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. Although intimal disruption is not present, the prognosis is similar to that of classic aortic dissection; therefore, early diagnosis is critical. In this review, symptoms and prognosis of aortic IMH are discussed, as well as current diagnostic techniques and therapy.

Key words: aortic dissection; aortic intramural hematoma; transesophageal echocardiography

Abbreviations: IMH = intramural hematoma; IVUS = intravascular ultrasound; TEE = transesophageal echocardiography

Aortic dissection is a life-threatening medical condition, and immediate diagnosis and treatment are crucial. The vast majority of aortic dissections begin with a primary intimal tear. Blood flow enters the media through this tear and propagates distally, creating a false lumen. A less common variant of aortic dissection exists, however, termed aortic intramural hematoma (IMH). In this entity, hemorrhage occurs within the aortic wall in the absence of initial intimal disruption. Although its existence has been validated by surgical and pathologic examination, the lack of an intimal flap has led to underdiagnosis by aortography in the past.1–6 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. Thus, a familiarity with IMH is useful during the evaluation of acute chest pain. Herein we will review the pathogenesis, natural history, diagnosis, and treatment of IMH.

Pathogenesis

The initial event in aortic dissection has long been a point of controversy. It is unclear who first described aortic dissection, but references are found as far back as 1761 in Nicholl’s description of the autopsy of King George II of England.7 The term aortic dissection is often credited to Laennec in 1826.8,9 Laennec and the scholars of his time believed that the intimal tear allowed blood to enter the media and cause distal dissection. Indeed, autopsy examinations have revealed that most dissections have intimal tears and that these tears usually occur where shear forces are greatest.10,11

An alternate theory of pathogenesis evolved in the early 1900s when several authors independently described the existence of IMH (that is, a hemorrhagic dissection of the media without an intimal tear). Of these authors, Krukenberg12 first proposed that a rupture of the vasa vasorum initiated the process of aortic dissection. Gore9 championed this view in the 1950s and suggested that underlying...
medial degeneration predisposed the vasa vasorum toward hemorrhage and that IMH “... may be the usual rather than the uncommon mechanism of hemorrhagic dissection.” In support of this theory, intimal tears are not always present in cases of aortic dissection. There are also well-documented cases of acute IMH rupturing through the intima and evolving into more typical dissections during the course of treatment.1,5,15–23

**Natural History**

We have a limited knowledge of the natural history of IMH, as much of the data comes from case reports and small retrospective series. In a review of 505 autopsy cases of aortic dissection from 1933 to 1954, Hirst et al9 found a 4% incidence of IMH. In 1982, an autopsy study10 noted that 13% of patients with a diagnosis of aortic dissection had IMH. Similarly, clinical series1,13–17,19 have shown that 13 to 27% of patients with a diagnosis of aortic dissection in fact had IMH. IMH is difficult to distinguish from classic dissection on purely clinical grounds. The most common initial presentations of IMH are chest pain (50 to 74%) and intrascapular back pain (44 to 84%).13–21 Other more rare presentations include neurologic or vascular complications (such as syncope or transient ischemic attack), hoarseness, paraplegia, mesenteric ischemia, and acute renal failure. Patients with IMH are typically elderly (mean age, 66 years) with a history of hypertension.13–21 Unlike classic aortic dissection, the ratio of men to women appears equal.13–21 Other traditional risk factors for aortic dissection, such as bicuspid aortic valve, Marfan syndrome, and collagen vascular disease, have been distinctly uncommon in case series of IMH. Several series13–21 have noted the development of IMH following trauma. Proximal aortic involvement occurs in 40% of cases (range, 20 to 60%). IMH has a variable clinical course. While some patients have limited hemorrhage and respond well to medical therapy, IMH may progress to classic dissection (ie, with intimal disruption) in up to 33% of cases.1,13,14,17,20–33 IMH of the ascending aorta may—similar to classic dissection—rupture through the adventitia and cause pericardial effusion, hemothorax, and mediastinal hemorrhage. These vascular catastrophes largely account for the observed acute mortality from IMH (up to 30%).13–21 The mortality from proximal aortic IMH is higher than that of distal IMH (34% vs 14%), and deaths tend to occur within the first 24 to 72 h after hospital admission (Table 1).13,14

