Inflammatory Aneurysm of the Thoracoabdominal Aorta With Associated Dissection

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The inflammatory variant of aortic aneurysms has 3 unique features: marked thickening of the aneurysm wall, fibrosis of the adjacent retroperitoneum, and rigid adherence of the adjacent structures to the anterior aneurysm wall. Abdominal tenderness with or without a pulsatile abdominal mass is the most common finding, although it is present in only about 33% of patients. Systemic symptoms, such as fever, malaise, and weight loss, are reported in about 20% to 50% of patients. A contrast-enhanced computed tomography scan, magnetic resonance imaging, and a transesophageal echocardiogram are among the best modalities to evaluate for inflammatory thoracoabdominal aneurysm, but a transthoracic echocardiogram can frequently be very suggestive. Medical treatment options include corticosteroids or other anti-inflammatory and immunosuppressive therapies. Surgical intervention usually consists of a transperitoneal approach with infrarenal aortic clamping. This case review describes a 64-year-old woman with a history of hypertension and dyslipidemia who presented with anemia, lower back pain, and a recent 30-pound weight loss.
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A 64-year-old woman with a history of hypertension and dyslipidemia presented to her primary medical doctor with complaints of lower back pain. She was currently a chronic smoker. After initial testing, magnetic resonance imaging (MRI) of the lumbar spine revealed radiculopathy. She was started on analgesics, including narcotics.

At a subsequent visit 2 months later, her hematocrit level had decreased from 38% to 20%, and she complained of a 30-pound weight loss over the past 3 months. She was referred for admission to the emergency department for

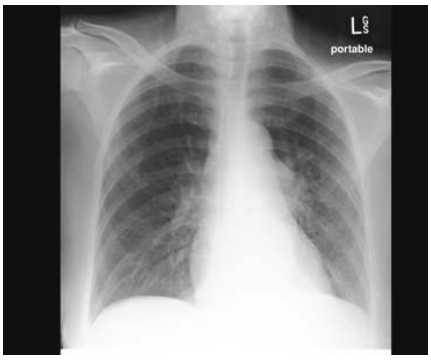

Figure 1. *The patient's chest x-ray was unremarkable, with a normal-sized mediastinum.*
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evaluation of the associated signs and symptoms. Her admission blood pressure was 152/80 mm Hg, without differential blood pressure in either arm. The chest x-ray was unremarkable, with a normal-sized mediastinum (Figure 1). The stool was positive for occult blood, and laboratory data were consistent with iron-deficiency anemia.

The patient was transfused with 2 units of packed red blood cells. The gastroenterology service was consulted to evaluate the patient for the decreased hematocrit level and to perform a colonoscopy in the setting of weight loss and anemia to exclude a gastrointestinal malignancy. A contrast-enhanced computed tomography (CT) scan of the abdomen and chest was ordered before the colonoscopy. The CT scan revealed an extremely large type B dissecting inflammatory aneurysm involving the distal descending aorta and upper abdominal aorta to the level below the renal pedicles, with multiple smaller aneurysms along the course (Figure 2). There was also associated periaortic thickening as well as irregularity of the aortic wall. The proximal end of the dissection was at the level of the carina, and the distal end was at the level of the left renal vein, with the lesion compressing the left main stem bronchus. The transthoracic echocardiogram (TTE) clearly showed that the descending thoracic aortic aneurysm was also compressing the posterior left ventricular wall and left atrium (Figure 3).

The patient was transferred to the coronary care unit and received a labetalol infusion for her elevated blood pressure. She was urgently referred for surgical intervention. Her erythrocyte sedimentation rate (ESR) was elevated at 55 mm/h, which also suggested an inflammatory process. A repeat CT scan revealed an acute mural thrombus in the proximal aneurysm with periaortic inflammatory changes associated with the descending aortic dissection.

