

Pictorial Essay

Pathogenesis in Acute Aortic Syndromes: Aortic Dissection, Intramural Hematoma, and Penetrating Atherosclerotic Aortic Ulcer

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Acute aortic syndromes refer to the spectrum of aortic emergencies that include aortic dissection, intramural hematoma, penetrating atherosclerotic ulcer of the aorta, aortic aneurysm leak and rupture, and traumatic aortic transection. The aortic wall is composed of three layers (Fig. 1): the inner layer of intima, the middle layer of media, and the outer layer of adventitia. Multiple mechanisms are involved in the disruption of the aortic wall layers, leading to various acute aortic syndromes. This pictorial essay focuses on the distinction of a typical aortic dissection from an intramural hematoma and penetrating atherosclerotic ulcer.

Aortic Dissection

A classic aortic dissection begins with a laceration of the aortic intima and inner layer of the aortic media, forming an entrance tear that allows entering blood to split the aortic media [1]. The splitting of the media is responsible for formation of a double-channel aorta, with an aortic dissection flap dividing the aortic lumen into true and false lumens (Figs. 2 and 3). The intima and the inner part of the aortic media form the intimomedial flap. The flap tissue is composed mainly of aortic media delaminated from the aortic

wall [2]. The outer portion of the aortic media and adventitia form the outer wall of the false channel. Reentrance tears are usually present in the intima, creating additional communication between the true and false lumens in the distal aorta. The true lumen is usually small with high-velocity flow, whereas the false lumen is larger with slower velocity, turbulent blood flow (Fig. 4).

Cystic medial necrosis associated with connective tissue disorders was once believed to contribute to degeneration of the aortic media leading to aortic dissection. However, a study showed that a minority of patients with aortic dissection exhibited medial degeneration [3]. In most patients, the primary event that allowed the blood to spread through the aortic media was the intimal tear. When present, degenerative changes

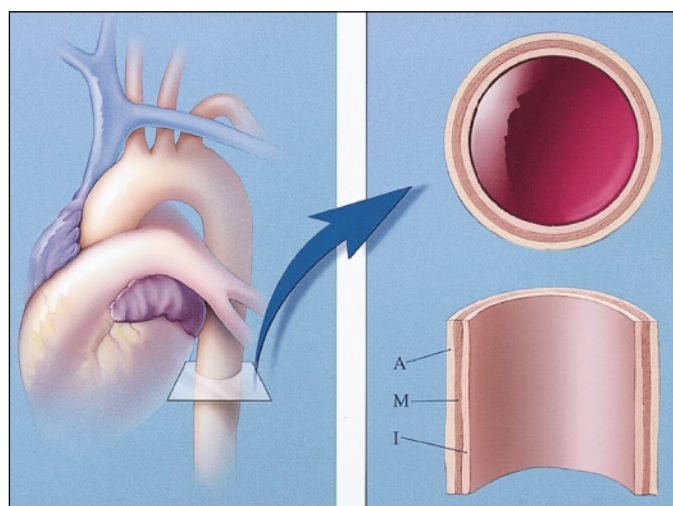


Fig. 1.—Diagram shows three layers of normal aortic wall, from inner to outer: intima (I), media (M), and adventitia (A).

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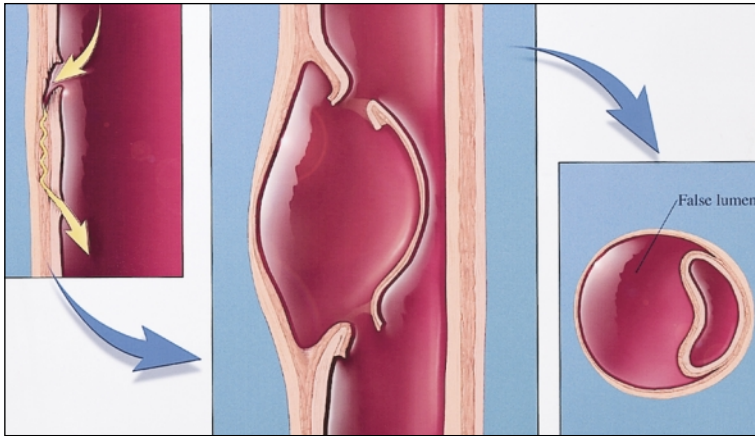


Fig. 2.—Diagram illustrates events leading to aortic dissection from formation of entrance tear and exit tear of intima to splitting of aortic media and formation of intimal flap. Blood under pressure dissects media longitudinally, and double-channel aorta is formed with blood filling both true and false lumens.

