Update on Aortic Intramural Hematoma

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Abstract
For the last decade aortic intramural hematoma (AIH), a variant form of classic aortic dissection (AD), has emerged as an increasingly recognized and potentially fatal entity of acute aortic syndrome (AAS). The successful clinical introduction of noninvasive imaging modalities for aortic diseases, including transesophageal echocardiography (TEE), has contributed to early differential diagnosis of AIH and investigation of its natural course or remodeling process after the initial event.

AIH, which is characterized by circular or crescentic aortic wall thickening without an intimal flap or tear, is easily differentiated from classic AD. Other findings suggestive of AIH are smooth contour of the aortic lumen, increased density of aortic wall thickening before contrast injection, and demonstration of an ‘echo-lucent area’ or ‘echo-free space’ within the thickened aortic wall. It is widely accepted that AIH is not just a precursor of AD but has diverse remodeling processes, and, compared to classic AD, has a more favorable clinical course including complete resorption with medical treatment, reported even in type A pathology. Further study has demonstrated that two important predictors for the development of AD or mortality are aortic diameter and hematoma thickness. Thus, noninvasive imaging modalities are useful not only for diagnosis but also for risk stratification and treatment strategy selection. Although a consensus regarding optimal management strategies, especially for type A AIH, has not been established, this is in part due to our incomplete understanding of AIH. With more information of this interesting disease entity, the role of TEE and other noninvasive imaging modalities is expected to be reestablished in the near future.

Key words: aortic intramural hematoma, transesophageal echocardiography

With the recent advances and successful clinical introduction of various noninvasive imaging modalities for aortic pathology [1], a considerable number of patients visiting an emergency room for severe ‘aortic’ pain are ultimately found to have aortic intramural hematoma (AIH), a variant form of classic aortic dissection (AD). As clinical symptomatology is very similar and not useful in distinguishing between these two important disease entities, a new cardiovascular syndrome, the ‘acute aortic syndrome’ (AAS) has been suggested. AAS would embrace a heterogeneous group of patients presenting a similar clinical profile including penetrating atherosclerotic aortic ulcer, AIH and the classic AD [2]. Accurate differential diagnosis of AAS is totally dependent on imaging techniques, and follow-up imaging is necessary as one entity of AAS may progress to another entity of AAS [2]. Thus, in clinical practice, the importance of noninvasive imaging modalities for aortic pathology, such as contrast enhanced X-ray computed tomography (CT), magnetic resonance imaging (MRI), and transesophageal echocardiography (TEE), cannot be overemphasized for patients with AAS. Among them, TEE is unique as it is the only technique directly used by attending cardiologists familiar with a patient’s medical history or condition to make clinical decisions at the bedside. In 1994, Dr. Erbel’s group in Mainz University the first to report a diagnosis of AIH by TEE [3]. Since then, AIH and AAS have emerged as interesting topics for clinical investigators, especially cardiologists. In this presentation, I would like to summarize updated information and discuss the role of
noninvasive imaging studies, including TEE, on the diagnosis and management of AIH.

A. Epidemiology: East Versus West

AIH occurs predominantly in the elderly population and the incidence of AIH shows a single peak in the 7th decade. As the population increasingly ages and survival is prolonged despite hypertension, a major risk factor for the development of AAS, the incidence of AIH is expected to increase significantly. In earlier studies, it was reported that AIH accounts for 10 to 20% of patients with a clinical presentation of AAS. However, with the wide application of noninvasive imaging modalities in clinical practice and the resulting increased recognition of AIH, the relative incidence of AIH might increase. One interesting point is that there might be a geographical difference in the relative incidence of AIH. Table 1 summarizes the incidence of AIH versus AD reported in literature [4-11]. In western countries, the relative incidence of AIH ranges from 5.1 to 22.5%. In Japan and Korea, the incidence is higher than 25%. The relatively larger population of patients with poorly controlled or uncontrolled hypertension in the East might be one of the factors explaining this difference. Further study is necessary to determine whether this difference represents any racial or genetic heterogeneity.

