Different Clinical Features of Aortic Intramural Hematoma Versus Dissection Involving the Ascending Aorta

Jae-Kwan Song, MD, FACC,* Hyun-Sook Kim, MD,* Duk-Hyun Kang, MD,* Tae-Hwan Lim, MD,† Meong-Gun Song, MD,‡ Seong-Wook Park, MD, FACC,* Seung-Jung Park, MD, FACC*

Seoul, Korea

OBJECTIVES
The goal of this study was to test the hypothesis that the absence of direct flow communication through intimal tear in aortic intramural hematoma (AIH) involving the ascending aorta has different clinical impact on clinical course compared with typical aortic dissection (AD).

BACKGROUND
Although emergent surgical repair has been applied for patients with proximal AIH as if it was typical AD, the natural history of proximal AIH is not known clearly yet.

METHODS
Direct comparison of the clinical data of 81 patients with proximal AD and 24 patients with AIH was performed retrospectively.

RESULTS
Patients with AIH were older (67 ± 10 vs. 50 ± 13, p = 0.001), and female gender was more predominant in AIH (19/24 vs. 29/81, p = 0.001). The development of mediastinal hemorrhage and pericardial and pleural effusion was more frequent in patients with AIH than it was in patients with AD. Although medical treatment was more frequently selected in the AIH group (75% vs. 29/81, p = 0.001). The development of mediastinal hemorrhage and pericardial and pleural effusion was more frequent in patients with AIH than it was in patients with AD (6% vs. 58%, p = 0.003). In follow-up imaging studies of 13 patients who survived AIH without surgical repair, seven patients showed complete resolution. Typical AD developed in three patients, and the other three patients showed focal AD only in the descending aorta. The two-year survival rate did not show significant difference (84% ± 6% in AIH vs. 76% ± 17% in AD, p = 0.47).

CONCLUSIONS
Absence of continuous flow communication can explain a more favorable clinical course of AIH than for AD, and medical treatment with frequent imaging follow-up and timed elective surgery in cases with complications can be a rational option for patients with proximal AIH.

(J Am Coll Cardiol 2001;37:1604–10) © 2001 by the American College of Cardiology

Aortic intramural hematoma (AIH), known as a variant form of aortic dissection (AD), is characterized by the absence of intimal tear and direct flow communication between true and false lumen (1–8). Initial clinical studies concluded that AIH is a precursor or very early stage of AD, having a high rate of progression to overt dissection, and the same treatment strategy has been applied for AIH as for classic AD (3,5,9). For patients with AIH involving the ascending aorta, surgical intervention has been considered as a standard treatment modality. However, the natural history of proximal AIH is not clearly known yet, and most clinical studies included only small numbers of patients with proximal AIH (3,5,8). From a pathological point of view, AIH is different from AD in terms of the absence of continuous direct flow communication through intimal tear; however, the impact of different false lumen hemodynamics on the clinical courses has not been seriously investigated.

Persistent flow communication between the true and the false lumen is a well-known adverse prognostic factor in patients with classic AD (10–18). It is also reported that, with the same medical treatment, AIH involving the distal descending thoracic aorta shows a much higher rate of complete resolution of the aortic pathology than typical AD (17). All these findings helped us develop a hypothesis that the absence of intimal tear and continuous flow communication in patients with AIH may have a different impact on clinical courses compared with AD. The previous studies were mainly focused on the description of the clinical features of AIH, and were limited by the heterogeneous patient population in terms of the affected site and treatment modality. We intended to test the hypothesis in patients with proximal aortic pathology by direct comparison of the clinical data of patients with AD and AIH involving the ascending aorta.