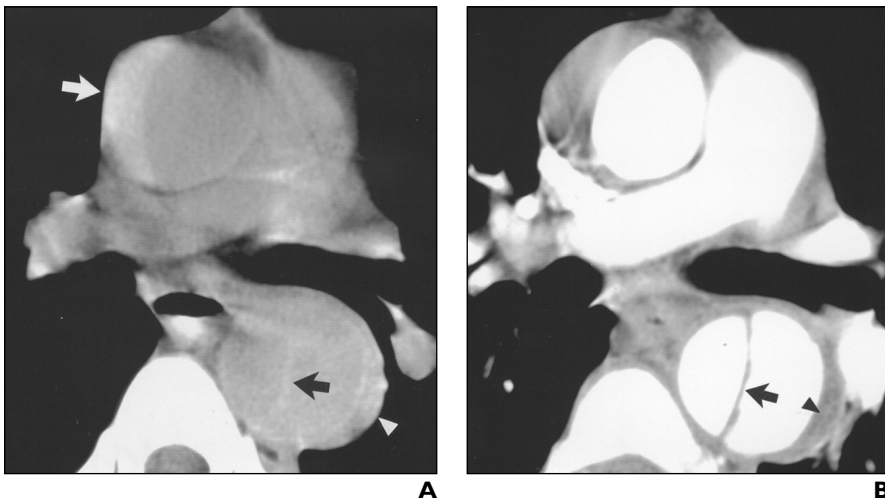


Fig. 3.—46-year-old man with concurrent intramural hematoma involving ascending aorta and communicating dissection involving descending aorta.

A, Axial unenhanced CT scan shows hyperdense crescentic hematoma in wall of ascending aorta (white arrow) with eccentric narrowing of lumen, type A intramural hematoma. Small intramural hematoma (arrowhead) is also noted at left lateral aspect of proximal descending aorta. High-attenuation dissection flap (black arrow) is seen in descending aorta.

B, Axial contrast-enhanced CT scan obtained at same level as **A** shows wall thickening in ascending and descending aorta, but high-attenuation intramural hematoma is less obvious. Classic intimal flap (arrow) dividing true and false lumens in descending aorta is more conspicuous after contrast administration. Note irregular margin of flap on false lumen side. Intramural hematoma (arrowhead) is seen along lateral wall of false lumen.

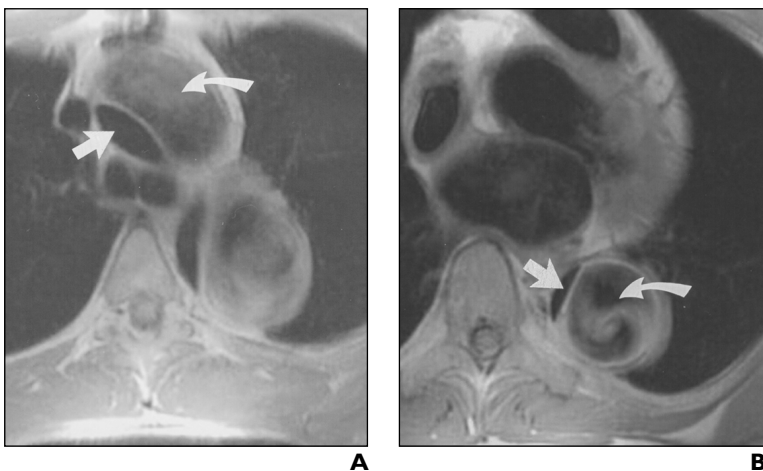


Fig. 4.—Axial double-inversion-recovery MR images (TR/TE, 1875/18; inversion time, 150 msec) of 37-year-old man with Marfan syndrome.

A, Image shows classic aortic dissection with double-channel aorta. True lumen (straight arrow) is smaller than false lumen (curved arrow). High-velocity flow in true lumen causes signal void. Slower flow with higher signal can be seen in false lumen.

B, Image shows swirling flow pattern in false lumen (curved arrow). True lumen (straight arrow) is significantly narrowed but patent.

