

# Abdominal Aortic Aneurysm in a Patient with Systemic Lupus Erythematosus

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## CASE PRESENTATION

### Initial Presentation and History

A 61-year-old African American woman presented to the hospital with bilateral lower extremity edema that had worsened over one week and associated shortness of breath on exertion. For several weeks, she awakened at night with dyspnea that resolved spontaneously after sitting up in bed. On the morning of presentation, she reported an episode of chest pain with worsening shortness of breath; the pain was midsternal and radiated down the left arm without other associated symptoms or aggravating factors and resolved within 2 hours.

The patient's past medical history included hypertension, congestive heart failure, severe mitral regurgitation, gastritis, chronic systemic lupus erythematosus (SLE) (diagnosed in the 1970s), and a 4 × 4-cm asymptomatic abdominal aortic aneurysm (AAA) found incidentally on an abdominal computed tomography (CT) scan performed 3 years ago. Her past surgical history included bilateral hip arthroplasty and a total abdominal hysterectomy for uterine fibroids. She reported a 20 pack-year smoking history and occasional alcohol use but denied illicit drug use. There was no family history of AAA or SLE.

Outpatient medications included cyclophosphamide 50 mg once daily, digoxin 0.125 mg once daily, furosemide 20 mg twice daily, potassium chloride 20 mEq once daily, diltiazem SR 240 mg once daily, isosorbide mononitrate once daily, quinapril 20 mg once daily, ranitidine 150 mg once daily, and 1 to 2 tablets of acetaminophen/codeine 4 times daily for arthralgia.

### Physical Examination and Laboratory Studies

On physical examination, the patient was an obese woman in no apparent distress. Vital signs were stable, including a blood pressure of 120/72 mm Hg, pulse rate of 94 bpm, respiratory rate of 16 breaths/min, temperature of 99.4°F (37.4°C), and pulse oximetry reading of 100% on 2 L/min of oxygen via nasal canu-

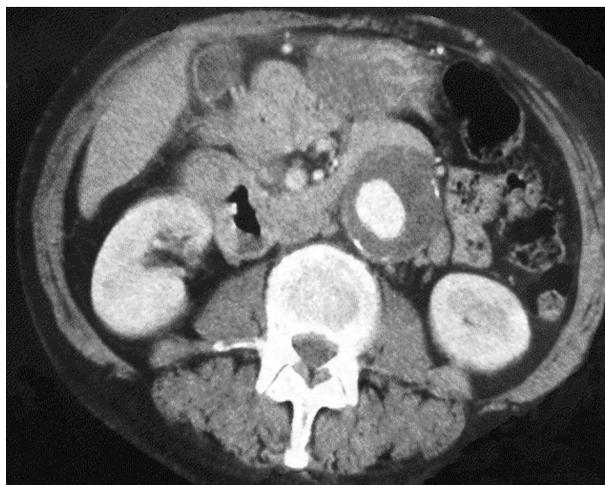
la. Neck examination showed no jugular venous distension and no audible carotid bruits. Cardiovascular examination demonstrated a regular rhythm with a grade IV/VI systolic murmur, and pulmonary examination revealed decreased breath sounds at the bases bilaterally. Abdominal examination yielded bowel sounds in all quadrants, and the abdomen was soft, nontender, and nondistended, with no palpable mass. Evaluation of the extremities demonstrated 1+ pitting edema bilaterally without cyanosis or calf tenderness, and Homans's sign was not elicited. Results of laboratory studies revealed a hemoglobin level of 9.0 g/dL (normal, 11.5–15.2 g/dL), normal levels of serum electrolytes, and an erythrocyte sedimentation rate of 110 mm/h (potentially indicating an inflammatory process such as an acute exacerbation of SLE). Initial measurement of troponin I was less than 1 ng/mL. The electrocardiogram (ECG) revealed nonspecific ST/T wave changes with a ventricular rate of 83 bpm. Chest radiography demonstrated clear lung fields but noted cardiomegaly with aortic calcification.

### Clinical Course

The patient was admitted with a diagnosis of an acute exacerbation of congestive heart failure and was evaluated for an acute myocardial infarction. During her hospital stay, cardiac enzymes and serial troponin I measurements remained within normal limits, and serial ECGs remained at their baseline reading. The patient's anemia was previously investigated and later confirmed as anemia of chronic disease. She was given

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**Figure.** An abdominal computed tomography scan of the patient demonstrating an abdominal aortic aneurysm extending to the iliac bifurcation.

a diuretic, and her daily fluid intake and output were closely monitored. She was also maintained on all outpatient medications. A Persantine stress test did not demonstrate any perfusion/reperfusion defects.

On hospital days 1 and 2, the patient remained pain-free, and her shortness of breath and leg edema improved. However, on hospital day 3, she began to complain of back pain, which she described as radiating to the abdomen. She denied any tearing sensation associated with the pain and had no nausea or vomiting. Her abdominal examination was benign, and her vital signs were stable. In view of her history of AAA, an abdominal CT scan with contrast was obtained. When compared with the previous CT scan, the aneurysm was noted to have increased from  $4 \times 4$  cm to  $4.5 \times 4.5$  cm, with a large intraluminal thrombus (patent intraluminal diameter, 2.5 cm). The aneurysm was noted to extend to the iliac bifurcation (**Figure**).