METHODS
From January 1992 to July 1999, routine clinical and diagnostic evaluation consisting of random combinations of
contrast-enhanced X-ray computed tomography (CT), magnetic resonance imaging (MRI) and transesophageal echocardiography (TEE) identified 81 patients as having acute proximal AD and 24 patients as having acute proximal AIH in the Asan Medical Center. All consecutive patients underwent diagnostic imaging studies within two days from chest pain onset. Patients with chronic AD or AIH were excluded. Typical double-channel aorta with dissecting membrane or intimal tear was an imaging criterion for diagnosis of AD (Fig. 1, A and C). Exclusion of dissecting flap or intimal disruption was a prerequisite for diagnosis of AIH by any imaging modality (5,18). Regional thickening of the aortic wall >7 mm in a circular or crescent shape or evidence of intramural accumulation of blood in TEE and a crescentic or circular high attenuation area along the aortic wall without enhancement after contrast injection in CT and MRI were considered diagnostic of AIH (Fig. 1, B and D). For all these patients, noninvasive diagnostic imaging studies were performed within two days after the onset of chest pain.

Retrospective analysis of clinical data including symptoms, signs, therapeutic modality, hospital course and follow-up data was done. Clinical data were obtained from the hospital records, including surgical reports. Clinical follow-up data were obtained in all patients by direct or telephone interview. Overall clinical outcome was compared using survival rates at two years.

Numerical values are expressed as mean ± SD. Statistical analysis of the difference between two groups was assessed by the Student unpaired t test. The chi-square test and Fischer exact test were used to compare frequency ratios between groups. Survival analysis was performed with Kaplan-Meier analysis, and differences in survival between groups were examined with the log-rank test. A p value <0.05 was considered statistically significant.

**Abbreviations and Acronyms**

AD = aortic dissection
AIH = aortic intramural hematoma
CT = contrast-enhanced X-ray computed tomography
MRI = magnetic resonance imaging
TEE = transesophageal echocardiography

**Figure 1.** Representative X-ray computed tomograms (A and B) and transesophageal echocardiograms at the basal horizontal view (C and D) in proximal aortic dissection and intramural hematoma. 'Double-channel aorta' with intimal flap is characteristic in aortic dissection (A and C), whereas crescentic aortic wall thickening without any evidence of flow communication in aortic intramural hematoma (B and D). LA = left atrium; PA = pulmonary artery; RA = right atrium.
or family members refused surgery due to old age. In the AIH group, only 25% (6/24) underwent surgery, which was significantly lower than in the AD group (p = 0.001); surgical intervention was rejected by the patients or family members due to old age in 14 patients. Other medical illnesses associated with old age, such as malignancy, asthma and stroke, made operation an unacceptable treatment modality (Table 2). Medical treatment for these patients with AD or AIH was general supportive care with aggressive antihypertensive medications to maintain the systolic blood pressure ≤120 mm Hg. Contrast-enhanced X-ray computed tomography or TEE was repeated once a week, and the patients stayed at the intensive care unit for the first two weeks. Without any clinical event with oral medications, the patients were discharged.

The overall in-hospital mortality of the proximal AIH group was 8%, which was not significantly different from that of the AD group (20%, p = 0.24). However, the mortality with medical treatment in the proximal AIH group was 6%, whereas the AD group showed much higher mortality (58%, p = 0.003). Operative mortality in the two groups did not show any significant difference (17% in AIH vs. 14% in AD, p = 0.96). In the AD group, medical treatment was done for 12 patients (15%); among them, seven patients died during the hospital admission, and the remaining five patients showed no significant interval change of typical ‘double-channel aorta’ in the follow-up imaging studies.

In the AIH group, medical treatment was selected for 18 patients (75%); one patient (case no. 11) was in a shock status during imaging studies and died before surgical consultation on the same day. The other patients could be effectively stabilized with supportive medical treatment. In two patients (case nos. 6 and 8), emergent pericardiocentesis was done on the day of admission. Surgical intervention was done during hospital stay in another two patients due to development of typical type I AD on the 17th day (case no. 12) and an increase of pleural effusion on the 23rd day (case no. 16). Fifteen patients were discharged without any event and followed for 15 ± 3 months. Among them, follow-up imaging study could be obtained in 13 patients (87%). Complete resorption of proximal AIH occurred in seven patients (Fig. 3). Typical AD developed in three patients about two months after the acute event (DeBakey type I/II = 1/2). Among them, one patient (case no. 17) underwent surgical repair successfully. Another one (case no. 2) died, who was a poor surgical candidate due to intractable asthma (Fig. 4). In the other three patients (case nos. 1, 7 and 13), focal AD developed in the descending aorta (Fig. 5), and they are under close follow-up observation without surgical intervention.

