of vasa vasorum and intramural hematoma of the aorta: a changing paradigm. J Vasc Bras 2010;9(2):57–60.
17. Osada H, Kyogoku M, Ishidou M, Morishima M, Nakajima H. Aortic dissection in the outer third of the media: what is the role of the vasa vasorum in the triggering process? Eur J Cardiothorac Surg 2013;43(3):e82–e88. Published December 31, 2012.
18. Kan CB, Chang RY, Chang JP. Optimal initial treatment and clinical outcome of type A aortic intramural hematoma: a clinical review. Eur J Cardiothorac Surg 2008;33(6):1002–1006.
19. Uchida K, Imoto K, Karube N, et al. Intramural haematoma should be referred to as thrombosed-type aortic dissection. Eur J Cardiothorac Surg 2013;44(2):366–369; discussion 369.
20. Song JK. Update in acute aortic syndrome: intramural hematoma and incomplete dissection as new disease entities. J Cardiol 2014;64(3):153–161.
21. Chirillo F, Salvador L, Bacchion F, Grisolia EF, Valfrè C, Olivari Z. Clinical and anatomical characteristics of subtle-discrete dissection of the ascending aorta. Am J Cardiol 2007;100(8):1314–1319.
22. Litmanovich D, Bankier AA, Cantin L, Raptopoulos V, Boiselle PM. CT and MRI in diseases of the aorta. AJR Am J Roentgenol 2009;193(4):928–940.
23. Wu MT, Wang YC, Huang YL, et al. Intramural blood pools accompanying aortic intramural hematoma: CT appearance and natural course. Radiology 2011;258(3):705–713.
24. Baliga RR, Nienaber CA, Bossone E, et al. The role of imaging in aortic dissection and related syndromes. JACC Cardiovasc Imaging 2014;7(4):406–424.
25. Sueyoshi E, Matsuoka Y, Sakamoto I, Uetani M, Hayashi K, Narimatsu M. Fate of intramural hematoma of the aorta: CT evaluation. J Comput Assist Tomogr 1997;21(6):931–938.
26. Park GM, Ahn JM, Kim DH, et al. Distal aortic intramural hematoma: clinical importance of focal contrast enhancement on CT images. Radiology 2011;259(1):100–108.
27. 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(3):435–440.
28. Sueyoshi E, Sakamoto I, Uetani M, Matsuoka Y. CT analysis of the growth rate of aortic diameter affected by acute type B intramural hematoma. AJR Am J Roentgenol 2006;186(6 suppl 2):S414–S420.
29. Sueyoshi E, Imada T, Sakamoto I, Matsuoka Y, Hayashi K. Analysis of predictive factors for progression of type B aortic intramural hematoma with computed tomography. J Vasc Surg 2002;35(6):1179–1183.
30. Schlatter T, Auriol J, Marcheix B, et al. Type B intramural hematoma of the aorta: evolution and prognostic value of intimal erosion. J Vasc Interv Radiol 2011;22(4):533–541.
31. Song JM, Kim HS, Song JK, et al. Usefulness of the initial noninvasive imaging study to predict the adverse outcomes in the medical treatment of acute type A aortic intramural hematoma. Circulation 2003;108(suppl 1):II324–II328.
32. Sueyoshi E, Matsuoka Y, Imada T, Okimoto T, Sakamoto I, Hayashi K. New development of an ulcerlike projection in aortic intramural hematoma: CT evaluation. Radiology 2002;224(2):536–541.
33. Bosma MS, Quint LE, Williams DM, Patel HJ, Jiang Q, Myles JD. Ulcerlike projections developing in noncommunicating aortic dissections: CT findings and natural history. AJR Am J Roentgenol 2009;193(3):895–905.
34. Ganaha F, Miller DC, Sugimoto K, et al. Prognosis of aortic intramural hematoma with and without penetrating atherosclerotic ulcer: a clinical and radiological analysis. Circulation 2002;106(3):342–348.
35. Williams DM, Cronin P, Dasika N, et al. Aortic branch artery pseudoaneurysms accompanying aortic dissection. I. Pseudoaneurysm anatomy. J Vasc Interv Radiol 2006;17(5):765–771.
36. Wu MT, Wu TH, Lee D. Multislice computed tomography of aortic intramural hematoma with progressive intercostal artery tears: the Chinese ring-sword sign. Circulation 2005;111(5):e92–e93.
37. Ferro C, Rossi UG, Seitun S, Scarano F, Passerone G, Williams DM. Aortic branch artery pseudoaneurysms associated with intramural hematoma: when and how to do endovascular embolization. Cardiovasc Intervent Radiol 2013;36(2):422–432.

38. Choi SH, Choi SJ, Kim JH, et al. Useful CT findings for predicting the progression of aortic intramural hematoma to overt aortic dissection. J Comput Assist Tomogr 2001;25(2):295–299.
39. Evangelista A, Dominguez R, Sebastia C, et al. Prognostic value of clinical and morphologic findings in short-term evolution of aortic intramural haematoma: therapeutic implications. Eur Heart J 2004;25(1):81–87.
40. Hiratzka LF, Bakris GL, Beckman JA, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with thoracic aortic disease: executive summary. J Am Coll Cardiol 2010;55(14):1509–1544.
41. Sueyoshi E, Onitsuka H, Nagayama H, Sakamoto I, Uetani M. Endovascular repair of aortic dissection and intramural hematoma: indications and serial changes. Springerplus 2014;3:670.
42. Watanabe S, Hanyu M, Arai Y, Nagasawa A. Initial medical treatment for acute type A intramural hematoma and aortic dissection. Ann Thorac Surg 2013;96(6):2142–2146.
43. Zhu FP, Luo S, Wang ZJ, Jin ZY, Zhang LJ, Lu GM. Takayasu arteritis: imaging spectrum at multidetector CT angiography. Br J Radiol 2012;85(1020):e1282–e1292. doi:10.1259/bjr/25536451.
44. Restrepo CS, Ocazionez D, Suri R, Vargas D. Aortitis: imaging spectrum of the infectious and inflammatory conditions of the aorta. RadioGraphics 2011;31(2):435–451.
45. Hayashida T, Sueyoshi E, Sakamoto I, Uetani M, Chiba K. PET features of aortic diseases. AJR Am J Roentgenol 2010;195(1):229–233.
46. Hartlage GR, Palios J, Barron BJ, et al. Multimodality imaging of aortitis. JACC Cardiovasc Imaging 2014;7(6):605–619.
47. Gonsalves CF. The hyperattenuating crescent sign. Radiology 1999;211(1):37–38.
48. Salhab KF, Said SM, Sundt TM 3rd. Pseudocoarctation of the aorta secondary to aortic intimal sarcoma. Ann Thorac Surg 2012;94(1):279–281.