Some medically treated patients with IMH may have complete resolution.1,2,5,16–19,22,24,25,32 Yamada et al2 followed up 10 survivors of IMH and found that 8 survivors had normal chest CT scan findings within 1 year. Ide et al22 performed serial CT scans in 27 patients and demonstrated a dynamic nature of IMH. At 2 months, 19 of 27 patients with IMH showed resolution, but in 2 other patients, late progression to aortic aneurysm occurred. Furthermore, Ide et al22 described three patients in whom IMH converted to classic aortic dissection within 4 to 40 months of presentation.

Sueyoshi et al33 followed up 32 patients with IMH with serial CT imaging at weekly intervals in the first month followed by further imaging two to three times per year. Their results were similar to those of

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*Data are presented as No. of deaths/total cases. Total mortality from IMH was 34 deaths in 160 cases (21%).
†Total mortality from proximal IMH was 22 deaths in 64 cases (34%): medical treatment (14 deaths in 30 cases; 47%) and surgical treatment (8 deaths in 34 cases; 24%).
‡Total mortality from distal IMH was 12 deaths in 96 cases (14%): medical treatment (9 deaths in 76 cases; 13%) and surgical treatment (3 deaths in 20 cases; 15%).
Ide et al., with 11 of 32 patients showing resolution by 24 months and 18 of 32 patients showing aneurysms eventually evolving in the affected portions of the aorta. Interestingly, the authors noted the development of “ulcer-like projections” on the aortic intima during the follow-up period. Of 21 patients with these projections, saccular aneurysms developed in 12 patients at a mean of 45 days following the initial event, with an average growth in aortic diameter of 1.2 cm/yr. These authors hypothesized that IMH causes structural weakness of the aortic wall—even after resolution—accounting for the high incidence of late aneurysms.

**Diagnosis**

Problems in defining the natural history of IMH stem from difficulty in diagnosing the entity. While the diagnosis of aortic dissection can be challenging, detection of IMH has been even more problematic. For many years, aortography was the only method available to diagnose aortic dissection premortem, and many still consider aortography the “gold standard.” Studies have shown, however, that aortography is not as accurate as direct visualization of an intimal flap or double lumen for diagnosis, although indirect signs such as aortic thickening and compression of the true lumen by the false lumen may also be present. By definition, IMH has neither an intimal flap nor intraluminal flow.

Newer imaging techniques have markedly enhanced our ability to detect IMH. These modalities, including MRI, spiral CT, and transesophageal echocardiography (TEE), have basically replaced aortography in the evaluation of IMH. Unquestionably, MRI yields strikingly accurate images of the aorta. It is noninvasive and requires no intravascular contrast. Several studies place the sensitivity of MRI for detection of aortic dissection at 98 to 100% and the specificity at 100%. MRI also detects complications of aortic dissection and IMH, such as pericardial effusion, hemomediastinum. Although experience with IMH is considerably less than with classic dissection, MRI has accurately diagnosed IMH in several series. Clinically, the technique has several drawbacks, including difficulty with critically ill patients and those with metallic prosthetic valves or permanent pacemakers. Finally, most MRI scanners cannot accurately quantify coronary involvement or aortic regurgitation.

With MRI, IMH is characteristically seen as a focal thickening of the aortic wall in the absence of dissection (Fig 1). There is usually minimal compression of the aortic lumen, and of course no intimal flap. An acute aortic IMH has an isodense intensity on T1-weighted images and has a high signal intensity on T2-weighted images. The signal intensity evolves over time, as oxyhemoglobin is converted to methemoglobin, resulting in an increased signal intensity on T1-weighted images in subacute IMH. Dynamic phase-contrast imaging can support no flow within the aortic wall.

