Acute Intramural Hematoma of Aorta: Still Mystery for Debate

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Aortic intramural hematoma (IMH) is defined as aortic dissection without identifiable intimal tear and lack of flow in the false lumen of the aorta. Hematoma forms within the aortic wall as a result of either vasa vasorum hemorrhage, a microscopic tear of the intima that is not detectable by imaging modalities or an atherosclerotic plaque with penetrating ulceration rupture. Type A aortic IMH is defined as involvement of the ascending aorta. Type B aortic IMH is defined as involvement of only the descending aorta. The diagnosis is confirmed by computed tomography (CT) scan, magnetic resonance imaging and/or transesophageal echocardiography (TEE). The regional thickening of the aortic wall greater than 7 mm with evidence of intramural blood accumulation is considered IMH. The management of acute type A IMH is still controversial. Asian cohort used serial scans to monitor complications, which was then used to determine the timing of surgery. The treatment policy of institutions was initial medical treatment with timed surgical intervention in cases with complications, according to clinical assessment and on follow-up imaging studies. The initial therapeutic goal during the acute phase of IMH included the elimination of pain and the reduction of systolic blood pressure to 100–120 mmHg. Close clinical monitoring with transthoracic echocardiography, TEE, and CT scan is carried out to minimize the risk of fatal complications. Aortic diameter >5.0 cm and hematoma thickness >12 mm are independent predictors of development of complications and may benefit from urgent surgery. In addition, regression may occur but is less common and not predictable in proximal IMH involving the ascending aorta. Therefore, IMH involving to the aortic valve is usually related to acute mortality and needs early surgical approach. Type B IMH are generally associated with favorable outcome as compared with type A lesion. There was no difference in survival between medical vs. surgical management in patients with type B IMH.

KEY WORDS — aortic dissection, intramural hematoma
Introduction

Acute aortic syndrome was first described in 2001 by Vilacosta and Roman [1]. It is characterized by acute chest pain of aortic origin and coexisting hypertension; the acute aortic syndrome includes classic aortic dissection (AD) and aortic intramural hematoma (IMH). Although indications of surgical and medical treatment have been established for patients with AD, the approach to acute aortic IMH remains controversial due to an incomplete knowledge of its natural history. The IMH was first described in 1920 by Krukenberg [2] as “dissection without intimal tear”. Hematoma forms within the aortic wall as a result of either vasa vasorum hemorrhage, a microscopic tear of the intima that is not detectable by imaging modalities or an atherosclerotic plaque with penetrating ulceration rupture [3]. Therefore, the pathophysiology of aortic IMH is different from classic AD. In aortic IMH involving the descending aorta, the consensus is the use of aggressive anti-hypertensive treatment. In contrast, there is less agreement for the management of ascending aortic IMH. In addition, evolution of the acute aortic IMH may lead to aortic dissection due to provoking disruption of the intima. The present review aims to provide a comprehensive overview for acute aortic IMH, including its definition, diagnosis, management strategies and outcomes.

Definitions and Diagnoses

Aortic IMH is defined as aortic dissection without identifiable intimal tear and lack of flow in the false lumen of the aorta [4]. Acute aortic IMH means that the onset of aortic IMH is less than 2 weeks. Type A aortic IMH is defined as involvement of the ascending aorta. Type B aortic IMH is defined as involvement of only the descending aorta. This diagnosis could be confirmed by computed tomography (CT) scan, magnetic resonance imaging (MRI) and/or transesophageal echocardiography (TEE). Regional thickening of the aortic wall greater than 7 mm with evidence of intramural blood accumulation is considered IMH. By TEE, there is crescentic or circular thickening of the aortic wall, often containing echolucent spaces (Fig. 1) [5,6]. On contrast CT scan, aortic IMH is visualized as a low-density space of the involved segment of the aortic wall, often with displacement of intimal calcium. No contrast enhancement effect within the IMH area was shown on the postcontrast CT scan (Fig. 2) [7]. By MRI, a thickened aortic wall will similarly be seen. The signal intensity characteristics of MRI depend on the age of the hematoma [8].

Type A IMH

The management of acute type A IMH is still controversial. Robbins et al [9] first reported that
patients with acute ascending aortic IMH had an associated mortality of 66%. They recommended early surgery for these patients [9]. However, several studies of Asian cohorts with type A IMH reported good results with medical management alone, ranging from 0% to 8% [10–12]. Moreover, Song et al [10] have reported complete absorption of the IMH in as high as 67% of patients. In contrast, others have reported poor early outcomes from medical management, with mortalities ranging from 54% to 80%, leading to the recommendation of early surgery [13,14].

Estrera et al [15] reported the basic characteristics of IMH and classic AD. They found that aortic diameter and gender were similar between typical dissection and IMH. However, patients with type A IMH showed more frequently with chest pain and less frequently with moderate to severe aortic insufficiency when compared with typical dissection. Additionally, patients with type A IMH were older and in less extremis condition and with less hypotension and tamponade. However, Song et al [16] from Korea reported that patients with IMH had more frequent pericardial and pleural effusion, a higher prevalence of cardiac tamponade, and syncope at clinical presentation when compared with patients with AD.