During follow-up procedure, two patients in the AIH group (case nos. 2 and 4) died; sudden death occurred two years after the acute event in case no. 4, who showed complete normalization of proximal AIH. In the AD group, three patients with postoperative complications, such as

### RESULTS

Table 1 summarizes the clinical features of both groups. Mean age of patients with proximal AIH was 67 ± 10 years, which was significantly higher than that of patients with proximal AD (50 ± 13, p = 0.001). Female gender was more frequently observed in the AIH group (79% vs. 36%, p = 0.001). The prevalence of hypertension, chest or back pain and syncpe did not show any difference in two groups. In patients with Marfan’s syndrome, AD was the only clinical presentation. There was no statistically significant difference between the two groups in DeBakey type and incidence of renal dysfunction, but pericardial effusion (88% vs. 57%, p = 0.007), pleural effusion (75% vs. 22%, p = 0.001) and mediastinal hemorrhage (21% vs. 1%, p = 0.002) occurred more frequently in the AIH group. Significant aortic regurgitation (moderate to severe) was observed more frequently in the AD group.

Selected treatment modalities and hospital course are summarized in Figure 2. For all these patients, surgical intervention was strongly recommended. In the AD group, 85% (69/81) underwent emergent surgical repair, and medical treatment was selected for the remaining 12 patients. Multi-organ failure at the time of clinical presentation was the major reason for selecting medical treatment in eight patients who were not suitable for surgical intervention. In the remaining four patients with proximal AD, the patients...
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yr)</th>
<th>Gender</th>
<th>Diagnostic Imaging</th>
<th>DeBakey Type</th>
<th>Initial Treatment</th>
<th>Underlying Disease</th>
<th>In-Hospital Outcome</th>
<th>Duration of Follow-Up (Months)</th>
<th>Follow-Up Imaging</th>
<th>Outcome (DeBakey Type)</th>
<th>Time to Dissection</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>74</td>
<td>F</td>
<td>CT, TEE, MRI</td>
<td>II</td>
<td>Medical</td>
<td>Alive</td>
<td>8</td>
<td>CT</td>
<td>Focal dissection (III)</td>
<td>20 days</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>78</td>
<td>F</td>
<td>CT</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>15</td>
<td>CT</td>
<td>Overt dissection (I)</td>
<td>2 months</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>80</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>12</td>
<td>CT, TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>71</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>35</td>
<td>MRI</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>87</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>37</td>
<td>TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>72</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical†</td>
<td>Alive</td>
<td>18</td>
<td>CT</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>48</td>
<td>M</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>15</td>
<td>TEE</td>
<td>Focal dissection (III)</td>
<td>2 months</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>71</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>17</td>
<td>CT, TEE</td>
<td>Overt dissection (II)</td>
<td>3 months</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>71</td>
<td>F</td>
<td>CT, TEE</td>
<td>II</td>
<td>Medical</td>
<td>Alive</td>
<td>2</td>
<td>CT, TEE</td>
<td>Focal dissection (III)</td>
<td>2 months</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>72</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>56</td>
<td>CT, TEE</td>
<td>Overt dissection (II)</td>
<td>3 months</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>54</td>
<td>F</td>
<td>CT, TEE</td>
<td>II</td>
<td>Medical</td>
<td>Alive</td>
<td>6</td>
<td>CT, TEE</td>
<td>Overt dissection (I)</td>
<td>17 days</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>48</td>
<td>M</td>
<td>CT, TEE, MRI</td>
<td>II</td>
<td>Medical*</td>
<td>Alive</td>
<td>7</td>
<td>CT, TEE</td>
<td>Focal dissection (III)</td>
<td>3 months</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>60</td>
<td>M</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>2</td>
<td>CT, TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>54</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>3</td>
<td>CT, TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>57</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>3</td>
<td>CT, TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>60</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical†</td>
<td>Alive</td>
<td>1</td>
<td>CT, TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>75</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>1</td>
<td>CT, TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>72</td>
<td>M</td>
<td>CT, TEE</td>
<td>I</td>
<td>Medical</td>
<td>Alive</td>
<td>1</td>
<td>CT, TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>75</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Surgery</td>
<td>Alive</td>
<td>37</td>
<td>CT, TEE</td>
<td>Overt dissection (II)</td>
<td>3 months</td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>68</td>
<td>F</td>
<td>CT, TEE</td>
<td>II</td>
<td>Surgery</td>
<td>Alive</td>
<td>30</td>
<td>TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>21</td>
<td>72</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Surgery</td>
<td>Alive</td>
<td>25</td>
<td>TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>62</td>
<td>F</td>
<td>CT, TEE</td>
<td>I</td>
<td>Surgery</td>
<td>Alive</td>
<td>68</td>
<td>CT, TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>23</td>
<td>66</td>
<td>M</td>
<td>CT, TEE</td>
<td>I</td>
<td>Surgery</td>
<td>Alive</td>
<td>19</td>
<td>CT, TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
<tr>
<td>24</td>
<td>68</td>
<td>F</td>
<td>TEE</td>
<td>I</td>
<td>Surgery</td>
<td>Alive</td>
<td>25</td>
<td>TEE</td>
<td>Complete resorption</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Delayed elective surgical repair after confirmation of the development of typical aortic dissection; †Emergent percutaneous pericardiocentesis on the day of admission.