Vascular surgery was consulted, and based on the aneurysm's rapid increase in size, the surgeon recommended surgical repair, which could include either open repair or endovascular stent-graft placement (EVS GP). After further evaluation, it was decided that emergent surgical repair was not indicated during hospitalization but needed to be arranged as an elective procedure soon after the patient was discharged. The next day (hospital day 4), the patient's back pain resolved after taking acetaminophen and rest, and as a result, the pain was believed to be musculoskeletal and not related to the AAA. Again, the abdominal CT findings were incidental, but they did reveal the asymptomatic yet rapid increase in the size of the AAA.

The patient was discharged on hospital day 5 in stable condition with instructions to follow-up with the vascular surgeon to schedule the recommended procedure as well as any necessary imaging for EVS GP. She was advised to maintain a low-salt diet and continue all previous medications as well as to see her primary care physician within 7 days.

#### **Follow-up of the Patient**

The patient remained asymptomatic and did not see her primary care physician until 5 months after discharge. She explained that she did not want to undergo surgery at that time. Another CT scan of the abdomen was performed and demonstrated a further increase of the AAA to  $4.5 \times 5.0$  cm, without any dissection. The patient was again advised to follow-up with the vascular surgeon for repair of the AAA.

#### **DISCUSSION**

There are few reports in the literature that describe an association between AAA and SLE and the resulting worse prognoses when these conditions occur in concert. In fact, there are only 16 reported cases of AAA occurring in combination with SLE, but this number may be increasing.<sup>1-16</sup> AAA is a rare, life-threatening complication in SLE patients.<sup>17,18</sup> Although only a few documented cases suggest that SLE is a primary cause for AAA formation, the inflammatory processes related to SLE, in combination with other causative factors (eg, hypertension, atherosclerosis, tuberculosis), may contribute to the rapid expansion and possible rupture of the AAA.

SLE is an autoimmune disease that affects multiple organ systems and is characterized by immune complex deposition, complement activation, and production of autoantibodies. While the exact etiology of SLE remains unknown, multiple autoantibodies are thought to be created against an array of nuclear and cytoplasmic components of the cell and are neither organ- nor species-specific, ultimately resulting in renal, pulmonary, cardiovascular, and other multisystem disorders. Several cardiovascular disorders (eg, pericarditis, endocarditis, vasculitis) can affect SLE patients. Valvular involvement, pericarditis, myocarditis, and early coronary artery disease are also frequently found in SLE patients. New vascular complications have been observed as a result of prolonged patient survival and improved noninvasive surveillance techniques.

Autoimmune vascular injury may initiate the inflammatory process, leading to atherosclerotic plaque formation via numerous mechanisms: (1) immune complexes deposited on the vascular wall may stimulate cholesterol accumulation; (2) the formation of autoantibodies to

oxidized low-density lipoproteins may accentuate these atherogenic particles within vessel wall macrophages; and (3) the formation of autoantibodies to lipoprotein lipase may lead to elevated triglyceride levels. Other more general effects of an impaired inflammatory process include vascular endothelial dysfunction, platelet hyperactivity, and impaired fibrinolysis, all of which are associated with atherosclerotic plaque formation.

The development of the AAA in the case patient was likely multifactorial. In addition to SLE, the patient had long-standing hypertension, as well as a history of cigarette smoking (a known risk for coronary and peripheral vascular disease development). However, if the SLE was not the primary cause of the AAA, the severe systemic inflammation associated with SLE may have contributed to the rapid progression of the AAA.

The presence of SLE influences the significance and prognosis of aortic aneurysms. SLE is an additional risk factor for the dissection and rupture of an existing aneurysm. In reported cases of dissected and ruptured AAA in SLE patients, the aneurysms were between 3 and 6 cm in size. Earlier surgical correction of AAA in SLE patients, as compared with patients without SLE, may be recommended, as several successful surgeries have been reported in the literature.<sup>15,16,19</sup> Surgical options include conventional open repair with a Dacron graft or EVSGP, which is a newer, less invasive procedure and has shown promise for treating AAA in SLE patients. However, the use of EVSGP is very limited because of the low prevalence of confirmed pathology relating AAA and SLE and because most aneurysms are discovered as incidental findings on abdominal ultrasound or CT scan or during a routine physical examination in asymptomatic patients.<sup>19</sup> Because surgical management of active vascular disease often requires a more aggressive approach to achieve secure anastomosis, EVSGP may offer a less invasive solution for patients with acute aortic pathology in underlying inflammatory conditions, such as SLE.

## CONCLUSION

Physicians should be aware that AAA can occur in SLE patients and be mindful that AAA may rapidly progress in this setting. Physicians should also have a low threshold for diagnostic testing, including abdominal CT scanning for back or abdominal pain or transesophageal echocardiograms for chest pain, in these patients<sup>15</sup> as aneurysms can quickly progress and potentially rupture early in the course of disease. Improved blood pressure control, use of steroid-sparing agents, and appropriate use of all imaging modalities are essential to prevent potentially fatal vascular dam-

age in the presence of SLE.<sup>20</sup> Family physicians, who follow these patients on a long-term basis, must bear in mind that recurrences have been reported after AAA repair or stent placement.<sup>21</sup> Aggressive monitoring of the AAA is critical for the successful long-term management of these patients.

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