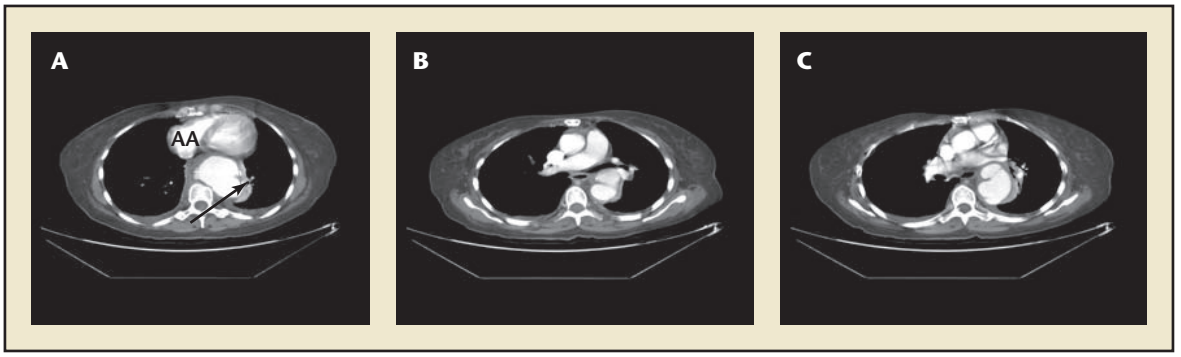
The patient underwent endovascular repair with a stent and aortic conduit placement. She continues to follow-up with her primary care physician with CT scans every 3 months, which show resolving inflammatory changes. The dimension of the aneurysm remains unchanged, and the patient is symptomatically improved.

Discussion

Aortic aneurysms have been recognized since the beginning of the 19th century. The first inflammatory aneurysms, however, were described in 1972 by Walker and colleagues (Table 1).[1] They reported that 5% to 10% of patients undergoing resection of abdominal aortic aneurysms (AAA) demonstrated 3 features unique to the inflammatory variant: marked thickening of the aneurysm wall, fibrosis of the adjacent retroperitoneum, and rigid adherence of the adjacent structures to the anterior aneurysm wall.

The development of noninflammatory aneurysm is primarily related to an alteration of elastic and fibrillar collagen in the media of the aortic wall.[2] The alteration is primarily dependent on the production of proteases by the medial smooth cells and adventitious fibroblasts.[2,3] These proteases are primarily responsible for thinning of the aortic media, which subsequently leads to aneurysm formation in the noninflammatory variant.[2,4] Conversely,

Figure 2. *Contrast-enhanced computed tomography scan. Multiple aneurysms with periaortic fibrosis and inflammation are evident (***A***). The type B dissection involves the descending aorta with irregular walls and periaortic thickening (***B*** and* **C***). AA, ascending aorta.* www.medreviews.com



