A 69-year-old woman was transferred to our intensive care unit with concerns of a possible rupture of abdominal aortic aneurysm (AAA). She presented with a 1-week history of sharp left upper quadrant abdominal pain radiating to her back. Her comorbidities included hypertension and chronic obstructive pulmonary disease with a >90 pack-year history of smoking. On examination, she was hemodynamically stable but was cachectic in appearance and had epigastric tenderness. Laboratory evaluation revealed hemoglobin of 11g/dL, white cell count of 12900 k/uL, and creatinine of 0.4 mg/dL. Both the erythrocyte sedimentation rate and c-reactive protein were elevated (100 mm/h and 6.9 mg/dL, respectively). Computed tomography scan performed at the outside facility demonstrated diffuse aortic plaque with descending thoracic aortic aneurysm measuring 4.2 cm (Figure 1, arrow) and a small juxtarenal abdominal aortic aneurysm measuring 3.9 cm with adjacent soft tissue mass along the left lateral aspect of the aorta (Figure 2). The celiac artery was occluded. Additional diagnostic testing included antinuclear and anti-neutrophilic cytoplasmic antibody titers and blood cultures, all of which were normal. Subsequent magnetic resonance angiography with gadolinium and edema-weighted imaging demonstrated enhancement and edema of the periaortic soft tissue mass extending into the left retroperitoneal space and around the inferior vena cava (Figure 3).

Given the differential diagnosis of inflammation versus neoplasm, a computed tomography-guided biopsy of the periaortic mass was performed and demonstrated chronic periaortic inflammation with infiltrate of neutrophils, lymphocytes, plasma cells, and eosinophils (Figure 4A), as well as reactive fibroblast proliferation (Figure 4B). Of note, the characteristic findings of giant cell arteritis were absent. There was no evidence of malignancy. Serum immunoglobulin G subclass panel did not show an excess of immunoglobulin G4. With the diagnosis of inflammatory AAA, the patient was started on prednisone 30 mg/d with subsequent slow taper and plan for surveillance imaging. Antiatherosclerotic therapies, including statin, aspirin, and angiotensin-converting enzyme inhibitor were instituted. The patient was counseled on the importance of smoking cessation.

Patient responded well to steroids initially and her flank/back pain resolved. The patient was discharged with plans for clinical follow-up at 1 month followed by an MRI of the aorta at 3 months to evaluate the response of periaortic inflammation and surrounding soft tissues. She was also counseled on need for periodic imaging surveillance to monitor the growth of her thoracic and abdominal aortic aneurysms. She opted to follow-up in her local community and was lost to follow-up at our facility.

Inflammatory AAA is on the spectrum of disorders known as chronic periaortitis, also including idiopathic thoracic and abdominal periaortitis and idiopathic retroperitoneal fibrosis (Ormond’s disease).1 This process can involve the inferior vena cava resulting in lower extremity swelling. Involvement of ureters may present as hydronephrosis.2 In this entity, inflammation of the aortic adventitia and surrounding structures generally does not involve the aortic vessel wall to the extent of Takayasu or giant cell arteritis. Recently, the association of chronic periaortitis and immunoglobulin G4-related inflammatory disease has been described.3 Inflammatory AAA is associated with a higher prevalence of aneurysm-related symptoms, such as back or abdominal pain and fever, and a lower risk of rupture than conventional AAA.1

Disclosures
Dr Clair is on the Advisory Board for Boston Scientific and Medtronic, and is a consultant for Cordis, Covidien, Endologix, vV3, and WL Gore. The other authors report no conflicts.

References

From the Departments of Vascular Surgery (J.F., D.G.C.), Rheumatology (C.G.), Anatomic Pathology (S.D.B.), and Cardiovascular Medicine (H.L.G.), Cleveland Clinic, Cleveland, OH.

Correspondence to Heather Gornik, MD, Department of Cardiovascular Medicine, Cleveland Clinic, 9500 Euclid Avenue – Desk F30, Cleveland, OH 44195. E-mail gornikh@ccf.org

(Circulation. 2014;130:1300-1302.)
© 2014 American Heart Association, Inc.

Circulation is available at http://circ.ahajournals.org

DOI: 10.1161/CIRCULATIONAHA.114.010173
Figure 1. Computed tomography (CT) scan demonstrating diffuse aortic plaque (arrow) with descending thoracic aortic aneurysm.

Figure 2. Juxtarenal abdominal aortic aneurysm with adjacent soft tissue mass along the left lateral aspect of the aorta (arrowhead).
Figure 3. MRI demonstrates edema within the enhancing peri-aortic soft tissue (A, arrow), within the retroperitoneum surrounding the aorta and IVC and extending into the left retroperitoneal space (arrow) and inferiorly to below the aortic bifurcation.

Figure 4. Histopathology slides showing infiltrate of neutrophils, lymphocytes, plasma cells and eosinophils (D1), as well as reactive fibroblastic proliferation (D2).
Inflamed Abdominal Aortic Aneurysm With Retroperitoneal Fibrosis
Javairiah Fatima, Carmen Gota, Daniel G. Clair, Steven D. Billings and Heather L. Gornik

*Circulation.* 2014;130:1300-1302
doi: 10.1161/CIRCULATIONAHA.114.010173
*Circulation* is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2014 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/130/15/1300

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in *Circulation* can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to *Circulation* is online at:
http://circ.ahajournals.org//subscriptions/