Diagnosis and treatment of thoracic aortic intramural hematoma

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Purpose: This report reviews our recent experience with nine patients who had intramural hematoma of the thoracic aorta.
Methods: This was a retrospective study of all patients who had intramural hematoma at our institution from 1989 to 1994. Patients who had identifiable intimal flap, tear, or penetrating aortic ulcer were excluded from the study.
Results: Among these nine elderly patients (mean age, 76 years), the most common presentation was chest or back pain. Intramural hematoma was diagnosed by a variety of high-resolution imaging techniques. The descending thoracic aorta alone was involved in seven patients, whereas the ascending aorta was affected in the other two patients. One patient had evidence of an aneurysm (5.0 cm diameter) in the region of the hematoma. All patients were initially managed nonsurgically with blood pressure control. Both patients who had ascending aortic involvement had progression of aortic hematoma, which resulted in death in one case and in successful surgery in the other. Six of the seven patients who had descending aortic involvement alone were successfully managed without aortic surgery. The patient who had intramural hematoma and associated aortic aneurysm, however, had severe, recurrent pain and underwent successful aortic replacement. Another patient had recurrent pain associated with hypertension, but was successfully managed nonsurgically with antihypertensive therapy. All eight survivors are doing well at a median follow-up of 19 months.
Conclusions: Intramural hematoma appears to be a distinct entity, although overlap with aortic dissection or penetrating aortic ulcer exists. Aggressive control of blood pressure with intensive care unit monitoring has been our initial management. Patients who have involvement of the descending thoracic aorta alone can frequently be managed without surgery in the absence of coexisting aneurysmal dilatation or disease progression. Our experience suggests that a more aggressive approach with early surgery is warranted in patients who have ascending aortic involvement or those who have coexisting aneurysm and intramural hematoma. (J Vasc Surg 1996;24:1022-9.)

Improved imaging techniques have led to better definition of the range of diseases that can affect the thoracic aorta. Intramural hematoma (IMH) of the thoracic aorta only recently has been described in the literature as a distinct entity.[1,2] Fewer than 50 cases of IMH have been previously reported,[1-6] which is most likely due to the fact that cases of IMH have often been diagnosed as acute aortic dissection. It has become clear, however, that a variety of pathologic processes can affect the thoracic aorta. Classic aortic dissection with intimal flap, which often occurs in an aorta that does not have atherosclerosis, is well known. In the setting of severe atherosclerotic disease, development of localized penetrating atherosclerotic ulceration (PAU) has been described.[7,8] Although PAU or aortic dissection can be associated with a variable amount of aortic wall hematoma, IMH appears to be a separate entity,[1,2] characterized primarily by aortic wall hematoma without demonstrable intimal flap or penetrating ulceration. The paucity of clinical experience with IMH has meant that the optimal therapy for this condition is still undefined. We report herein our recent experience with thoracic aortic IMH in nine patients.

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MATERIALS AND METHODS

Patient identification and imaging methods. Our recent experience (1993 to 1994) with thoracic aortic IMH in eight patients was reviewed. A ninth

