Intimal Tear Without Hematoma
An Important Variant of Aortic Dissection That Can Elude Current Imaging Techniques

Lars G. Svensson, MD, PhD; Sherif B. Labib, MD; Andrew C. Eisenhauer, MD; John R. Butterly, MD

Background—The modern imaging techniques of transesophageal echocardiography, CT, and MRI are reported to have up to 100% sensitivity in detecting the classic class of aortic dissection; however, anecdotal reports of patient deaths from a missed diagnosis of subtle classes of variants are increasingly being noted.

Methods and Results—In a series of 181 consecutive patients who had ascending or aortic arch repairs, 9 patients (5%) had subtle aortic dissection not diagnosed preoperatively. All preoperative studies in patients with missed aortic dissection were reviewed in detail. All 9 patients (2 with Marfan syndrome, 1 with Takayasu’s disease) with undiagnosed aortic dissection had undergone ≥3 imaging techniques, with the finding of ascending aortic dilatation (4.7 to 9 cm) in all 9 and significant aortic valve regurgitation in 7. In 6 patients, an eccentric ascending aortic bulge was present but not diagnostic of aortic dissection on aortography. At operation, aortic dissection tears were limited in extent and involved the intima without extensive undermining of the intima or an intimal “flap.” Eight had composite valve grafts inserted, and all survived. Of the larger series of 181 patients, 98% (179 of 181) were 30-day survivors.

Conclusions—in patients with suspected aortic dissection not proven by modern noninvasive imaging techniques, further study should be performed, including multiple views of the ascending aorta by aortography. If patients have an ascending aneurysm, particularly if eccentric on aortography and associated with aortic valve regurgitation, an urgent surgical repair should be considered, with excellent results expected. (Circulation. 1999;99:1331-1336.)

Key Words: aorta ■ dissection ■ imaging ■ surgery

Dissection can be defined as the process of separating tissue planes along intervening layers of connective tissue.1 Thus, aortic dissection is defined as the separation of the lamellae of the aortic wall. Extraluminal blood under pressure usually advances the intramural process of dissection and may later clot. The extraluminal blood usually arises from an intimal tear, and in only 3% to 13% of autopsies is a noncommunicating intramural hematoma found without an intimal tear site.2–6 The extent of undermining of the intimal layer by dissection can vary from being only a few millimeters to extending from the aortic valve down into the iliac arteries.1,4,5 This creates a false lumen that varies from only a few millimeters to the larger classic false lumen with an associated flap or septum.

There are thus 2 well-recognized forms of aortic dissection in the aortic wall (Table 1 and Figure 1): dissection of the aortic wall, resulting in the classic septum associated with an intimal tear (class 1), and the less common intramural hematoma-type dissection of the aortic wall in which the dissection is usually filled with blood clot without a detectable intimal tear1,3–5,7–10 (class 2). In patients with a classic intimal flap or septum, detection of the presence of aortic dissection by available imaging techniques is very accurate, with a reported sensitivity of 97% to 100% for both transesophageal echo (TEE) and MRI.1,3,8–18 O’Gara and DeSanctis4 stressed the importance of variants such as intramural hematoma (class 2) dissections and penetrating ulcer (class 4) aortic dissections and the difficulty of diagnosis in these classes. For patients with an intramural hematoma-type dissection, the sensitivity for detection of dissection is difficult to document accurately because it is not known how many patients are missed (false negatives) in the total denominator. Furthermore, noninvasive and invasive testing may overestimate the incidence of this type of dissection because a tear is often found at the time of surgery or autopsy.1

We have noted another variant of aortic dissection. This class 3 dissection is characterized by a stellate or linear intimal tear associated with exposure of the underlying aortic media or adventitial layers but without the progression and separation of the medial layers, resulting in extensive undermining of the intimal layers. Of particular concern is the fact that current imaging techniques may be inadequate for diagnosing this type of aortic dissection as demonstrated in our series because of a limited extent of undermining of the intimal layers and a minimal amount of blood in the dissected

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TABLE 1. Classification of Variants of Aortic Dissection