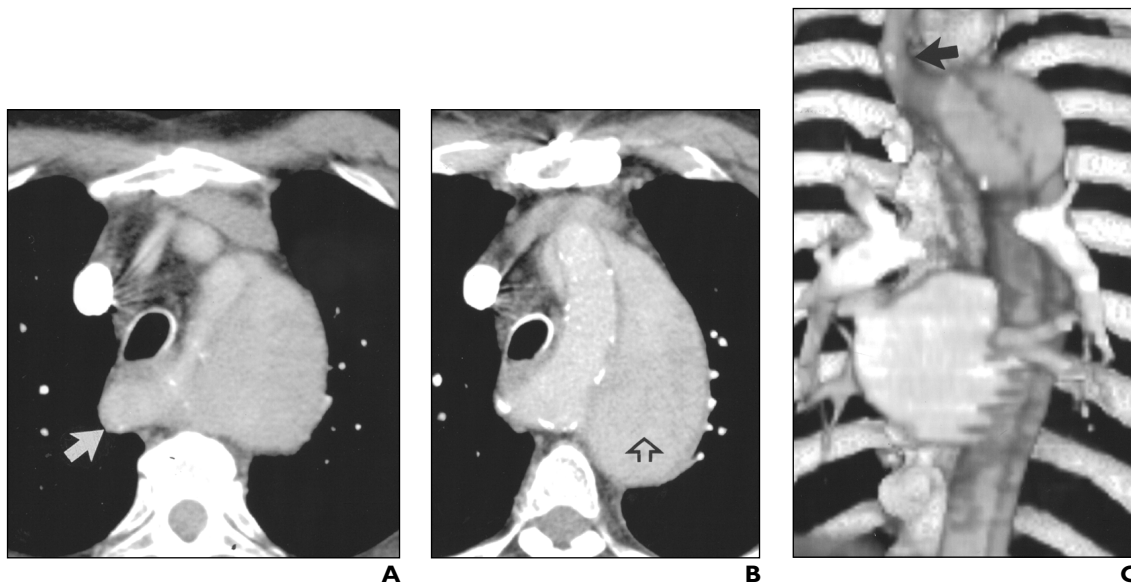


Fig. 5.—68-year-old man with aberrant right subclavian artery and horseshoe kidney.

A, Axial contrast-enhanced CT scan obtained at level of origin of aberrant right subclavian artery shows aberrant vessel (*arrow*) crossing midline behind trachea and esophagus. **B,** Axial contrast-enhanced CT scan shows dissection involving aortic arch with calcifications within intimal flap and different attenuation of enhanced blood within true and false (*arrow*) lumens. Intimal tears leading to dissection frequently form in areas of elevated hydraulic stress, such as region of aberrant vessel origin. **C,** Anteroposterior volume-rendered CT image of origin of aberrant subclavian artery depicts aberrant vessel course (*arrow*) better than axial scans **A** and **B**.

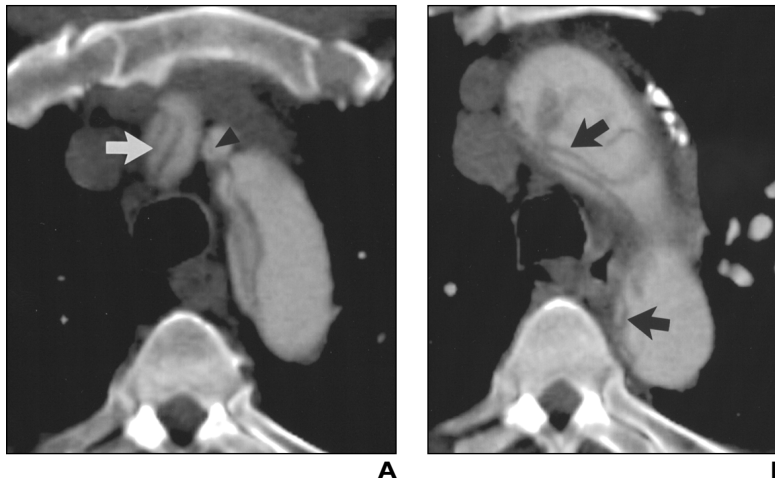


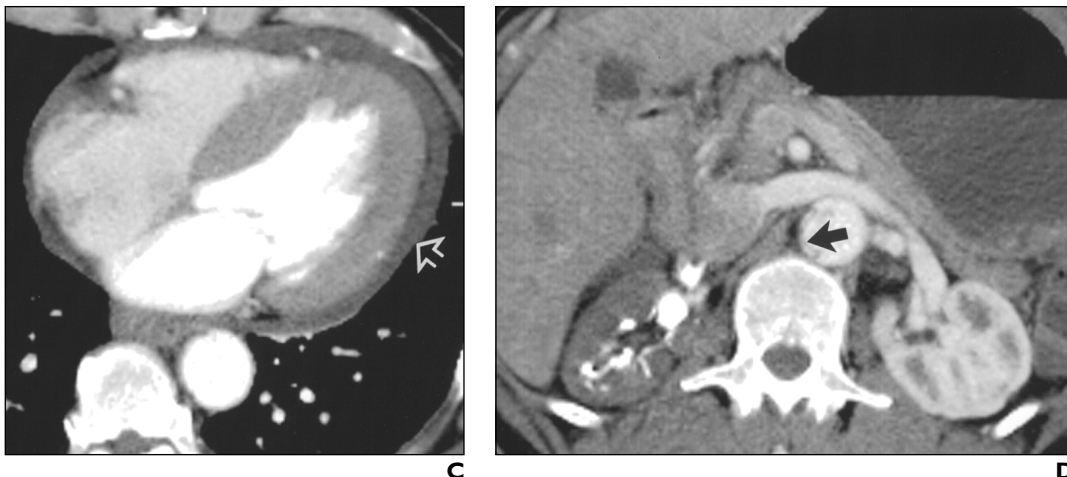
Fig. 6.—61-year-old man with symptoms of right hemispheric stroke who was found to have marked blood pressure discrepancy between arms and hypertension. Urgent CT scan (not shown) revealed type A aortic dissection. Patient went into asystole and died 15 hr after imaging.