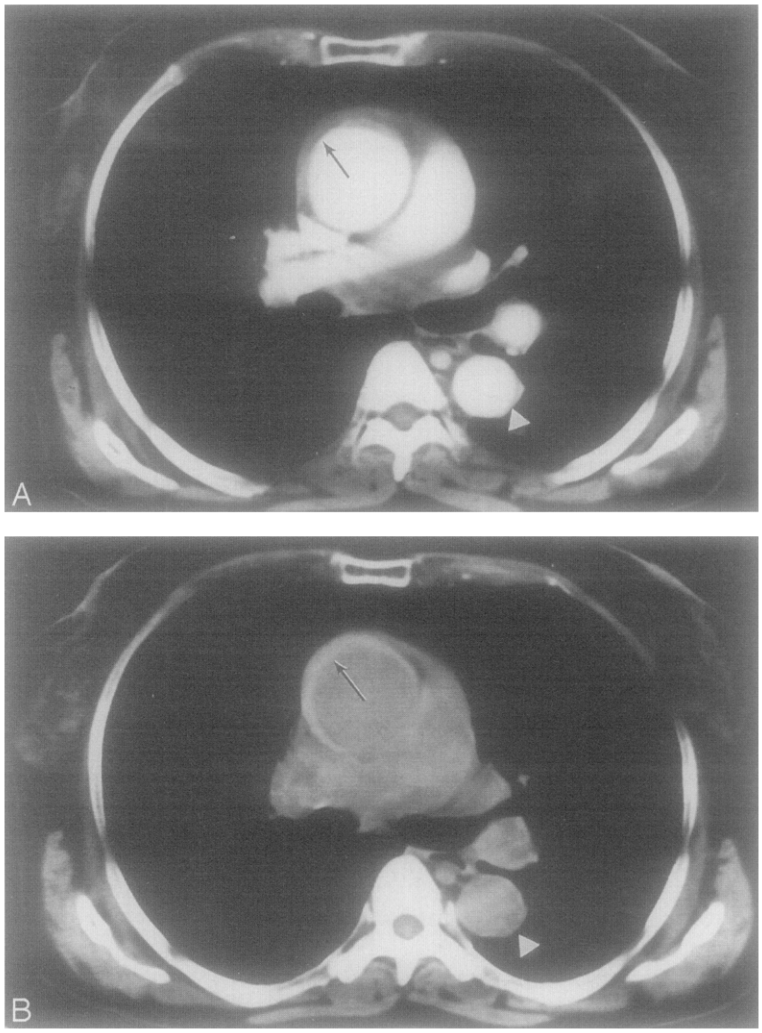

Fig. 1. Admission CT scan from patient #7 with **(A)** and without **(B)** intravenous contrast administration. A large hematoma surrounding ascending aorta is evident *(black arrow)*. Note that descending aorta appears normal at this time *(white arrowhead)*.

Table I. Demographic and clinical characteristics of patients with IMH

Patient	*Age*	*Sex*	*Htn*	*Diabetes*	*Cardiac disease*	*Renal dysfunction*	*Presenting symptom*	*CT*	*Angio*	*MRA*	*TEE*	*Site*
1	88	F	Severe	None	Moderate	None	Midscapular pain	+	+	−	−	Desc
2	80	F	Severe	None	Moderate	Moderate	Midscapular pain	+	−	+	−	Desc
3	81	M	Mild	None	None	None	Back and neck pain	+	−	+	−	Desc
4	76	M	Severe	None	None	None	Midscapular pain	+	+	+	+	Desc
5	71	M	Severe	Mild	None	Mild	Left chest and flank pain	−	−	+	−	Desc
6	77	F	Mild	None	None	None	Syncope	+	+	−	−	Asc
7	65	F	None	None	None	None	Left chest and neck pain	+	+	+	−	Asc
8	64	F	Mild	None	None	None	Midchest pain	+	+	−	−	Desc
9	69	F	Moderate	None	Moderate	None	Asymptomatic	+	−	+	−	Desc

Htn, Hypertension; *CT,* computerized tomography; *Angio,* conventional angiography; *MRA,* magnetic resonance angiography; *TEE,* transesophageal echocardiography; *Desc,* descending thoracic aorta; *Asc,* ascending aorta.
Risk factors graded as per Methods section.