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Separation of intima/media; dual lumens (classic)</td>
</tr>
<tr>
<td>2</td>
<td>Intramural hematoma separation of intima/media; no intraluminal tear or flap imaged</td>
</tr>
<tr>
<td>3</td>
<td>Intimal tear without hematoma (limited dissection) and eccentric bulge</td>
</tr>
<tr>
<td>4</td>
<td>Atherosclerotic penetrating ulcer; ulcer usually penetrating to adventitia with localized hematoma</td>
</tr>
<tr>
<td>5</td>
<td>Iatrogenic/traumatic dissection</td>
</tr>
</tbody>
</table>

aortic wall. The inability to identify this third type of dissection may confer a dismal prognosis because without diagnosis treatment may be withheld, allowing progression of dissection, cardiac tamponade, or aortic rupture.

Methods

Of 181 consecutive patients referred to us for ascending or aortic arch repairs until June 30, 1997, there were 9 patients (9 of 181, 5%) in whom, despite multiple preoperative noninvasive and invasive studies (Table 2), the definitive diagnosis of aortic dissection was made during surgery. All 9 patients underwent ≥3 diagnostic studies, including TEE, CT, MRI, or aortography (Table 2). These studies were done preoperatively for ascending and/or aortic arch aneurysms associated with sudden onset of pain or aortic valve regurgitation.

Results

At the time of surgery, 9 patients were found to have aortic dissection despite negative imaging studies, with stellate or linear intimal tears identified. These were between 1 and 7 cm wide and exposed the deeper media and adventitia with limited undermining of the intimal layer (Table 2). In none of the patients was a classic type of flap or septum seen, nor was there an intramural hematoma type of dissection. In 6 of the 9 patients, aortic dissection had been suspected on the basis of clinical presentation but had never been proved preoperatively. The ascending aorta measured between 4.7 and 9 cm in diameter in these patients, and in 6 patients, there was a subtle eccentric bulge or bubble seen on aortography (Figures 1, 2, 3 and 4). In 7 patients, severe aortic valve regurgitation was also present. All 9 patients underwent operative repair of the aorta. Eight patients had a composite valve graft inserted by use of previously described techniques,1,18 and 1 patient had the aortic valve resuspended with tube graft repair of the ascending aorta. In addition, 3 patients had aortic arch repairs with deep hypothermic circulatory arrest, and 1 patient had an ascending aorta to abdominal aorta bypass.

None of the 9 patients with limited intimal tears who underwent surgery had postoperative complications, and 6 patients required no operative blood transfusions. The intraoperative findings are detailed in Table 2. Figure 5 shows an illustration of a limited aortic intimal tear. All patients had TEEs before discharge that showed good valve function with no regurgitation. All patients were 30-day survivors, and all were alive 13 to 75 months after surgery. In the larger series of 181 patients, 98% were 30-day survivors.

Discussion

Physicians usually request a TEE or CT scan of the aorta for patients who present with chest pain suggestive of aortic dissection.1 In our 9 patients, however, aortic dissection was not proven before surgery by any of the usual imaging techniques of TEE, CT scan, aortography, or MRI. The patients all had ascending aortic aneurysms, often with aortic regurgitation but without a separate intimal flap or septum to allow definitive diagnosis of aortic dissection. Furthermore, even careful retrospective review of the old preoperative studies failed to confirm aortic dissection by the usual criteria. The importance of this is that physicians who first see patients with classic symptoms of aortic dissection should be aware that current imaging techniques may not fully exclude the presence of subtle forms of aortic dissection despite reports of 97% to 100% sensitivity for detecting classic (class 1) aortic dissection.14,15 It should be noted that one third of patients with aortic dissection are not diagnosed as having aortic dissection before death.19 At least 3 of our patients would probably have died shortly from cardiac tamponade or rupture if they had not undergone surgery. Thus, if a physician encounters a patient with chest pain suggestive of aortic dissection but fails to diagnose aortic dissection, whether by TEE, CT, or MRI, and if an aneurysm, particularly if eccentric, or aortic valve regurgitation is found, aortography should be considered because the more subtle class 3 form of limited intimal aortic dissection may be present. This would be particularly important in patients at risk for aortic dissection such as those with Marfan syndrome, bicuspid aortic valve, coarctation of the aorta, Takayasu’s disease, Turner’s and Noonan’s syndromes, polycystic kidney disease, Ehlers-Danlos syndrome, and osteogenesis imperfecta; patients on corticosteroids; those with family histories of dissection; pregnant women; those who have had cardiovascular procedures; and patients with inflammatory