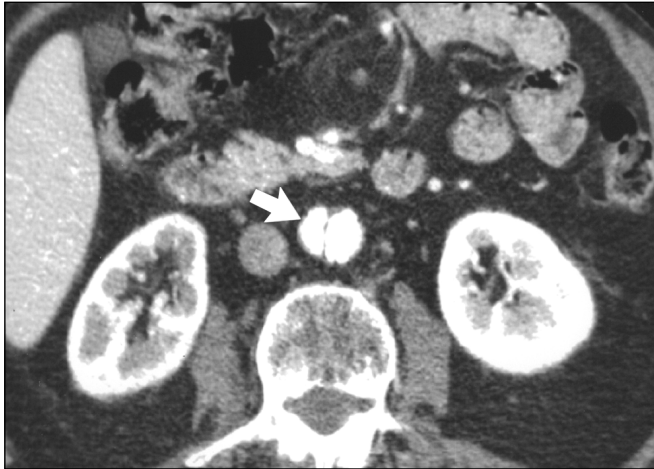
A, Axial contrast-enhanced CT scan obtained at level of aortic arch shows complex dissection with intimal flap involving arch and brachiocephalic artery (*arrow*). Dissection extended into left common carotid artery (*arrowhead*) and into left subclavian artery (not shown).

B, Axial CT scan shows irregular dissection flap within lumen of ascending and descending aorta (*arrows*).

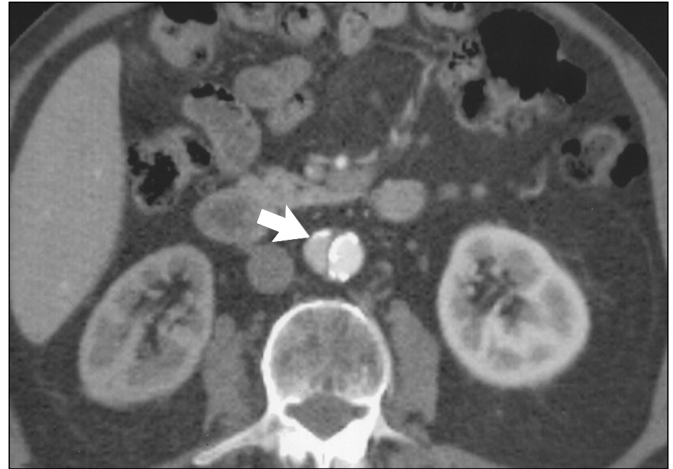
C, Axial CT scan shows hemopericardium (*arrow*) that was confirmed at echocardiography (not shown) as large circumferential hyperechoic pericardial effusion with evidence of right ventricle compression.

D, Axial CT scan shows dissection continuing along right wall of abdominal aorta (*arrow*). No enhancement of right kidney parenchyma was present.

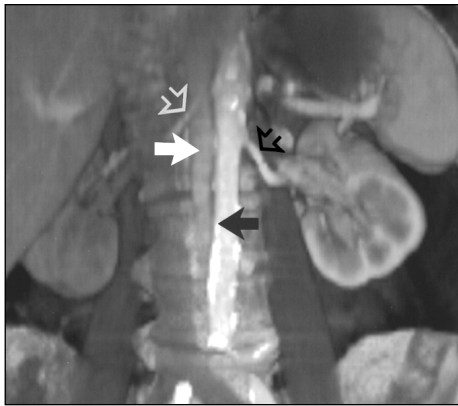




A



B



C

Fig. 7.—82-year-old man with thoracic aortic aneurysm type B and thoracoabdominal aortic dissection extending from just distal to left subclavian artery to proximal right common iliac artery. Patient was first diagnosed with aortic dissection 12 years ago. For more than 10 years, symmetric perfusion of kidneys was seen, until recently when CT showed hypoperfusion of right kidney.

A, Contrast-enhanced CT scan shows both true and false (*arrow*) lumens to be well opacified with contrast material. There is minimal delay in enhancement and thinning of cortex of right kidney.

B, CT scan obtained 1 year after **A** shows decrease in attenuation of contrast-enhanced blood in false lumen (*arrow*) when compared with true lumen. Enhancement of right kidney is markedly diminished, which is compatible with progressive hypoperfusion.

