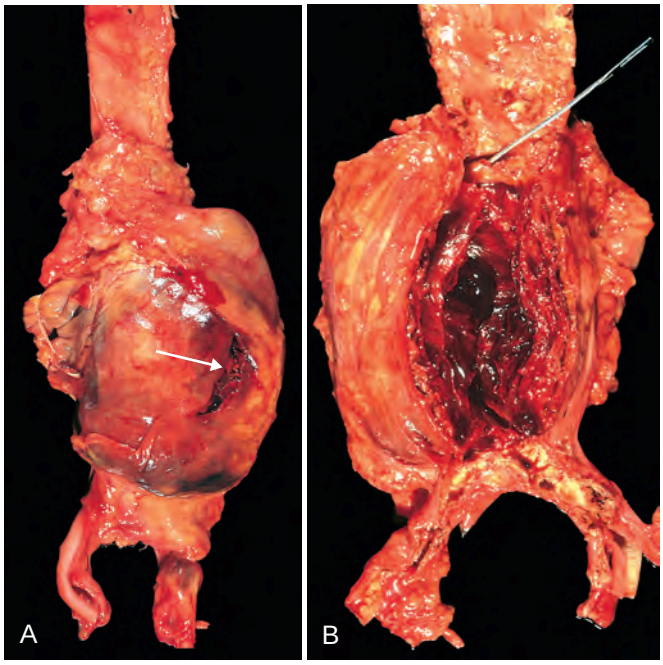

Figure 11-20 Abdominal aortic aneurysm. **A,** External view, gross photograph of a large aortic aneurysm that ruptured (rupture site is indicated by the *arrow*). **B,** Opened view, with the location of the rupture tract indicated by a probe. The wall of the aneurysm is exceedingly thin, and the lumen is filled by a large quantity of layered but largely unorganized thrombus.

AAAs occur more frequently in men and in smokers, rarely developing before age 50. Atherosclerosis is a major cause of AAA, but other factors clearly contribute since the incidence is less than 5% in men older than age 60 years, despite the almost universal presence of abdominal aortic atherosclerosis in this population.

MORPHOLOGY

Usually positioned below the renal arteries and above the bifurcation of the aorta, AAA can be saccular or fusiform, up to 15 cm in diameter, and up to 25 cm in length (Fig. 11-20). There is severe complicated atherosclerosis with destruction and thinning of the underlying aortic media; the aneurysm frequently contains a bland, laminated, poorly organized mural thrombus. AAA can occasionally affect the renal and superior or inferior mesenteric arteries, either by direct extension or by occluding vessel ostia with mural thrombi. Not infrequently, AAA is accompanied by smaller aneurysms of the iliac arteries.

Even though they are less common than the usual atherosclerotic aneurysm, three AAA variants merit special mention because of their unusual features:

- **Inflammatory AAA** account for 5% to 10% of all AAA; these typically occur in younger patients, who often present with back pain and elevated inflammatory markers (e.g., elevation of C-reactive protein). Inflammatory aneurysms are characterized by abundant lymphoplasmacytic inflammation with many macrophages (and even giant cells) associated with dense periaortic scarring that can extend into the anterior retroperitoneum. The cause is a presumed localized immune response to the abdominal aortic wall; remarkably, most cases are not associated with inflammation of other arteries.
- A subset of inflammatory AAA may be a vascular manifestation of a recently recognized entity called **immunoglobulin G4 (IgG4)-related disease**. This is a disorder marked by (in most cases) high plasma levels of IgG4 and tissue fibrosis associated with frequent infiltrating IgG4-expressing plasma cells. It may affect a variety of tissues, including pancreas, biliary system, and salivary gland. The affected individuals have aortitis and periaortitis that weaken the wall sufficiently in some cases to give rise to aneurysms. Recognition of this entity is important since it responds well to steroid therapy.
- **Mycotic AAA** are lesions that have become infected by the lodging of circulating microorganisms in the wall. In such cases, suppuration further destroys the media, potentiating rapid dilation and rupture.

Clinical Features. Most cases of AAA are asymptomatic and are discovered incidentally on physical exam as an abdominal mass (often palpably pulsating) that may mimic a tumor. The other clinical manifestations of AAA include:

- Rupture into the peritoneal cavity or retroperitoneal tissues with massive, potentially fatal hemorrhage
- Obstruction of a vessel branching off from the aorta, resulting in ischemic injury to the supplied tissue; for example, iliac (leg), renal (kidney), mesenteric (gastrointestinal tract), or vertebral arteries (spinal cord)
- Embolism from atheroma or mural thrombus
- Impingement on an adjacent structure, for example, compression of a ureter or erosion of vertebrae

The risk of rupture is directly related to the size of the aneurysm, varying from nil for AAA 4 cm or less in diameter, to 1% per year for AAA between 4 and 5 cm, 11% per year for AAA between 5 and 6 cm, and 25% per year for aneurysms larger than 6 cm. Most aneurysms expand at a rate of 0.2 to 0.3 cm/year, but 20% expand more rapidly. In general, aneurysms 5 cm or larger are managed aggressively, usually by surgical bypass with prosthetic grafts, although treatment via endoluminal approaches using stent grafts (expandable wire frames covered by a cloth sleeve) rather than surgery is now available for selected patients. Timely surgery is critical; operative mortality for unruptured aneurysms is approximately 5%, whereas emergency surgery after rupture carries a mortality rate of more than 50%. It is worth reiterating that because atherosclerosis is a systemic disease, a patient with AAA is also very likely to have atherosclerosis in other vascular beds and is at a significantly increased risk of IHD and stroke.

Thoracic Aortic Aneurysm

Thoracic aortic aneurysms are most commonly associated with hypertension, although other causes such as Marfan syndrome and Loeys-Dietz syndrome are increasingly recognized. These can present with signs and symptoms referable to (1) respiratory difficulties due to encroachment on the lungs and airways, (2) difficulty in swallowing due to compression of the esophagus, (3) persistent cough due to compression of the recurrent laryngeal nerves, (4) pain caused by erosion of bone (i.e., ribs and vertebral bodies), (5) cardiac disease as the aortic aneurysm leads to aortic valve dilation with valvular insufficiency or narrowing of