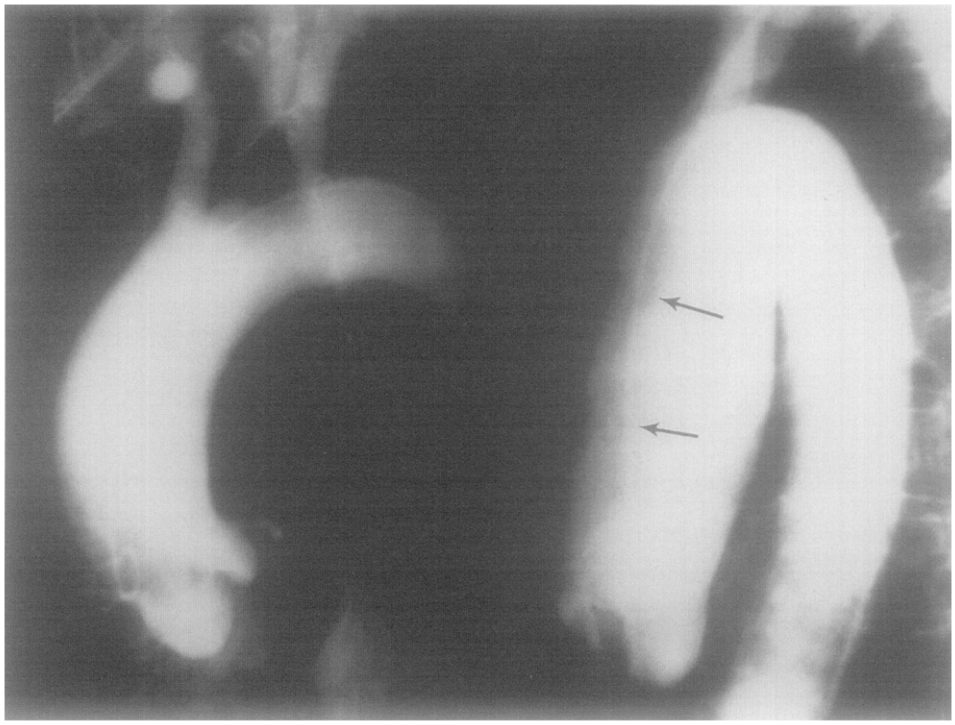

Fig. 2. Admission angiogram from patient #7 shows no evidence of intimal defect. *Arrows* indicate overlying lung shadow.

patient included in the study has been observed for this condition since 1989. In all nine patients, a hematoma of the thoracic aorta was identified, but high-resolution angiographic imaging failed to show an intimal tear, disruption, flap, or penetrating ulcer of the aorta. Computed tomographic (CT) examinations for all patients were performed with contiguous slices (no gap between slices), and each image spanned a thickness of 1 cm. All contrast aortograms were performed with a minimum of two projections, but four projections were used in three of the five cases (patients #1, #4, and #7 in Table I) in which the diagnosis was made by this method.

Data collection. Demographic data, including risk factors and associated diseases (hypertension, diabetes, cardiac, and renal disease) were classified and graded as mild, moderate, or severe according to the suggested standards for reports dealing with lower extremity ischemia as formulated by the Ad Hoc Committee on Reporting Standards of the SVS/ISCVS-NA.[9] The diagnosis of IMH was established by a review of imaging studies by one of the authors, a vascular radiologist. Information regarding the clinical course and outcome of the study patients was determined by a review of the hospital and office records and by telephone communication with surviving patients.

RESULTS

Table I summarizes demographic and clinical features in these patients. There were three men and six women, ranging in age from 64 to 88 years (mean, 76 years). All except one patient had hypertension, which was severe in four patients. The presence of other medical conditions was typical for patients of this age group. Seven of the nine patients had back, chest, or flank pain; another patient had syncope alone. A final patient was asymptomatic, the lesion having been diagnosed after workup for an abnormal routine chest roentgenogram. CT was used for diagnosis in eight of the nine patients, but in every case the diagnosis of IMH was definitively made only after either conventional contrast angiography or magnetic resonance angiography was performed. A single patient (#8) had associated aneurysmal dilatation (5.0 cm diameter) of the aorta in the region of the IMH.