C, Anteroposterior volume-rendered CT image shows right renal artery (*open white arrow*) originating from false lumen (*solid white arrow*). Left renal artery (*open black arrow*) originates from true lumen. Note dissection flap with calcifications (*solid black arrow*) that separates true and false lumens.

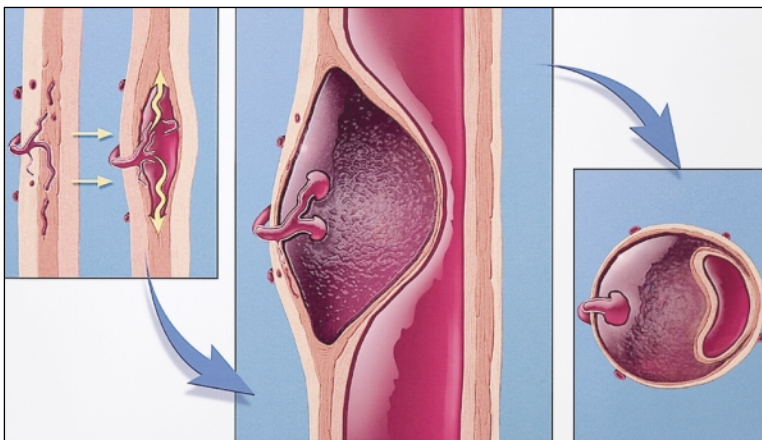
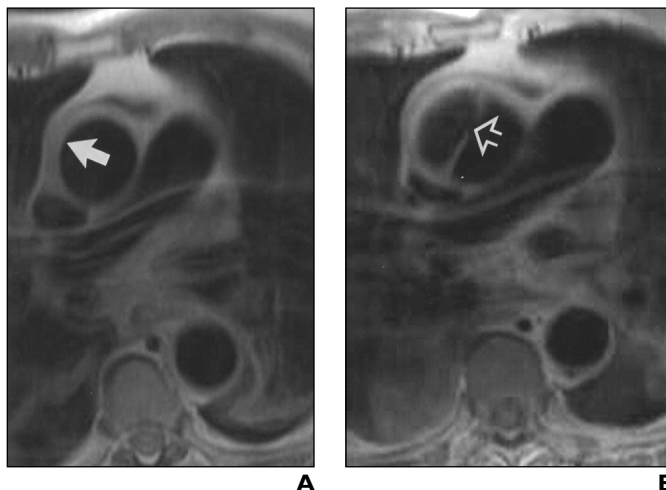


Fig. 8.—Diagram shows events leading to intramural hematoma, from rupture of vasa vasorum feeding aortic media to creation of intramural hematoma with intact intimal layer.

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Fig. 9.—Axial double-inversion-recovery MR images (TR/TE, 1690/29; inversion time, 150 msec) of 76-year-old man with progression of intramural hematoma to overt dissection in ascending aorta within 6 days.

A, Image shows high-signal-intensity crescentic intramural collection in ascending aorta (*arrow*), consistent with early subacute type A intramural hematoma. **B,** Image obtained 6 days after **A** shows that intramural hematoma progressed to type A aortic dissection within 6 days. Note signal intensity difference between true and false lumens. Signal void within true lumen reflects high-velocity blood flow, whereas higher signal within false lumen is related to slower, turbulent flow. Also note defect in intimomedial flap (*arrow*) representing intimal tear.



within the media and the loss of the elastic tissue reduce the resistance of the aortic wall to hemodynamic stress, leading to subsequent dissection. Hypertension-related spontaneous rupture of the aortic vasa vasorum might lead to intramural hematoma and subsequently to intimal tear. Intramural hematoma precedes intimal rupture because hemorrhage of the vasa vasorum weakens the media, and the arterial pressure from blood flow in the aortic lumen subsequently favors the entrance of blood from the lumen into the aortic media [1]. Atherosclerosis was once thought to cause aortic dissection. However, there is an association between an atheroma and the location of dissection in only a small number of patients [1]. Dissection in the region of gross atherosclerosis is usually limited by neighboring fibrosis and calcification.

Mechanical forces contributing to aortic dissection include flexion forces of the vessel

at fixed sites, the radial impact of the pressure pulse, and the shear stress of the blood. During the cardiac cycle, the heart and aorta produce rhythmic movements, allowing all but fixed segments to move. These fixed points of the aorta are exposed to the most significant flexion forces. Classic type A and B aortic dissections produce an intimal tear at the areas of greatest hydraulic stress: the right lateral wall of the ascending aorta or the descending aorta in proximity to the ligamentum arteriosum (Fig. 5). Hypertension adds to a mechanical strain on the aortic wall and the shearing forces exerting a longitudinal stress along the aortic wall (Figs. 6 and 7). Decreased vasa vasorum flow, occurring in arterial hypertension, may increase the stiffness of the outer ischemic media of the aorta to produce interlamellar shear stresses contributing to the development of aortic dissection.