Figure 1. Classes of aortic dissection: class 1, classic dissection with flap between true and false aneurysm and clot in false lumen; 2, intramural hematoma; 3, limited intimal tear with eccentric bulge at tear site; 4, penetrating atherosclerotic ulcer with surrounding hematoma, usually subadventitial; 5, iatrogenic or traumatic dissection illustrated by coronary catheter causing dissection.
TABLE 2. Presentation, Studies, Findings, and Operative Procedure

<table>
<thead>
<tr>
<th>Age, y</th>
<th>Sex</th>
<th>Presentation</th>
<th>Studies/Findings</th>
<th>Operative Findings</th>
<th>Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>30</td>
<td>M</td>
<td>Neck pain, vomiting, dyspnea</td>
<td>TEE/anerysm, AR 4+, TEE/8.6-cm anerysm, AR 4+, MR/8.9-cm anerysm, coarctation</td>
<td>Bloody cardiac tamponade, acute ascending dissection, 3×5-cm intimal tear</td>
<td>Composite valve graft and 25-mm tube graft from composite graft to abdominal aorta</td>
</tr>
<tr>
<td>36</td>
<td>M</td>
<td>Marfan, chest pain, acute MI (CK-MB + Q waves)</td>
<td>TEE/anerysm</td>
<td>Bloody effusion, acute posterior 4×2-cm ascending aortic dissection tear (Figure 5), no intimal undermining</td>
<td>Composite valve graft</td>
</tr>
<tr>
<td>67</td>
<td>F</td>
<td>Lifting “heavy boulders,” chest pain</td>
<td>TEE/anerysm, AR 3+, TEE/5-cm anerysm, AR 3+</td>
<td>Anterior and posterior aortic dissection tears, valve commissure prolapse, 2- to 3-cm intimal undermining</td>
<td>Composite valve graft</td>
</tr>
<tr>
<td>67</td>
<td>F</td>
<td>CHF, fever of unknown origin 6 months before</td>
<td>TEE/AR 4+, MR 2+, 9-cm anerysm</td>
<td>Adhesions and scarred pericardium, partially healed 7×4-cm chronic ascending dissection, valve commissure prolapse, no intimal undermining</td>
<td>Composite valve graft and aortic arch repair</td>
</tr>
<tr>
<td>58</td>
<td>F</td>
<td>Aerobic exercise, chest pain</td>
<td>TEE/bicuspid valve, anerysm MR/anerysm</td>
<td>Acute small intimal tear ascending aorta</td>
<td>Composite valve graft</td>
</tr>
<tr>
<td>60</td>
<td>M</td>
<td>Marfan, mild chest pain, dyspnea</td>
<td>TEE/AR 4+, anerysm</td>
<td>Partially healed 1×2-cm intimal tear involving valve commissure</td>
<td>Composite valve graft</td>
</tr>
<tr>
<td>61</td>
<td>F</td>
<td>Marfanoid, CHF, chest pain</td>
<td>TEE/AR 4+, anerysm</td>
<td>Healed 10×3-cm intimal tear</td>
<td>Composite valve graft, aortic arch and descending “elephant trunk,” endarterectomy of innominate artery</td>
</tr>
<tr>
<td>61</td>
<td>M</td>
<td>Takayasu’s disease, chest pain for 6 years</td>
<td>3×TEE/AR 4+, ascending anerysm 6×MRI/ascending anerysm</td>
<td>Posterior ascending intimal tear involving valve commissures</td>
<td>Composite valve graft, hemiaortic arch, coronary bypass</td>
</tr>
<tr>
<td>37</td>
<td>M</td>
<td>Chest and neck pain</td>
<td>CT/4.5-cm ascending aorta TEE/pericardial effusion, AR 1+ Intraoperative TEE/cartic tamponade, AR 1+</td>
<td>Cardiac tamponade, acute dissection, 2½×2-cm intimal tear</td>
<td>Tube graft, valve resuspension</td>
</tr>
</tbody>
</table>