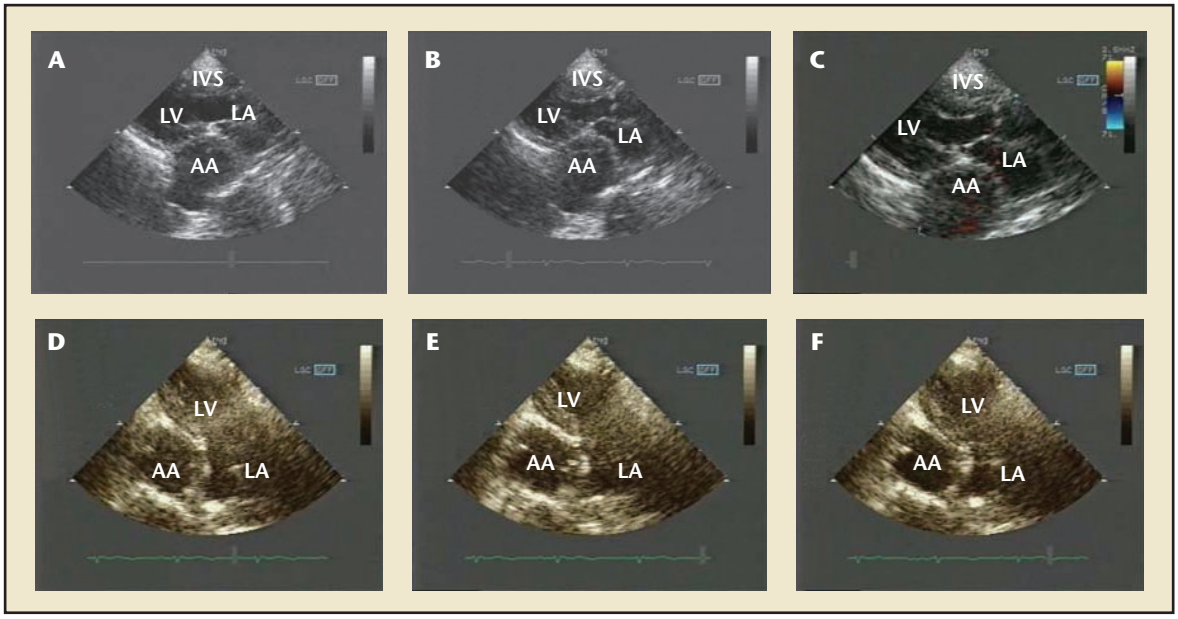


Figure 3. *Transthoracic echocardiogram. Parasternal views show the aneurysm compressing the LA and LV (***A-C***). Apical 2-chamber views show the thoracic aneurysm (***D-F***). IVS, interventricular septum; LV, left ventricle; LA, left atrium; AA, ascending aorta.*
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the exact pathophysiology for inflammatory AAA remains unclear. The histopathology suggests an immune response to antigen localized to adventitia, which has been hypothesized to be an oxidized low-density lipoprotein and ceroid.[4] The antigen's ability to promote an inflammatory response may depend upon thinning or disruption media caused by proteases, which has been thought to be dysfunctional in the inflammatory AAA.[2,3,5] The possibility that extensive inflammation is related to inflammatory AAAs more so than to the noninflammatory variant has not reached consensus because retroperitoneal fibrosis has been shown in certain cases to regress after surgery.[5]

The majority of inflammatory aneurysms present as iliac aneurysms. Only about 1% of all inflammatory aortic aneurysms present in the ascending aorta, and their presence in the descending aorta is even more rare.[6,7] An associated dissection occurring concomitantly with an inflammatory aneurysm of the descending thoracic and abdominal aorta in continuity is an extremely rare presentation. In our literature search, we came across only 2 cases with inflammatory aneurysm of the descending aorta and none with associated dissection.[8,9] An associated dissection with inflammatory aortic aneurysm is a life-threatening condition, and early identification remains important for management.[10] When left untreated, approximately 30% to 35% of patients die within the first 24 hours, and about 50% die within 48 hours.[10,11]

Inflammatory aortic aneurysm is more common in men than in women.[1,3,6,7] Smoking has universally been associated with inflammatory abdominal aortic aneurysm.[3,6,7] Hypertension and dyslipidemia, which are risk factors for atherosclerotic

Table 1
Differences in Inflammatory Versus Atherosclerotic Aneurysms

	Epidemiology	Signs and Symptoms	Imaging Studies
Inflammatory Aneurysm	Family history More common in men than in women Younger age	Weight loss (+) Systemic findings (+) ESR (↑), CRP(↑)	Periaortic inflammation (+) Fibrosis (+)
Atherosclerotic Aneurysm	HTN Dyslipidemia, as with inflammatory variant	Weight loss (−) Systemic findings (−) ESR (−), CRP (−)	Periaortic inflammation (−) Fibrosis (−)

ESR, erythrocyte sedimentation rate; CRP, C-reactive protein; HTN, hypertension.

AAA, have also been associated with the inflammatory variant. However, the more common risk factors of the inflammatory variant are younger age of presentation and positive family history.[3]