Aortic Intramural Hematoma

Aortic intramural hematoma may occur as a primary event in hypertensive patients in whom there is spontaneous bleeding from vasa vasorum into the media or may be caused by a penetrating atherosclerotic ulcer. Intramural hematoma may also develop as a result of blunt chest trauma with aortic wall injury. Intramural hematoma is thought to begin with the rupture of the vasa vasorum, the blood vessels that penetrate the outer half of the aortic media from the adventitia and arborize within the media to supply the aortic wall (Fig. 8). The hematoma propagates along the media layer of the aorta [2]. Consequently, intramural hematoma weakens the aorta and may progress either to outward rupture of the aortic wall or to inward disruption of the intima, the latter leading to communicating aortic dissection [4] (Fig. 9).

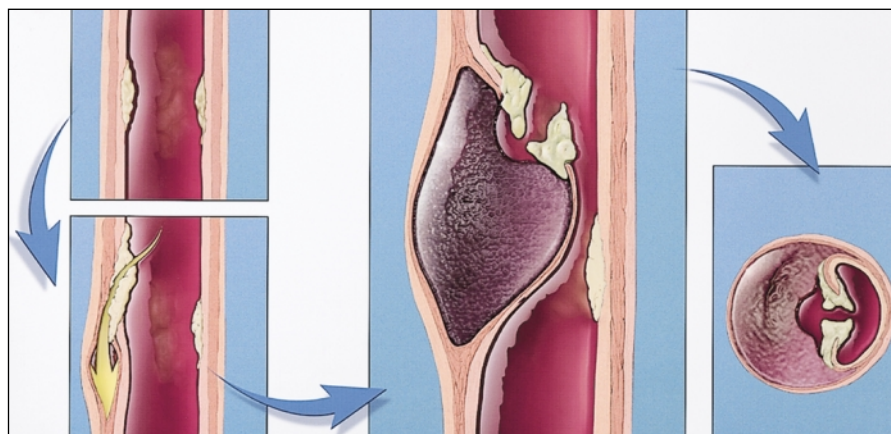


Fig. 10.—Diagram shows events leading to penetrating aortic ulcer from formation of extensive aortic atheroma confined to intimal layer, through lesion progression to deep ulceration of plaque with penetration into media, to entrance of blood from aortic lumen into media and splitting of media with intramural hematoma. Hematoma formation may extend along media, resulting in long-segment intramural hematoma.