AR indicates aortic valve regurgitation; CATH+AG, selective coronary catheterization with simultaneous aortogram; MI, myocardial infarction; CK-MB, creatinine kinase myocardial band; Q, ECG Q waves; CAD, coronary artery disease; MR, mitral valve regurgitation; and CHF, congestive heart failure.

collagen vascular diseases. Although aortography is not as sensitive as other noninvasive tests in the initial diagnosis of classic (class 1) aortic dissection when the false lumen is fully thrombosed, it remains useful in patients with limited intimal tears.1,4,12,16,17 The eccentric bulge on aortography seen in patients with limited intimal tears, described in this series, is a subtle but important finding suggestive of a class 3 limited dissection. On retrospective review of available TEE studies, this eccentric bulging was not noted, although it would be expected to be present. The diagnosis of dissection may be missed if conventional diagnostic criteria that are dependent on the presence of an intimal flap are applied.

Typically, validation studies comparing the sensitivities and specificities of the various imaging modalities in the diagnosis of aortic dissection have depended critically on the identification of a classic dissection flap (septum) separating the aortic lumen into true and false channels. Although the reported sensitivities in these studies are very high, the diagnosis of dissection was based on a single “gold standard” modality, concordance between 2 of the imaging tests, or anatomic inspection in a limited number of patients who underwent surgery or autopsy.11,13,15,20 Lack of anatomic follow-up by surgery or autopsy of patients with negative tests may lead to underestimation of the true prevalence of
aortic dissection among those who are clinically suspected to have the condition, particularly in the detection of variants of dissection such as those without the classic flap, and overestimation of the sensitivity of diagnostic testing. Indeed, 6 of the 9 patients in our series had multiple negative or nondiagnostic tests that resulted in their being discharged from the hospital after initial studies but were found to have dissection on subsequent admissions.

There is unanimous agreement that in patients with aortic dissection involving the ascending aorta of the classic type (class 1) with a septum or an intraluminal flap, surgical repair is required in most cases to prevent death from rupture or cardiac tamponade. We and others believe that the intramural hematoma type of dissection (class 2) of the ascending aorta or aortic arch, increasingly diagnosed by TEE and MRI, should also be surgically treated. An intramural hematoma is often found intraoperatively to have only 1 short linear intimal entry tear without extensive destruction of the septum between the true and false lumens, and distal reentry sites are often absent. Thus, an intramural hematoma may exhibit a less pronounced displacement of the intimal septum layer from the outer adventitial layer with clot in the false lumen but has a prognosis just as lethal as that of a classic type of acute aortic dissection, as has been noted by others. The reason is that as with classic class 1 dissections, the adventitial layer is extremely thin and can rupture, or fluid seeps through the thin injured layers, resulting in cardiac tamponade from the exudate.