CT has successfully identified IMH in several reports. Compared to MRI, CT appears somewhat less accurate in identifying intimal flaps, which may lead to the overdiagnosis of IMH in cases of dissection. As the treatments for dissection and IMH are currently similar, however (see below), this point may be clinically moot. Like MRI, CT cannot identify coronary involvement or aortic regurgitation, and it has a low sensitivity for detection of branch artery occlusions. Management of an intubated or critically ill patient is often easier with CT than with MRI, but as with angiography, CT carries with it the inherent risks of intravascular contrast. A CT image (with intravascular contrast) is shown in Figure 2. The intramural hematoma is seen as an area of low attenuation within the aortic wall on contrast-enhanced CT (and of high attenuation on noncontrast CT). Absence of contrast within

![Figure 1. T2-weighted transverse MRI of descending aortic IMH. The descending aorta is dilated, and the wall is thickened. The signal from the wall is abnormally intense, consistent with IMH.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21968/ on 06/25/2017)
the aortic wall helps differentiate IMH from a classic aortic dissection (where contrast enters the false lumen). Distinguishing IMH from aortic dissection with thrombosis of the false lumen remains problematic. In addition, there may be difficulty in differentiating IMH from penetrating aortic ulcer with adherent intraluminal thrombus. In this regard, the presence of intimal calcium often can be used to distinguish intramural (subintimal) hematoma from intraluminal thrombus.\(^2,26,32,33\)

With the advent of TEE, echocardiography has become an important imaging modality for the diagnosis of aortic dissection and IMH. Early studies of transthoracic echocardiography were limited by poor image quality, especially in cases of distal aortic dissection. TEE has allowed better visualization of the aorta, and has a sensitivity and specificity of about 95% for detection of dissection (with multiplane probes).\(^4,14,29,34,36,37\) Unlike CT, TEE is remarkably accurate in the detection of intimal flaps, and therefore is an accurate technique for differentiating dissection from IMH.\(^14,16,17\) TEE is accurate in the detection of aortic regurgitation, pericardial effusion/tamponade, and proximal coronary artery involvement.\(^14,27,36\) In addition, TEE yields a diagnosis more quickly than MRI\(^28\) and is often easier to perform in critically ill and/or intubated patients than either CT or MRI.

The entities that must be distinguished from IMH are aortic aneurysm with associated thrombus, dissection with thrombosed false lumen, and severe aortic atherosclerosis (with or without penetrating aortic ulceration).\(^1,14–16\) The typical TEE findings of IMH include a focal or diffuse thickening of the aortic wall in the absence of an intimal flap or any communication between the aortic lumen and IMH (Fig 3, top and bottom). This finding distinguishes IMH from a dissection with thrombosed false channel. The luminal wall of the aorta is typically curvilinear and smooth in patients with IMH, as opposed to the rough, irregular borders seen with aortic atherosclerosis and penetrating ulcer (Fig 4). In addition, IMH tends to be homogeneous in appearance, while atheroma is often heterogeneous, speckled, and calcified on ultrasound imaging. Differentiation between IMH and aortic aneurysm with associated thrombus can be challenging, but the presence of a smooth intimal border next to the lumen (as opposed to thrombus with an irregular border interposed between the lumen and intima) favors the diagnosis of IMH or a thrombosed false lumen (Fig 3, 4).\(^38\)
A relatively new diagnostic tool that has been described for the diagnosis of IMH is intravascular ultrasound (IVUS) as an adjunct to aortography.3,19,39 The largest study involving IVUS included eight patients with a diagnosis of IMH, including four patients with diagnoses missed by other imaging procedures such as angiography and TEE. Unfortunately, IVUS also carries many of the risks of angiography, and its role in the diagnosis of IMH is not yet well defined.

Treatment

Lacking data from clinical trials, treatment of IMH has been mostly empiric. Most authorities1,3,6,13,14,40 currently recommend treatment of IMH similar to that of classic aortic dissection, with early surgery for patients with proximal IMH and medical management for patients with distal IMH. Others16,17,22 favor a conservative approach, with initial medical management for all patients in stable condition.