In two patients the site of the IMH was the ascending aorta, whereas in the other seven the descending thoracic aorta was involved. The two pa-

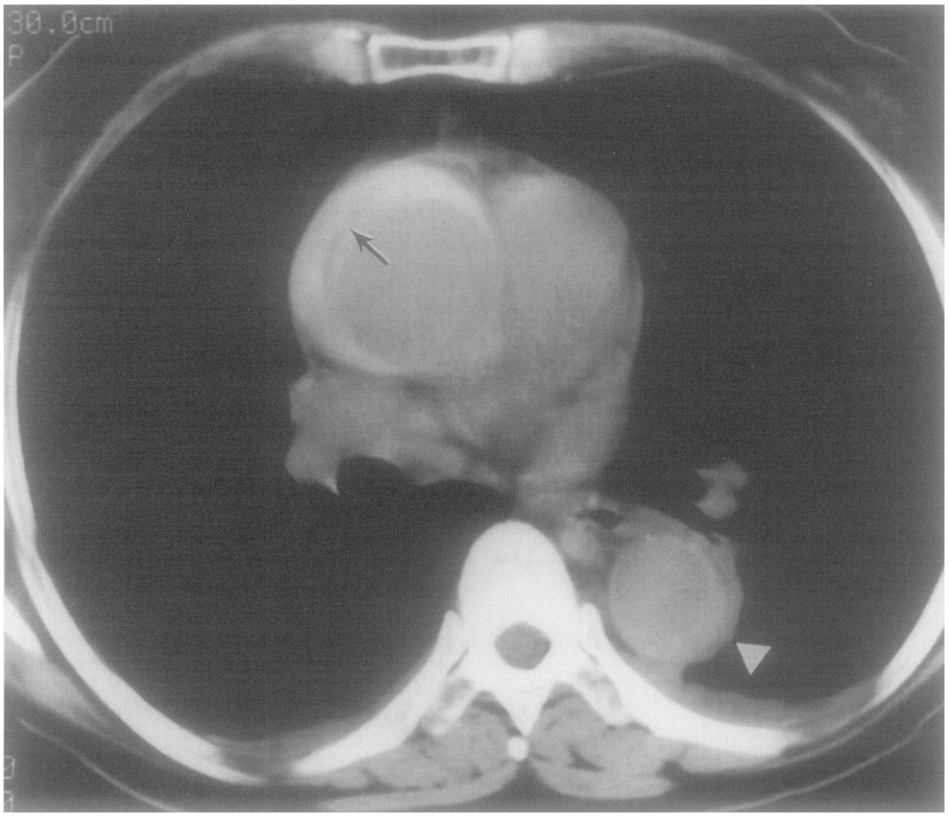

Fig. 3. Follow-up CT scan of patient #7 on hospital day 4 shows interval increase in size of ascending aortic IMH *(black arrow)*, as well as extension of hematoma to descending thoracic aorta *(white arrowhead)*.

tients who had ascending aortic involvement had no or mild hypertension, whereas five of the seven patients who had descending aortic IMH had moderate or severe hypertension.

Representative imaging studies are shown in Figs. 1 through 5. Fig. 1, the presenting CT in patient #7, shows hematoma surrounding the ascending aorta. On the angiographic evaluation (Fig. 2), no intimal defect could be demonstrated. After 4 days of aggressive antihypertensive therapy in the intensive care unit (ICU), a follow-up CT scan (Fig. 3) showed a larger hematoma around the ascending aorta, as well as distal extension of the IMH. The patient was urgently taken to surgery, where replacement of the ascending aorta was performed uneventfully. No intimal defect was seen at surgical exploration, confirming the angiographic findings.

A representative CT panel from patient #2 shows IMH of the descending thoracic aorta (Fig. 4), associated with a pleural effusion. Because the IMH originated in the arch, and given the patient's overall condition and risk of arch replacement, conservative therapy was elected in this patient. Fig. 5 shows a CT image from patient #8, who had IMH in association with dilatation (5.0 cm diameter) of the descending thoracic aorta.