IMH was diagnosed more frequently relative to typical AD in Asian centers versus Western centers (31.7% vs. 10.9%) [17]. One possibility may be that clinicians in Asian centers have a heightened awareness of the IMH diagnosis and different diagnostic criteria. The other possibility is that the cause of IMH is distinct in these regions based on either genetic or environmental influences [18]. In patients with IMH, 33–40% were noted to have conversion to classic AD [15,19,20]. The risks factors for conversion included initial aortic diameter > 5.0 cm and hematoma thickness > 12 mm [10,21,22]. Interestingly, Estrera et al [15] observed that no patients converted within 3 days, but an increasing risk of conversion of up to 8 days and beyond was noted.

Despite increasing experience with IMH, much still remains unclear. Some Asian cohort studies with type A IMH reported good results with medical management alone. It should be noted that the Asian cohort used serial scans to monitor complications, which was then used to determine the timing of surgery. Song et al [16] reported the largest series of type A IMH, which enrolled 101 cases from a single center. The treatment policy of that institution was initial medical treatment with timed surgical intervention in cases with complications according to clinical assessment and on follow-up imaging studies. The initial therapeutic goal during the acute phase of IMH included the elimination of pain and the reduction of systolic blood pressure to 100–120 mmHg. Close clinical monitor was using transthoracic echocardiography (TTE), TEE and CT scan to minimize the risk of fatal complications. The overall hospital mortality was lower in IMH patients than in aortic dissection patients.

Type A IMH converting to AD requires surgical intervention. Cases with aortic diameter > 5.0 cm and hematoma thickness > 12 mm are independent predictors of development of complications and may benefit from urgent surgery. Also, IMH involving the aortic valve was related to acute mortality [4]. Regression may occur but is less common and not predictable in proximal IMH involving the ascending aorta [14]. Early surgical approach should be considered in patients with proximal part of IMH. Patients with hemodynamic instability (with cardiac tamponade, impending rupture, or rupture) or persistent pain also underwent early surgery on an emergency basis. However, Kaji et al [11] reported performing pericardiocentesis in 5 patients with complications caused by cardiac tamponade and these patients were treated medically after pericardiocentesis. On the other hand, the International Registry of Aortic Dissection (IRAD) data showed the overall hospital mortality for IMH was similar to that of classic AD (20.7% vs. 23.9%) and nearly 16% of patients with IMH had developed AD. Therefore, the IRAD data favor consideration for timely surgical approach to such patients [4]. Another report also suggested that IMH involving aortic valve had a high mortality and urgent surgical intervention should be considered [23]. A meta-analysis
study reported by Maraj et al [24] enrolled 81 cases of IMH involving the ascending aorta. The mortality rate among those treated surgically was 14%, whereas among those treated medically, mortality was 36%. The obvious conclusion is that surgical management carries a lower risk than does medical therapy [24].

Type B IMH

Type B IMH are generally associated with favorable outcomes when compared to type A lesion. In type B IMH, the natural history and prognosis are not completely understood; few studies examining the predictors for progression or regression in patients with type B IMH exist. The meta-analysis study by Maraj et al [24] reported that there was no significant difference in survival rate between medical vs. surgical management in patients with type B IMH. Patients with classic AD and persistent flow communication have been reported to develop more frequent in-hospital complications and have a worse prognosis than those with IMH and absence of flow communication [6,25]. In type B IMH, distensibility of aorta may play an important role in progression or regression. Sueyoshi et al [26] reported that type B IMH with aortic diameter > 40 mm or aortic wall thickness > 10 mm must be carefully followed up so as to monitor the progression of the condition. Recommendation for early intervention in type B AD should be considered when the descending aortic diameter approaches > 55 mm in IMH patients [27]. Additionally, Falconi et al [28] reported that aortic diameter > 50 mm and persistent back pain were associated with complications, while old age and hypotension/shock were associated with in-hospital mortality in patients with type B IMH.

IMH and Penetrating Atherosclerotic Ulcer (PAU)

PAU occurs often in patients with severe aortic atherosclerosis. Previous studies reported that IMH with PAU commonly affect those at an older age and with a history of hypertension [29,30]. Stanson et al [30] first defined the PAU as an atherosclerotic lesion with ulceration penetrating the intimal elastic lamina (Fig. 3). They found that patients with PAU had a high risk for rupture or disease progression to AD. However, subsequent studies suggested a more benign course under a series of radiographic monitor [31,32]. The controversy has continued since then, with the rarity of the condition depriving us of large observational series. Interestingly, PAU with acute symptoms has a worse prognosis, while asymptomatic patients present a lower incidence of life-threatening complications. The IMH and PAU diagnosed are usually located at descending thoracic aorta and associated with aortic rupture [32,33]. Therefore, surgical intervention and endovascular treatment are suggested in type B IMH with PAU. Ganaha et al [33] demonstrated that patients with PAU > 20 mm in maximum diameter or > 10 mm in depth had a high risk of disease progression and thus should be considered candidates for early surgical or endovascular repair.

References