To our knowledge, the only study to address therapy prospectively was published by Kang et al16; these investigators suggested that IMH might have a more favorable prognosis than aortic dissection, as the hematoma is "noncommunicating" with the aortic lumen. In this study,16 27 patients with a diagnosis of IMH were initially treated with medical management. Seven patients had proximal IMH; of these seven patients, three patients eventually required surgery. Twenty patients had distal IMH, and only 1 patient required surgery. There was no mortality in the proximal dissection group; the only fatality occurred in a patient with distal IMH treated medically. The results and patient outcomes in this study16 are in stark contrast to the majority of published series on IMH (Table 1), which have shown much higher mortality rates overall for IMH. On review, however, the results of these studies are quite variable. Treatment in these reports was not randomized, and so a selection bias is likely (for example, the sickest and most moribund patients may have been treated medically rather than surgically). Although Kang et al16 attempted to avoid a selection bias, the majority of patients followed up in their study presented > 48 h after symptom onset. Given the high early mortality of aortic dissection and IMH, the population in the study of Kang et al16 may have excluded the sickest patients (leaving a group with a somewhat better prognosis). Additional possible explanations for the disparity between published series include differences in patient populations and comorbidities, rapidity of diagnosis, and accuracy of the diagnostic technique(s) employed. Despite the differences between reports, the cumulative data are instructive. Total cumulative mortality from IMH (in the 11 studies listed in Table 1) was 21%. Mortality in proximal IMH cases was 34% (47% for medical management and 24% for surgically management). Mortality in distal IMH cases was 14% (13% with medical management, 15% with surgery).

Of interest, Ide et al22 noted that those patients with IMH who progressed to classic dissection tended to have aortic diameters > 5 cm. Ide et al22 suggested that this may be a way to identify patients at risk for dissection and chronic aneurysm formation. Kaji et al41 reported similar findings. Thus, the optimal treatment for IMH is not completely clear. In some patients, medical treatment of IMH with antihypertensives and negative inotropes appears adequate, but in other patients a surgical approach may be preferable. Surgical options have usually involved the placement of Dacron grafts, although resection of the affected aorta with end-to-end anastomosis (without graft insertion) has been described.42 While endovascular stenting is emerging as a treatment option for type-B dissections,43 it has not been described in the treatment of IMH. Given the lack of an intimal flap, it is unclear what benefit endovascular stenting would have in the treatment of IMH.

In the absence of definitive clinical trials, we believe it is prudent to treat ascending aortic IMH surgically and descending aortic IMH medically (assuming there is no evidence of rupture or compromise of organ perfusion). The fact that IMH has a high incidence of late complications and aneurysm formation underscores the need for periodic aortic imaging after successful medical therapy.

**Figure 4.** TEE image of a large atherosclerotic plaque in the descending thoracic aorta. The plaque (arrow) protrudes into the aortic lumen, has an irregular border, and is heterogeneous in echocardiographic density.
Conclusion

Aortic IMH is a difficult diagnosis that requires a high index of suspicion. TEE is an excellent imaging modality for diagnosis, especially in critically ill patients. MRI is also very accurate and is useful in patients in stable condition. As scanning times decrease, the role of MRI will likely increase. CT can diagnose aortic dissection reasonably well, but it may not be completely reliable in distinguishing IMH from classic aortic dissection (as noted, the clinical importance of this point is currently unclear). CT also gives less information regarding complications and prognosis than TEE or MRI. As no single imaging modality is optimal for IMH, there may be utility in combining imaging modalities to confirm the diagnosis.

It is clear that IMH has a high rate of mortality and morbidity. Although a recent report suggested an improved prognosis of IMH over aortic dissection, survivors of IMH are at significant risk for progressive aortic abnormalities, including aortic rupture, aneurysm, and ulceration. The optimal therapy for IMH is uncertain and will remain so until randomized trials are performed. Until then, we prefer to perform immediate surgery in patients with proximal IMH (as with classic acute ascending aortic dissection). In patients with descending IMH (without rupture or compromised end-organ perfusion), we believe that medical therapy is appropriate. In this latter group, serial imaging of the aorta is recommended, as aneurysm formation is not uncommon.

After submission of this review, a retrospective analysis of very elderly patients with ascending aortic IMH was published. Compared to patients with type-A aortic dissections, the group with ascending IMH had a somewhat better prognosis. The authors suggested that supportive medical treatment with frequent follow-up imaging may be a reasonable alternative to surgery in the very old patient group with ascending aortic IMH.

References

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