In contrast to atherosclerotic abdominal aneurysms, patients with inflammatory AAA are more likely to have symptoms and signs at presentation.[1,3] Abdominal tenderness with or without a pulsatile abdominal mass is the most common finding but is present in only about 33% of patients.[12] The presentation may also be of lower back or abdominal pain, mainly in the lumbar area. Patients with associated dissection may present with pain described as tearing and ripping in quality. Systemic symptoms, such as fever, malaise, and weight loss, are reported in approximately 20% to 50% of patients with inflammatory aneurysms.[2-4] Inflammatory AAAs are accompanied by extensive retroperitoneal fibrosis, and there may be symptoms and signs of duodenal obstruction, ureteral colic, or inferior vena caval obstruction.[4] The lifetime risk of rupture posed by the inflammatory variant is less than 5%.[3] However, altered mental status with neurological symptoms, syncope, orthopnea, dyspnea, and dysphagia may occur with associated dissection and subsequent rupture.[10,11] Laboratory abnormalities reflecting systemic inflammation are common and include elevated ESR and C-reactive protein. In fact, in initial studies, almost all patients with inflammatory AAA had elevated ESR, compared with only 29% of patients with the atherosclerotic variant.[1] In subsequent studies, the percentages of patients with an elevated ESR have ranged from 40% to 90%.[2,4] The elevation is often modest, with values in the 20 to 50 mm/h range.[2,8,9] Anemia is less common, and leukocytosis is rare.[4] Some studies have reported a high frequency of antinuclear antibodies and antineutrophil cytoplasmic antibodies.[12,13] One study of 31 patients with inflammatory AAA also found that 19% had an associated autoimmune disease.[5]

The diagnosis of inflammatory aneurysm is first made primarily with imaging studies macroscopically, and then it is confirmed microscopically with tissue diagnosis. The CT scan plays an important role in delineating the aneurysm and the thickened aortic wall, as well as the surrounding periaortic inflammation and fibrosis.[14] The periaortic inflammation is isodense to muscle but enhances with administration of intravenous contrast material.[3,14] Dynamic scanning reveals rapid intraluminal enhancement, slightly delayed enhancement of the inflammatory mass, and nonenhancement of the thick fibrous adventitia.[3,14] MRI is more sensitive and specific than a CT scan in demonstrating the aneurysm, the thickening of the wall, and the surrounding periaortic inflammation.[15] MRI also avoids the radiation exposure and administration of the nephrotoxic intravenous contrast material required with CT.[2,14,16] The periaortic cuff of inflammation also enhances strikingly with administration of intravenous gadolinium.[16] However, because the MRI provides less information about the ureters and is less readily available, some physicians prefer CT imaging and reserve MRI for patients who have renal insufficiency.[14,16]

Systemic symptoms, such as fever, malaise, and weight loss, are reported in approximately 20% to 50% of patients with inflammatory aneurysms.

Transesophageal echocardiography (TEE) is as reliable as CT or MRI for confirming or ruling out thoracic aortic dissection and aneurysms in patients at risk of aortic pathology.[17,18] It is a simple and accurate method for screening of aortic aneurysms and considered more sensitive and specific than the TTE. Any echocardiographic method can be performed at the bedside, thus eliminating the need to move critically ill patients. A disadvantage of TEE is that it cannot be performed in patients with esophageal pathology, and it increases the risk of aspiration pneumonia in people who are not fasting or who are critically ill. TTE, on the other hand, which is often considered insufficient for diagnosis of descending aortic pathology, can be as diagnostic and accurate as TEE when 3-dimensional transthoracic images are obtained. Great 2-dimensional images can also be suggestive and even conclusive in almost 50% of cases.[19] Three-dimensional transthoracic images can clearly differentiate true aortic aneurysm and dissection flaps from artifacts, which had been a limitation of 2-dimensional transthoracic images.[20] TTE can also be safely performed on all critically ill patients, even those who need to be monitored closely.[17,18,20] In fact, a live 3-dimensional TTE alleviates the need for esophageal probing and can give rapid information on the aortic pathology without any of the artifacts that are commonly produced by the patient's motion and the transducer.[19] Our patient's 2-dimensional TTE provided sufficient information that, along with the clinical presentation, clinched the diagnosis.

Initial therapy should begin when the diagnosis is suspected. The possibility that inflammatory AAA results primarily from inflammation of the periaortic tissues has led many

physicians to use corticosteroids or other anti-inflammatory and immunosuppressive therapies.[3,8] Anti-inflammatory therapy has been associated with improvement of symptoms and signs and with CT or MRI evidence of resolving inflammation.[2,4] Other agents such as methotrexate, cyclophosphamide, and azathioprine have also been reported to be effective.[3,5] However, the ultimate efficacy of the anti-inflammatory approach has not yet been proven.