The hospital course and outcome data for the patients who had IMH are displayed in Table II. All of these patients were treated with a regimen of anti-hypertensive therapy. One of the patients (#9) was treated as an outpatient because of her clinical stability and asymptomatic presentation, and the remainder were treated with antihypertensive medications in an ICU setting.

Of the seven patients who had descending thoracic IMH, six were managed without surgery. The one patient (#8) who had aneurysmal aortic dilatation was initially treated medically, but because of recurrent severe pain 10 days after the initial episode, she underwent surgical replacement of the descending thoracic aorta. At surgery, she was found to have a small ulcer in the proximal descending aorta (above the aneurysm), which appeared to be the source of the IMH. Patient #2 had recurrent pain in the setting

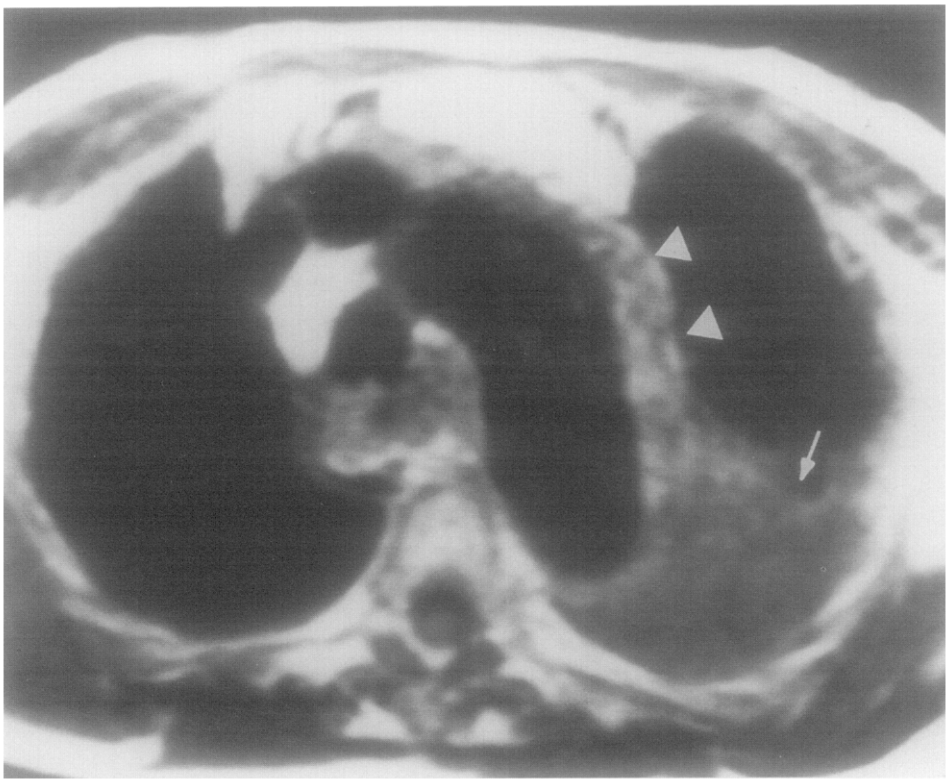

Fig. 4. Admission CT image of patient #2 demonstrates IMH of proximal descending thoracic aorta *(arrowheads)*, with accompanying pleural effusion *(small arrow)*.

of severe hypertension, but this subsided with blood pressure control. Bloody pleural effusions (confirmed by pleural tap) developed in two patients (#2 and #5), but because of their clinical stability and overall high surgical risk, surgery was deferred. Both patients who had renal insufficiency at presentation (#2 and #5) required surgical intervention (nephrectomy) or radiologic intervention (renal artery dilatation) to control renovascular hypertension. All seven patients who had descending thoracic IMH had favorable short-term outcomes and were well at follow-up. Both patients who had ascending aortic IMH had progression despite receiving medical therapy. One died of cardiac tamponade before surgery could be performed, and the other was successfully treated with ascending aortic replacement for extension of the IMH. Among the hospitalized patients who survived (n = 7), the median hospital stay was 14 days (range, 0 to 103 days; mean, 23 days). All eight surviving patients are well at a median follow-up of 19 months (range, 7.3 to 70 months; mean, 24 months). Serial imaging studies have shown stable or resolved IMH in these patients at a median follow-up of 11 months (range, 6.4 to 65 months; mean, 13.5 months).