Contrary to AD, which more frequently affects the ascending aorta, AIH has a strong tendency to involve the distal aorta. According to recently reported data of the International Registry of Acute Aortic Dissection (IRAD), the total number of patients with AAS involving the proximal (type A) and distal (type B) aorta were 547 and 328, respectively [12-13]. Among the 547 patients with type A AAS, 8.0% (44/547) were due to AIH. The prevalence of AIH in type B AAS was 15.6% (60/384). Based on our data of AAS from January 1993 to March 2003, AIH accounted for 27.4% of type A AAS (51/186) and 58.3% of type B AAS (108/185). Thus, the relative incidence of AIH affecting the distal aorta

<table>
<thead>
<tr>
<th>Author</th>
<th>Country</th>
<th>Duration</th>
<th>AD (n)</th>
<th>AIH (n)</th>
<th>AIH (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vaccari [5]</td>
<td>Italy</td>
<td>'91 – '99</td>
<td>166</td>
<td>9</td>
<td>5.1 %</td>
</tr>
<tr>
<td>Ganaha [6]</td>
<td>USA</td>
<td>'90 – ’00</td>
<td>663</td>
<td>65</td>
<td>9.1 %</td>
</tr>
<tr>
<td>Evangelista [7]</td>
<td>Spain</td>
<td>'90 – ’00</td>
<td>234</td>
<td>68</td>
<td>22.5 %</td>
</tr>
<tr>
<td>Sohn [8]</td>
<td>Korea</td>
<td>'95 – ’99</td>
<td>48</td>
<td>16</td>
<td>25.0 %</td>
</tr>
<tr>
<td>Kaji [9], [10]</td>
<td>Japan</td>
<td>'88 – ’00</td>
<td>158</td>
<td>83</td>
<td>34.4 %</td>
</tr>
<tr>
<td>Moizumi [11]</td>
<td>Japan</td>
<td>'90 – ’02</td>
<td>139</td>
<td>94</td>
<td>40.3 %</td>
</tr>
<tr>
<td>Song</td>
<td>Korea</td>
<td>'93 – ’03</td>
<td>212</td>
<td>159</td>
<td>42.9 %</td>
</tr>
</tbody>
</table>

AD, aortic dissection; AIH, aortic intramural hematoma.

Fig. 1. Circadian (left) and seasonal (right) variation in onset of aortic intramural hematoma.
is about two times higher than that affecting the proximal aorta; there seems to be no geographical or racial difference of this tendency.

Characteristic chronobiological patterns can be found in the occurrence of AIH (Figure 1). According to the analysis of 159 patients with AIH in our center, a significantly higher frequency of AIH occurred from 6:00 AM to 12:00 PM with a peak at 8:00 AM. Another small peak was demonstrated at 9:00 PM, which had a significant association with patients less than sixty years old and a history of hypertension. This finding may suggest the importance of more aggressive hypertension control in groups with a relatively more active social life style. Although no significant variation was found among days of the week, the frequency of AIH was significantly higher during the winter months (Figure 1, right). These significant circadian and seasonal variations observed may have important implications for the prevention of AIH by tailoring treatment strategies to ensure maximal benefits during these vulnerable periods. These variations or chronobiological patterns are also very similar to those of classic AD [14].

B. Diagnosis of AIH: Typical Cases and Difficult Scenarios

In classic AD, flow communication occurs through a demonstrable primary intimal tear: Flow propagation creates a so called ‘double-channel aorta’ with a true and false lumen separated by an intimal flap. In AIH, it is believed that hemorrhage occurs within the aortic wall in the absence of an initial intimal disruption [2]. Thus, conventional aortography, which is useful for detection of an intimal flap or double channel aorta in classic AD, will fail to identify AIH [15] and make ante-mortem diagnosis of AIH difficult. The hallmark of imaging diagnosis in AIH is the demonstration of crescentic or circular aortic wall thickening without an intimal flap or tear [16]. Noninvasive radiological techniques, such as CT and MRI, were first used for the diagnosis of AIH [17]. In CT (Figure 2), the characteristic finding is a crescentic, high attenuation area along the aortic wall without an intimal flap (Figure 2, A and C) that fails to enhance after injection of contrast medium (Figure 2, B and D). This characteristic finding is
also very easily detected by MRI; the signal intensity of the thickened aortic wall might vary depending on the amount of methemoglobin formation within the hematoma.