The aim of surgical treatment is to prevent rupture, which is often catastrophic.[10,11,15] Although data suggest that an inflammatory AAA is less liable to rupture than an atherosclerotic AAA, surgical intervention is recommended once the diameter exceeds 5.5 cm and when an associated dissection occurs, as was the case in our patient.[15,21,22]

The surgical management of inflammatory aneurysm presents a unique challenge for the surgical team. An inflammatory AAA forms dense adhesions that surround the aneurysm and frequently involve the duodenum (97% to 100%), inferior vena cava (63% to 70%), and ureters (20% to 44%).[4,15] Early surgical experience showed that lyses of adhesions and mobilization of periaortic structures frequently caused complications, including enterotomies, especially of the duodenum; injuries to the ureters and vena cava; and increased mortality.[22,23] Most surgeons now favor a transperitoneal approach to inflammatory AAA, and infrarenal aortic clamping is usually performed.[23,24] The current guideline to surgical management of inflammatory thoracic aneurysm is insufficient and is based primarily on the surgeon's experience. Postoperative mortality for inflammatory aortic aneurysm remains higher than for the atherosclerotic variant.[2] Minimizing dissection of these tissues has greatly reduced the likelihood of complications.[10] The definitive treatment for associated type B dissections, as in our patient, is surgical. Surgical intervention is also recommended if there is a leak, rupture, compromised blood flow to a vital organ, or failure of medical management with hemodynamic instability.[10,11]

There are no clear guidelines for follow-up in inflammatory AAA. The usual practice consists of follow-up examinations with radiological studies at 3-month intervals for the first year and 6-month intervals for the next 2 years. A similar approach is usually followed after surgical repair of aortic dissection.

Conclusion

An inflammatory aortic aneurysm should be suspected in patients presenting with associated history of hypertension, smoking, and familial history of aneurysm. The symptoms commonly are abdominal or back pain, associated weight loss, and systemic finding of inflammation, such as fever, elevated ESR, and elevated C-reactive protein.[1,3,6-8] Of note, the presentation of weight loss, as in our patient, is present in 20% to 50% of patients with inflammatory aneurysms. This symptom may be related to chronic blood loss and underlying systemic inflammation. Dissection should be suspected in patients presenting with classic tearing chest pain and hemodynamic instability.[10,11] Initial medical therapy with corticosteroids has been shown to be useful in some studies. When an inflammatory aneurysm is larger than 5.5 cm (as in the atherosclerotic variant) or is associated with dissection or hemodynamic instability, the patient should be referred for emergent surgery and managed in an intensive care setting.[10,15,21-23] Despite stringent management, 1% to 2% of patients with underlying aortic dissection die per hour during the first 24 to 48 hours[11]; therefore, this diagnosis remains a very important and

Main Points

- Aortic aneurysms have been recognized since the beginning of the 19th century. Inflammatory aneurysms were first described in 1972.
- Abdominal tenderness with or without a pulsatile abdominal mass is the most common finding, although it is present in only about 33% of patients.
- Transesophageal echocardiography is as reliable as computed tomography or magnetic resonance imaging for confirming or ruling out thoracic aortic dissection and aneurysms in patients at risk of aortic pathology. Transthoracic echocardiography with 3-dimensional imaging can, however, be almost as suggestive.
- The possibility that inflammatory abdominal aortic aneurysms result primarily from inflammation of the periaortic tissues has led many physicians to use corticosteroids or other anti-inflammatory and immunosuppressive therapies.
- Most surgeons favor a transperitoneal approach, and infrarenal aortic clamping is usually performed.

critical one to make. Early diagnosis and treatment could be life-saving for this otherwise fatal disease.

A contrast-enhanced CT scan and TEE are some of the best modalities to evaluate for inflammatory thoracoabdominal aneurysms, as they not only assess for the aneurysm but also detect underlying dissection.[14,16] These tests are more readily available than MRI and angiography.[14,16] Although a CT scan, TEE, and MRI are considered the gold standards for the diagnosis of this disease, one should not undervalue the contributions of TTE in diagnosing aortic pathology. TTE images can provide rapid information and alleviate the need for esophageal intubation and contrast administration as needed for TEE and CT, respectively.[19,20] The future will hold much more promise for 3-dimensional TTE, which has the potential to become a new standard. ■

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