In patients with limited intimal tears (class 3), the very thin outer adventitial layer is also present and thus also is prone to rupture or to allow fluid to seep through it, resulting in cardiac tamponade, as happened in 3 of our patients. Before the aorta is opened, the area of the tear has the same external appearance as an acute dissection with either a subadventitial ecchymosis or visible flowing blood seen through the translucent adventitia. We also suspect that this limited type of dissection may propagate to become the classic type of aortic dissection, particularly in patients with Marfan syndrome, because we have seen older healed class 3 intimal tears associated with acute dissection. We have also reported these types of tears in 6.6%
It is not clear what determines whether a classic dissection with a septum, intramural hematoma, or limited intimal tear occurs. It is of interest that although elastic tissue loss and medial degeneration are found in most aneurysms, loss of smooth muscle cells is found mostly in patients with Marfan syndrome or acute dissection.1,2,4 TEE is usually considered the gold standard for detection of aortic dissection with a reported sensitivity of 97% to 100%.11–15 The aortic dissection may not be detected, however, if there is not extensive separation of the intimal layer resulting in a flap or septum or if there is only a slightly thickened wall with hematoma and clot occluding the tear site.7 MRI is reported to have a 98% to 100% sensitivity but suffers the same shortcomings.1,3,5,13–16 Nonetheless, MRI is considered more accurate in detecting intramural blood or blood clot in the class 2 intramural hematoma type of dissection. CT scanning with contrast is reported to have an 83% to 94% sensitivity for detection of aortic dissection1,5,11,13; however, an intimal tear is very rarely visualized on CT scan. Indeed, detection of the limited type of intimal tears that would require better resolution may be beyond the capability of current TEE, MRI, or CT studies. The newer techniques of spiral and helical CT, particularly 3-dimensional, shaded-surface-display angiography, will probably be much more accurate in detecting subtle forms of class III aortic dissection because the intimal layer of the wall is imaged in 3 dimensions and there is a greater likelihood of detecting sites of intimal aortic tears.27,28 Intra-aortic ultrasound has been found useful in the diagnosis of traumatic rupture of the aorta. This is of interest because subtle forms of intimal tears also need to be detected in this entity.29 It can also be combined with aortography, which was of value in this series of patients.

The classes of dissection described above refer to the form of dissection in the aortic wall, not the site of the intimal tear or the extent of the aorta involved.21,30–32 The extent of the dissection and the usual sites of the intimal tear are classified by the well-known DeBakey and Stanford classifications21,30–32 or by referring to the dissection as being proximal or distal to the left subclavian artery because the latter determines whether surgery is required.33

A potential weakness of this study is our inability to characterize with certainty the risk of the development of extensive dissection or rupture after development of a limited dissection that is not “imagable.” It is unlikely that a controlled study to do so can be designed from either a scientific or an ethical standpoint. Despite our inability to define the precise natural history of limited intimal dissection, several clinical guidelines emerge. All 9 patients had either a dilated ascending aorta (>4.5 cm) or frank aneurysm, and all had developed chest pain that had remained unexplained by multiple imaging modalities. Obstructive coronary artery disease was not present in these patients, a finding that argues against myocardial ischemia as a cause. Yet all patients at operation had evidence of a limited intimal aortic dissection that would have resulted in at least 3 patients’ immediate deaths if the aortas had not been repaired, and the remaining patients probably would have had limited long-term survival.

**Conclusions**

All current imaging modalities used in the diagnosis of aortic dissections are critically dependent on the presence and
identification of an intimal flap or separate true and false lumens. In this series, we describe an important variant of aortic dissection in which an intimal tear is present but without an intimal flap or hematoma. Therefore, this entity may elude most current imaging modalities yet have life-threatening consequences if unrecognized or untreated.

When a physician encounters a patient with classic symptoms of aortic dissection, including sudden chest pain radiating from the front of the chest to the back with or without hypertension, a TEE or CT scan should be performed, depending on which is available first. If these tests fail to detect aortic dissection and an ascending aortic aneurysm >4.5 cm in diameter is measured, particularly if either eccentric, or if the patient has Marfan syndrome, a bicuspid aortic valve, aortic valve regurgitation, or a pericardial effusion, then a second imaging test, preferably aortography, should be performed. The finding of an eccentric bulge on aortography should heighten the suspicion of this uncommon class 3 variant of aortic dissection. Ultimately, because intimal tears may be missed, patients with classic symptoms of aortic dissection who are confirmed to have ascending aortic aneurysms and aortic regurgitation or pericardial effusion should undergo urgent surgery because of the risk of rupture or cardiac tamponade from undetected aortic dissection. Because results with ascending and aortic arch repairs for acute dissection have improved and most centers report >90% survival rate for acute aortic dissection and ≥95% for elective ascending or aortic arch repairs, 20–25,33,34–36 we believe an aggressive operative approach offers benefit over clinical observation in this high-risk population.

References
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