DISCUSSION

A large variety of acute, life-threatening diseases can affect the thoracic aorta. Although aortic dissection that is associated with primary intimal disruption has been extensively described, other conditions that have similar clinical presentation, including PAU and IMH (Table III), have only been recognized more recently. The hallmarks of classic aortic dissection are intimal flap and false lumen. Although aortic dissection can be seen in the atherosclerotic aorta, it is frequently seen in aortae that are relatively free of atherosclerosis. Roberts[10] actually found that atherosclerotic plaque hindered the dissection process, which is possibly related to the transmural inflammation and relative fusion of aortic wall layers. In contrast, PAU is characterized by deep ulceration of the aortic wall in the setting of extensive atherosclerosis.[7,8] Both aortic dissection and PAU can be accompanied by aortic wall hematoma, but IMH, as defined by us and others,[1-6,11] occurs without radiographi-

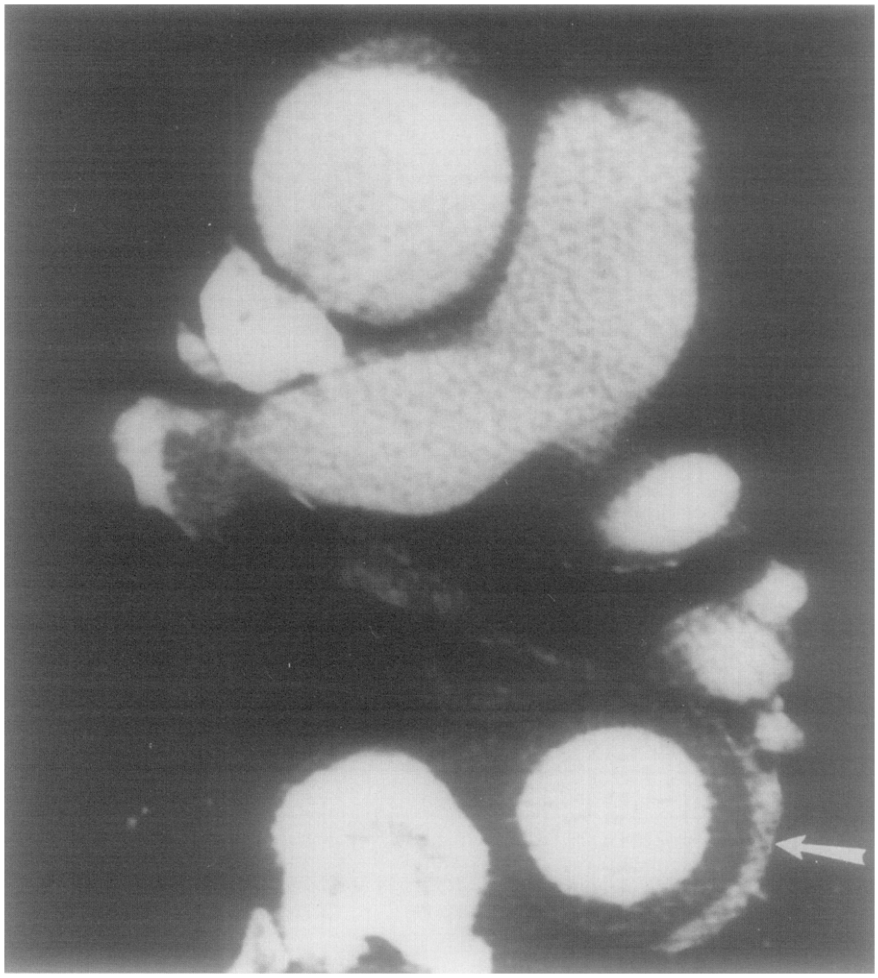

Fig. 5. Admission CT image of patient #8 demonstrates IMH of mid-descending thoracic aorta *(arrow)* associated with aneurysmal dilatation of aorta (5.0 cm diameter).

cally demonstrable intimal flap or penetrating ulcer. The term "aortic dissection without intimal defect" has been frequently applied to cases that would be classified as IMH by our definition.