TEE can also be used to demonstrate circumferential or crescentic aortic wall thickening without an intimal tear as well as displacement of the intimal calcification caused by the hemorrhage within the aortic media (Figure 3). According to our data, the mean thickness (±SD) of type A AIH (n=48 patients) and type B AIH (n=103 patients) was 12.7±6.9 mm (range 5-40 mm) and 10.5±3.5 mm (range 5-23 mm), respectively. As the normal thickness of the aorta is less than 3 mm by any imaging modality, aortic wall thickness greater or equal to 5 mm was adequate for diagnosis of AIH in patients with typical clinical symptoms suggesting AAS.

AIH is quite easily differentiated from classic AD; an intimal tear or flap is absent, and there is no evidence of direct flow communication (Figure 4). However, as AIH is prevalent in elderly patients with hypertension, other common pathologies in this population, such as aortic wall thickening caused by atherosclerotic changes or aneurysmal dilatation with mural thrombi, can cause a challenging diagnostic problem. Identification of the intima and careful observation of the thickened aortic wall's inner surface are helpful to distinguish AIH from other aortic conditions. Usually the inner margin of AIH is smooth, and aortic thickening occurs beneath a bright, echo-dense intima (Figure 4).

**Fig. 3.** Representative transesophageal echocardiographic (TEE) images of aortic intramural hematoma demonstrating crescentic aortic wall thickening without intimal flap or flow communication in the descending (A) and ascending aorta (B).

**Fig. 4.** Representative images of classic aortic dissection (A, B, C) and intramural hematoma (D, E, F).
3), whereas an irregular margin caused by thickening above the intima from a dilated aorta (Figure 5) is commonly observed in patients with significant atherosclerotic changes or mural thrombi.

Another differential point is the tissue characterization information from each imaging modality. For a confirmative diagnosis of AIH, evidence of an ‘acute hemorrhage’ or ‘accumulation of blood’ causing aortic wall thickening is very helpful. In CT, ‘high attenuation’ of the thickened aortic wall before contrast injection seems to represent ‘acute hemorrhage’ (Figure 2); this increased density disappears with time (Figure 6). As this kind of high attenuation without contrast medium is absent in chronic aortic illnesses such as atherosclerotic changes or aneurysmal dilatation with mural thrombi, careful observation of both pre- and post-contrast injection CT films is very important for the cor-

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**Fig. 5.** A representative transesophageal image of aortic aneurysm with mural thrombi. An irregular inner aortic margin above the intima in a notably dilated aorta is a characteristic finding.

**Fig. 6.** Computed tomographic images of aortic intramural hematoma showing temporal change of high attenuation in a pre-contrast study. In the initial study (middle), increased density of aortic wall thickening in the ascending and descending aorta (arrows) was prominent, which disappeared at follow-up study (right) performed 9 days later.

**Fig. 7.** Transesophageal echocardiographic images showing various patterns of an ‘echo-free space’ or ‘echo-lucent area’ in patients with aortic intramural hematoma.
rect diagnosis of AIH.

TEE is superior to any imaging modality as it allows direct observation of the entire aortic wall. One characteristic TEE finding of AIH is the presence of an ‘echo-free space’ within the thickened aortic wall; the prevalence of this finding is reported to be higher than 60% [18]. The extent or size of the ‘echo-free space’ or ‘echo-lucent area’ found in patients with AIH varies significantly (Figure 7) and can be larger than half of the whole aortic wall thickening in some patients. Demonstration of an ‘echo-free space’ or ‘echo-lucent area’ is quite unique or pathognomonic for AIH, and we believe it represents the different patterns of liquefaction after initial bleeding. Although some patients with a large ‘echo-free space’ detected in TEE also show contrast enhancement during CT, suggesting flow communication with a true lumen (Figure 8), the presence of an ‘echo-free space’ is not a poor prognostic sign and is not associated with development of classic AD [18].