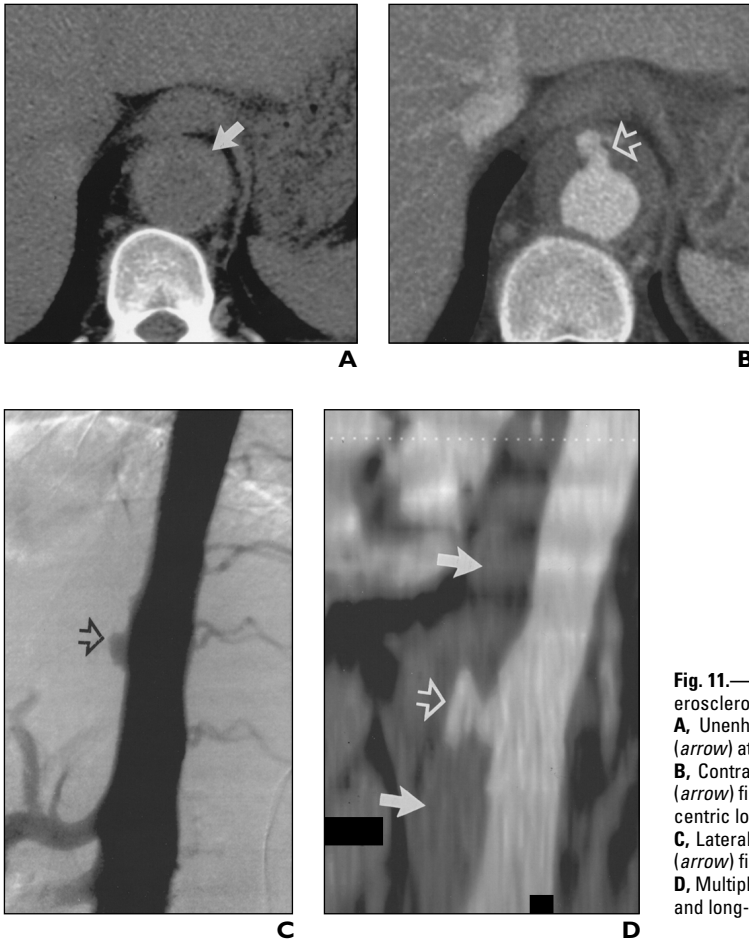


Fig. 11.—58-year-old woman presenting with severe back pain and penetrating atherosclerotic ulcer of aorta.
A, Unenhanced CT scan shows crescentic high-attenuation intramural hematoma (arrow) at distal thoracic aorta.
B, Contrast-enhanced CT scan obtained at level corresponding to **A** shows ulcer (arrow) filling with contrast material. Note that intramural hematoma presents as eccentric low-attenuation thickening of aortic wall.
C, Lateral angiogram of distal thoracic aorta shows anterior ulcerlike aortic lesion (arrow) filling with contrast material above level of celiac axis.
D, Multiplanar reformatted CT scan in sagittal view shows ulcer crater (open arrow) and long-segment intramural hematoma (solid arrows) in descending aorta.

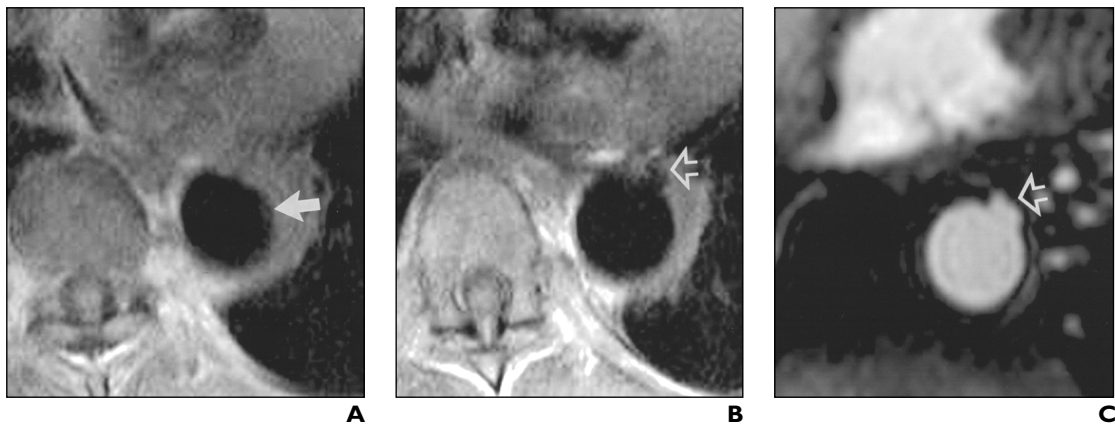


Fig. 12.—48-year-old man with penetrating atherosclerotic ulcer.
A, Axial double-inversion-recovery MR image (TR/TE, 1017/20; inversion time, 150 msec) shows intermediate-signal-intensity eccentric intramural hematoma in distal thoracic aorta (arrow).
B, Axial double-inversion-recovery MR image (1017/20; inversion time, 150 msec) shows distinct ulcer crater with signal void (arrow).
C, Contrast-enhanced spoiled gradient-refocused echo source MR image (3.7/1.3; flip angle, 30°) shows ulcer crater (arrow) filling with contrast material.
(Fig. 12 continues on next page)

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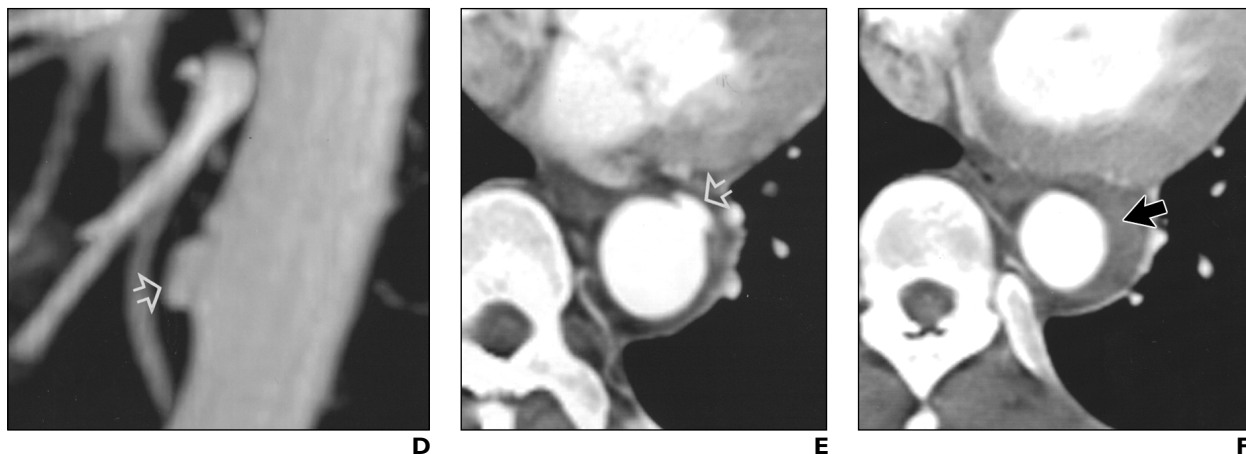


Fig. 12. (continued)—48-year-old man with penetrating atherosclerotic ulcer.
D, Multiplanar reformatted MR scan in oblique sagittal view shows ulcer crater (*arrow*).
E, Contrast-enhanced CT scan shows small focal contrast-filled outpouching (*arrow*) in distal thoracic aorta.
F, Axial CT scan obtained below ulcer crater level shows intramural hematoma (*arrow*), compatible with aggressive behavior of lesion.