Notwithstanding this classification, we speculate that some cases of IMH originate from ulceration in an atherosclerotic aorta. For example, at surgery to replace her descending thoracic aorta, patient #8 was found to have a small ulcer in the proximal descending aorta, which appeared to be the origin site of the IMH. This ulcer had not been seen on preoperative studies, including the contrast angiographic study. In addition, the patient's pathologic characteristics differed from cases of PAU[7] in that her hematoma was not localized to the site of the ulcer. In contrast to the case of patient #8, no intimal defect was identified as the cause of the IMH either before surgery or at surgery for patient #7. To explain such cases, Yamada et al.[1] speculated that IMH can result from the rupture of vasa vasorum of the aortic wall. Thus the cause of IMH appears to be multifactorial.

It is reasonable to consider whether the distinctions between aortic dissection, PAU, and IMH are clinically meaningful or merely semantic. A certain amount of overlap exists because the lesions occur in similar locations, they tend to occur in hypertensive patients, and they have some common pathologic features (Table III). Despite such overlap, we believe that these are distinct pathologic entities that can be separated from one another by using high-resolution imaging techniques. We propose that distinguishing among them is not merely an issue of semantics, but instead is likely to lead to better management of each.

Anecdotal data suggest that IMH can predispose to classic intimal flap in some cases, perhaps by weakening the aortic media and overlying intima. For

Table II. Clinical course and outcome of patients with IMH

Patient	*Site*	*Treatment*	*Course*	*Stay (days)*	*Short-term outcome*	*Follow-up (months)*	*Status at follow-up*
1	Desc	ICU	Brief period of congestive heart failure requiring intubation	19	Good	20	Good
2	Desc	ICU	Recurrent pain associated with uncontrolled hypertension; left nephrectomy performed for renovascular hypertension	103	Good	20	Good
3	Desc	ICU	Uncomplicated	3	Good	16	Good
4	Desc	ICU	Mild renal insufficiency	13	Good	70	Good
5	Desc	ICU	Renal insufficiency, uncontrolled hypertension; responded to percutaneous renal artery dilatation	14	Good	18	Good
6	Asc	ICU	Cardiac tamponade	2	Died	n/a	n/a
7	Asc	ICU, surgery	Developed extension of IMH while on aggressive antihypertensive therapy; taken to surgery for ascending aortic replacement	27	Good	27	Good
8	Desc	ICU, surgery	Initial nonoperative therapy undertaken, but recurrent severe pain led to surgical replacement of descending thoracic aorta	22	Good	7	Good
9	Desc	Outpatient	Uncomplicated	0	Good	17	Good

Desc, Descending thoracic aorta; *Asc,* ascending aorta.

Table III. Characteristics of thoracic aortic AD, PAU, IMH

	AD	*PAU*	*IMH*
Defining features	Intimal flap; false lumen	Large ulcer penetrating internal elastic lamina	Aortic wall hematoma
Degree of atherosclerosis	Variable (often minimal)	Always severe	Variable
Aortic wall thrombus	Possible (if false lumen thromboses)	Localized hematoma possible	Yes
Extent of lesion	Usually extensive	Focal	Usually extensive

example, on a magnetic resonance angiographic scan patient #7 in this report was found to have a typical intimal flap in the descending aorta 2 weeks after she underwent surgical replacement of the ascending aorta. As noted, she initially had ascending aortic IMH and a normal descending aorta, and subsequent extension of the IMH into the descending aorta led to the operative intervention (Figs. 1 through 3). It seems likely that the intimal flap developed secondarily, after the IMH extension into the descending aorta. Such a sequence has been reported by others.[2,4,5]

With wider recognition of IMH as a distinct entity, and because of the increasing availability of high-resolution imaging methods of the thoracic aorta, more cases of IMH are likely to be identified. Treatment will remain empiric until more experience is gained. The patients reported herein were generally managed initially with aggressive antihypertensive therapy in an ICU setting.