It is generally believed that AIH is caused by rupture of the vasa vasorum [2], which then separates the medial wall layers eventually leading to a secondary tear or to a communication with the adventitial space. However, some sporadic case reports showing accidental development of typical AIH by percutaneous catheter manipulations, such as the insertion of a balloon pump [19], coronary angioplasty [20], or catheter ablation of a left sided bypass tract (Figure 9), support the presence of a ‘primary intimomedial tear’ in AIH. This hypothesis is also supported by the demonstration of a small intimal communication at the time of surgery [21, 22]. These observations raise the issue of the diagnostic ability of various noninvasive imaging modalities to identify an ‘intimal tear’, which is considered a critical criterion to differentiate AD from AIH. The only difference between AD and AIH might be whether the intimal tear is big enough to be detected by contemporary noninvasive imaging techniques. Further investigation is necessary to verify if the

Fig. 8. Two cases of aortic intramural hematoma with a large ‘echo-free space’ detected by transesophageal echocardiography. Usually, in contrast-enhanced computed tomography, there is no contrast enhancement or evidence of flow communication (B). In some patients, focal contrast enhancement can be observed (A); however, ‘echo-free space’ is not a predictor of development of typical aortic dissection.
‘micro-tear’ is an initiating event of AIH or just a decompression vent.

C. Natural History and Treatment

As clinical symptomatology is very similar between AD and AIH, and rapid progression of AIH to classic AD or aortic rupture had been reported in earlier clinical studies, the same therapeutic strategies were recommended for patients with AIH and those with classic AD [4]. However, from a pathological viewpoint,
AIH is different from AD in terms of the absence of an intimal tear or a continuous flow communication between the true and false lumen; this difference may have a different impact on the clinical course of patients with AIH. This hypothesis was proved in part by a report showing that with the same medical treatment, AIH involving the distal descending thoracic aorta shows a much higher rate of complete resorption of the aortic pathology than typical AD [23]. Even in type A AIH involving the ascending aorta (Figure 10) [24-26], the clinical results of ‘aggressive’ medical treatments are better than in classic AD [27]. These observations, that the complete resorption of the hematoma is possible with medical treatment, suggest that AIH and AD are two distinctly different entities of AAS.

It is now widely accepted that AIH is not just a precursor of AD but has diverse remodeling processes after the initial event [7, 26]. With serial imaging studies, AIH was proven to have four different possible courses: Persistent hematoma; reabsorbed hematoma, so that the appearance of the aortic wall returns to normal; progression to aortic aneurysm; or conversion to a classic AD, with the development of a typical intimal flap and flow in a false lumen. Although the development of classic AD from AIH with medical treatment is the most disastrous remodeling process, its frequency is less than 30%. Thus, risk stratification based on imaging, including identification of predictors for clinical events including the development of AD or hospital mortality, has been a hot issue. Hitherto, the two most important variables reported as useful predictors for the development of adverse clinical events are aortic diameter and hematoma thickness [6, 24, 28, 29]. These observations are important as they demonstrate that noninvasive imaging modalities including TEE is useful not only for correct differential diagnosis but for risk stratification and selection of treatment strategy [30].

Although controversial, the initial medical treatment for patients with type A AIH has been reported to have acceptable results that are not so significantly different from that of initial surgical intervention [24, 27, 31]. A selective treatment strategy of initial medical treatment and timed surgery with frequent follow-up imaging studies has been proposed [8, 9] and is still in debate [32]. Selection of treatment options and results with medical treatment in these patients vary significantly among institutions (Table 2). In Germany [32], institutions report that most patients undergo surgery, and that with medical treatment only, early mortality was very high. This is very similar to recently published IRAD data on patients with classic type A AD undergoing medical treatment only [33]. However, other institutions in Spain, Japan, and Korea reported significantly lower mortality in a relatively high prevalence of patients without early surgical intervention undergoing medical treatment only. When aggressive medical treatment is selected for type A AIH, frequent follow-up imaging with surgical ‘back up’ is absolutely necessary, as wall configuration of AIH can change very rapidly, especially in patients with a very thick hematoma in the initial imaging study (Figure 11) [16]. Development of an ulcer-like projection at a follow-up imaging study is an excellent predictor for progression to classic AD and close monitoring is warranted despite a stable clinical condition [16].