Intramural hematoma can be distinguished from mural thrombus by identification of the intima: mural thrombus lies on top of the intima, which is frequently calcified, whereas intramural hematoma is subintimal. On unenhanced CT, intramural hematoma is hyperdense (Fig. 3). MR imaging can aid in the distinction of slow flow in the false lumen of a dissection from no flow in an intramural hematoma. Gradient-refocused echo pulse sequences, with the use of cine image acquisition, show cyclical flow-related enhancement in the false lumen of aortic dissection, whereas images of intramural hematoma show no signal intensity change. Dynamic phase-contrast MR imaging is more sensitive

than gradient-refocused echo sequences for excluding slow flow in the thickened aortic wall that would indicate aortic dissection rather than intramural hematoma [5].

Penetrating Atherosclerotic Aortic Ulcer

In a penetrating aortic ulcer, an atheromatous plaque ulcerates and disrupts the internal elastic lamina, burrowing deeply through the intima into the aortic media [2, 6]. When an atherosclerotic plaque penetrates into the media, the media is exposed to pulsatile arterial flow, which causes hemorrhage into the wall that then leads to intramural hematoma [7]

(Figs. 10–12). The plaque may precipitate a localized intramedial dissection associated with a variable amount of hematoma within the aortic wall, may break through into the adventitia to form a pseudoaneurysm, or may rupture. Ulceration of an aortic atheroma occurs in patients with advanced atherosclerosis. On imaging, a penetrating aortic ulcer can be distinguished from an atheromatous plaque by presence of a focal, contrast-filled outpouching surrounded by an intramural hematoma (Fig. 12), which confirms the aggressive behavior of the lesion. The atheromatous plaque with ulceration but without penetration through the intima shows irregular margins, but no contrast material extends beyond the level of intima,

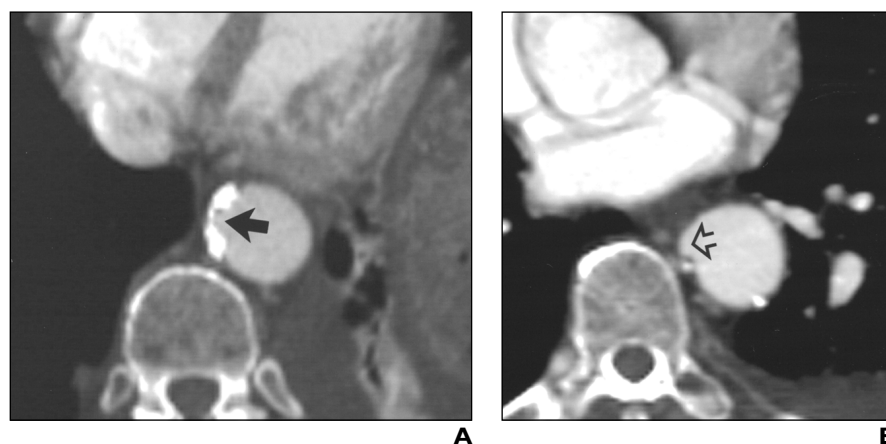


Fig. 13.—83-year-old man with chronic obstructive pulmonary disease and hypertension.
A, Contrast-enhanced CT scan shows calcified atheromatous plaque with focal ulceration (*arrow*) but without contrast extravasation beyond plaque.
B, Axial CT scan shows plaque-related intraluminal irregularity (*arrow*), but no contrast material is extending beyond level of intima (marked with calcification) and no intramural hematoma is present.

which is frequently calcified, and no intramural hematoma is present (Fig. 13).

Summary

Patients presenting with acute aortic syndromes usually have a similar clinical profile: aortic pain with coexisting history of hypertension. However, the pathophysiology and appearance of these syndromes differ in many ways. The classic aortic dissection involves an intimal flap, which traverses the aortic lumen. Intramural hematoma and penetrating aortic ulcer are nonflap lesions, with intramural hematoma showing no intimal disruption and penetrating aortic ulcer

showing an ulcer at the atherosclerotic plaque burrowing through the aortic intima and media. Radiologic evaluation plays a key role in assessing patients with acute disease of the aorta, and imaging techniques should aim both to diagnose the condition and to characterize the underlying pathology.

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