Both patients who had ascending aortic IMH had progression of their lesion despite medical therapy. One died of acute cardiac tamponade as a result of aortic rupture, and the other underwent urgent ascending aortic replacement because of extension of the IMH (Figs. 1 through 3). Interestingly, these two patients had no or mild hypertension (Table I). This suggests, at least in some cases, that hypertension may not be a major contributor to the development of ascending aortic IMH. This may explain the lack of clinical response to aggressive blood pressure control. Our experience with ascending aortic IMH is similar to that of Robbins and associates,[2] who reported that all three patients in their series who had ascending aortic IMH required surgery after initial medical therapy (two of the three patients died). Thus we suggest that the presence of ascending aortic IMH may be an indication for early surgery. This approach is consistent with the widely-accepted approach of early surgery performed for aortic dissection involving the ascending aorta. The ascending aorta may be more prone to rupture because of its high elastin/

collagen ratio, or because it is exposed to a larger rate of increase in aortic pressure in comparison with the descending aorta.

In contrast, six of the seven patients in this report who had descending thoracic aortic IMH were managed without aortic replacement. Interestingly, the development of bloody pleural effusion in two patients was not a harbinger of aortic rupture. Both patients in whom pleurocentesis-confirmed bloody effusions developed were managed without aortic replacement. We speculate that the bloody effusions developed from microperforations in the outer wall of the IMH.

The one patient with descending thoracic IMH who required surgery (#8) had both IMH and aneurysmal dilatation. Although this patient was initially managed without surgery, the recurrence of severe pain in the setting of good blood pressure control prompted surgical intervention. Our experience with this patient is akin to our reported series of patients in whom aortic dissection develops in the setting of aneurysm.[12] In that study, we found that the coexistence of acute dissection and aneurysm increased the risk of aortic rupture. On the basis of this experience and the clinical course of patient #8, we propose that the combination of IMH and aneurysm should be managed with aortic replacement.

Our overall experience with descending thoracic IMH is similar to that of Robbins et al.,[2] who reported that eight of their 10 patients who had descending aortic IMH were managed without aortic replacement and that all 10 patients survived. Thus it appears that descending aortic IMH may be more appropriately treated with aggressive antihypertensive therapy as the sole method in a majority of cases, and surgical intervention should be reserved for patients in whom aneurysmal dilatation coexists or in whom the lesion is seen to be progressing on serial studies.

Mohr-Kahaly and associates[6] suggest a more ominous prognosis in IMH patients than we have noted (a 47% mortality rate compared with the 13% mortality rate in our current series). Angiographic confirmation of the lack of intimal tear, however, was not documented in the European report, and it is therefore unclear whether those patients are directly comparable with the patients in the group we have reported. Furthermore, the outcome data in the study by Mohr-Kahaly et al. were not stratified according to IMH location, which precludes direct comparison of results. Nevertheless, their data underscore the need for close surveillance of patients who have IMH, including initial ICU monitoring, aggressive blood pressure control, and frequent serial follow-up imaging studies.

As IMH is more widely recognized as an entity distinct from aortic dissection, more clinical experience will be gained. Such experience will be needed to support the management recommendations that are put forth above, especially in regard to the possible differential approach to ascending and descending aortic IMH. Further studies and longer follow-up observation of the natural history of IMH will also be needed to determine the optimal imaging method and frequency for the follow-up of patients who have IMH.

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