The revolutionary change in the treatment of various aortic pathologies is the successful introduction of endovascular management. This treatment option is

### Table 2. Comparison of treatment option and results in patients with type A aortic intramural hematoma reported in the literatures.

<table>
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<tr>
<th>Author</th>
<th>Study Duration</th>
<th>Year Published</th>
<th>Study Design</th>
<th>Number of Cases</th>
<th>Patients with Medical Treatment Only</th>
<th>In-hospital Mortality without Surgery</th>
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<tr>
<td>Evangelista [7]</td>
<td>'90-'00</td>
<td>2003</td>
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<td>12</td>
<td>50.0% (6/12)</td>
<td>16.7% (1/6)</td>
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<tr>
<td>von Kodolitsch [32]</td>
<td>'94-'00</td>
<td>2003</td>
<td>Multicenter (4)</td>
<td>38</td>
<td>28.9% (11/38)</td>
<td>54.5% (6/11)</td>
</tr>
<tr>
<td>Sueyoshi [31]</td>
<td>'84-'95</td>
<td>1997</td>
<td>Multicenter (2)</td>
<td>13</td>
<td>62.5% (8/13)</td>
<td>12.5% (1/8)</td>
</tr>
<tr>
<td>Kaji [24]</td>
<td>'91-'97</td>
<td>1999</td>
<td>Multicenter (2)</td>
<td>22</td>
<td>59.1% (13/22)</td>
<td>7.7% (1/13)</td>
</tr>
<tr>
<td>Shimizu [25]</td>
<td>'95-'99</td>
<td>2000</td>
<td>Single center</td>
<td>13</td>
<td>84.6% (11/13)</td>
<td>27.3% (3/11)</td>
</tr>
<tr>
<td>Moizumi [11]</td>
<td>'90-'02</td>
<td>2004</td>
<td>Single center</td>
<td>41</td>
<td>51.2% (21/41)</td>
<td>4.8% (1/21)</td>
</tr>
<tr>
<td>Song</td>
<td>'93-'03</td>
<td>submission</td>
<td>Single center</td>
<td>51</td>
<td>68.6% (35/51)</td>
<td>8.6% (3/35)</td>
</tr>
</tbody>
</table>

especially useful for a localized ulcer or dissection (Figure 12), which can develop during medical treatment of type B AIH [34]. Stent-graft management for localized complications of type B AIH seems to be very beneficial for the natural healing or remodeling process and has almost replaced surgical management. For ideal candidate selection for stent-graft management, noninvasive imaging modalities, including TEE, provide invaluable information, such as the location, extent, and size of the localized ulcer or dissection. Monitoring of the descending thoracic lesion during the procedure is another advantage and indication of TEE.

Fig. 11. Two cases of aortic intramural hematoma showing very rapid changes of aortic wall configuration at the hyper acute stage. The case presented in panels A and B shows a dramatic decrease of hematoma within 7 hours, whereas the case presented in panels C and D shows development of a typical aortic dissection within 12 hours after pain onset.

Fig. 12. A case showing evolution of type B aortic intramural hematoma to localized dissection and endovascular management. Aortogram and computed tomography showed complete normalization of descending thoracic aorta after successful stent-graft deployment and medical treatment.
D. Conclusion

For the past decade, we have witnessed dramatic changes in the old concept of AAS. These changes include diagnostic identification of AIH, risk stratification, and selection of treatment strategies based on information provided by imaging studies. No one can deny that TEE has performed a pivotal role in making this change possible; through this modality, attending cardiologists have had an excellent means to study this interesting topic of AAS. Although a consensus regarding optimal management strategies, especially for type A AIH, has not been established, this is in part due to our incomplete understanding of AIH as it was only first clearly established in the mid-1980s. With more information of this rapidly changing and interesting disease entity, the role of TEE and other noninvasive imaging modalities is expected to be reestablished in the near future.

References