CT = contrast-enhanced X-ray computed tomography; MRI = magnetic resonance imaging; TEE = transesophageal echocardiography.
cerebrovascular accident and low cardiac output status, died. The two-year survival rate of the AIH group was 84% ± 6%, which was not significantly different from that in the AD group (76% ± 17%, p = 0.47, Fig. 6). The power to detect a 20% difference in survival rates with a two-sided significance level of 0.05 was 0.512.

DISCUSSION

The main finding of this study was that patients with proximal AIH show different clinical features and a much better prognosis with medical treatment compared with those with AD. This supports our initial hypothesis that AIH is not just a precursor of overt AD but a distinct disease entity, and absence of a continuous flow communication between true and false lumen in AIH can have different clinical impact. This difference in the false lumen hemodynamic may explain the excellent prognosis with medical treatment in the AIH group.

Natural history of proximal AIH. In the literature, most initial clinical investigations reported a very poor prognosis.
of proximal AIH without surgical repair (3,5,8,19) and concluded that AIH is associated with a clinical profile and prognosis similar to AD, and the outcome of proximal AIH appears favorable only with immediate surgical repair (5). However, recent reports from Japan (20,21) showed an excellent clinical course of proximal AIH with medical treatment, and they concluded that they have some concerns about the strategy that all patients with proximal AIH need urgent surgical repair (20).

Although pericardiocentesis in the clinical setting of typical proximal AD has been reported to be harmful rather than beneficial (22), Kaji et al. (21) and Shimizu et al. (23) reported successful pericardiocentesis and rapid stabilization of clinical conditions in several patients with proximal AIH. Thus, the results of our study and others suggest that, if we can successfully stabilize patients’ condition with general supportive care, elective surgery with frequent imaging follow-up can be a rational option for the old patients with AIH. Clinical availability of various noninvasive imaging modalities for aorta (24) and surgical backup are absolute prerequisite conditions for this option.

The development of AD has been described as a serious complication of AIH. Interestingly, in our study, development of typical AD is not as frequent as reported, and the pattern of AD after proximal AIH is quite variable. In some patients, typical AD developed in the same aortic lesion of previous AIH, whereas, in other patients, focal AD developed only in the localized area of the descending aorta with near complete normalization of the ascending aorta. Maximum aortic diameter estimated by the initial CT images has been reported to be associated with progression of proximal AIH (21), but age-related normal values should be reconsidered. Further systematic investigation with frequent imaging follow-up in these patients is necessary.

Clinical implications. In conclusion, considering a very old patient population and the relatively high mortality and morbidity of aortic surgery, supportive medical treatment with frequent follow-up imaging studies and timed surgical repair in cases with complications can be a rational option in management of patients with AIH involving the ascending aorta. The role of elective surgery in proximal AIH should be established by future investigations.
REFERENCES