Aortic dissection and accelerated aneurysmal degeneration in a patient with giant cell arteritis

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ABSTRACT

Giant cell arteritis (GCA) is associated with nonatheromatous aortic pathology. Here we present a case in which a 76-year-old woman with a biopsy-proven history of GCA and a previous repair of her ascending aortic aneurysm presents with an acute dissection of a 4-cm aneurysm in the descending thoracic aorta. It was treated using endovascular techniques. This report adds to a growing body of evidence that GCA is a risk factor for aortic dissection and nonatheromatous aortic aneurysms. (J Vasc Surg Cases and Innovative Techniques 2020;6:598-602.)

Keywords: Giant cell arteritis; Aortic dissection; Aortic aneurysm; Aortitis

Aortitis describes an inflammatory pathology of the aortic wall. These are classified as either rheumatologic, infectious disease related, or isolated aortitis.[1] Within the rheumatologic causes of aortitis, giant cell arteritis (GCA) and Takayasu arteritis are most common.[1] However, GCA is generally uncommon, usually affecting those of Northern European descent at a rate of 20 per 100,000 individuals beyond 50 years of age with the highest incidence between the seventh and eight decades of life.[2]

Here we present a case in which a 76-year-old woman with a biopsy-proven history of GCA experienced dissection of a 4 cm descending thoracic aortic aneurysm (TAA). Written consent was obtain from the patient for publication.

PATIENT HISTORY

The patient's past medical history was positive for hypertension, dyslipidemia, and osteopenia. From a past surgical perspective, she previously had a cholecystectomy, hysterectomy, and an ascending aortic aneurysm measuring 6 cm, which was repaired in 2010. In November 2010, she was diagnosed with GCA that was biopsy proven at another center. Unfortunately, after relocating to another city, she was lost to follow-up with her local rheumatologist. She did not have a family history of vascular pathology. She is a former smoker with a 50-pack-year smoking history.

Her home medication included metoprolol, aspirin, calcium, vitamin D, methylphenidate (Ritalin), and hydrochlorothiazide. No immune-modulating or anti-inflammatory therapy was noted on her medication regimen.

CASE REPORT

The patient presented to the emergency department with acute onset of severe epigastric pain and nausea. Chest and abdominal radiographs revealed marked unfolding of the thoracic aorta and widening of the mediastinum. A subsequent computed tomography (CT) angiographic scan discovered a type $B_{3,5}$ dissection[3] in association with a descending TAA, measuring a maximum of 6.0 $\times$ 6.8 cm, and a thickened aortic wall. A penetrating ulcer with and an intramural hematoma and was seen along the proximal descending thoracic aorta with a second focal dissection inferior to the ulcer (Fig 1).

The patient was admitted to the intensive care unit and continuous intravenous labetalol was initiated to maintain systolic blood pressures of less than 120 mm Hg. Rheumatology was consulted and methylprednisolone 50 mg/d intravenously was initiated for 7 days to decrease the inflammatory process and transitioned to prednisone 60 mg/d. Serial CT scans were completed to follow the progression of the dissection. Within 11 days, the dissection progressed caudally into the diaphragmatic hiatus to a length of 11.8 cm and an aneurysmal expansion to 6.3 cm $\times$ 6.9 cm.

Blood work. Blood work was obtained in the emergency department. Rheumatologic blood work was within normal limits and C-reactive protein was elevated at 125.4 mg/L.

Intervention. During the admission, the patient developed sudden back pain described as 9 out of 10 in severity. A CT angiography scan showed a rapidly expanding descending TAA. Given the dynamic symptoms and changes noted on imaging, she was consented for an endovascular repair of her TAA and dissection. A Zenith Alpha Thoracic Endovascular Graft by

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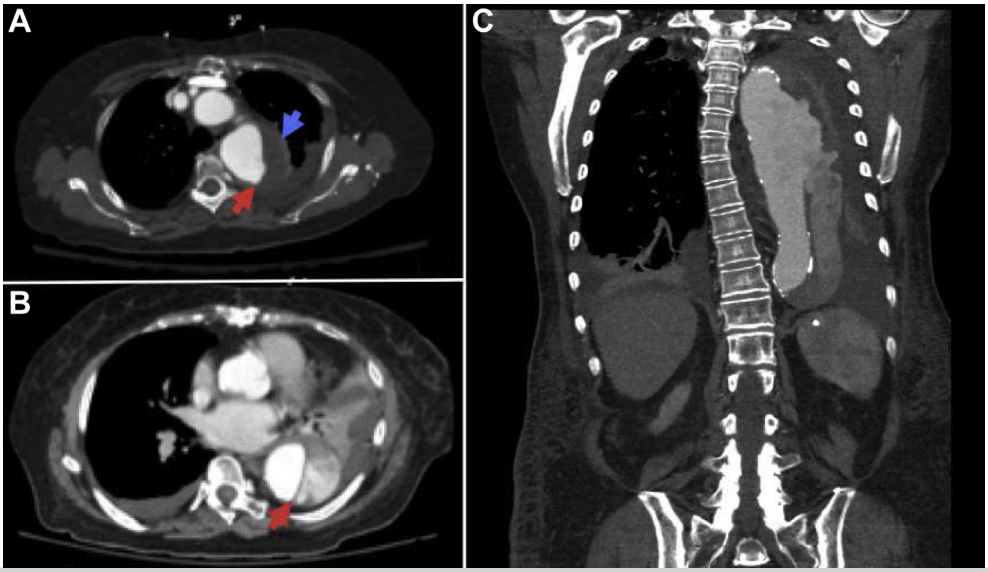

Fig 1. Type B dissection with interval expansion of the false lumen and increased aortic diameter of 6.3 × 6.9 cm. **A,** Axial view with evidence of aortic ulcer (*red arrow*) and mural hematoma (*blue arrow*) at the proximal aorta. **B,** Axial view with dissection flap seen within the aorta dividing native and false lumens and thickened aortic wall (*red arrow*). **(C)** Coronal view of the aorta showing extensiveness of the false lumen and aneurysmal degeneration.

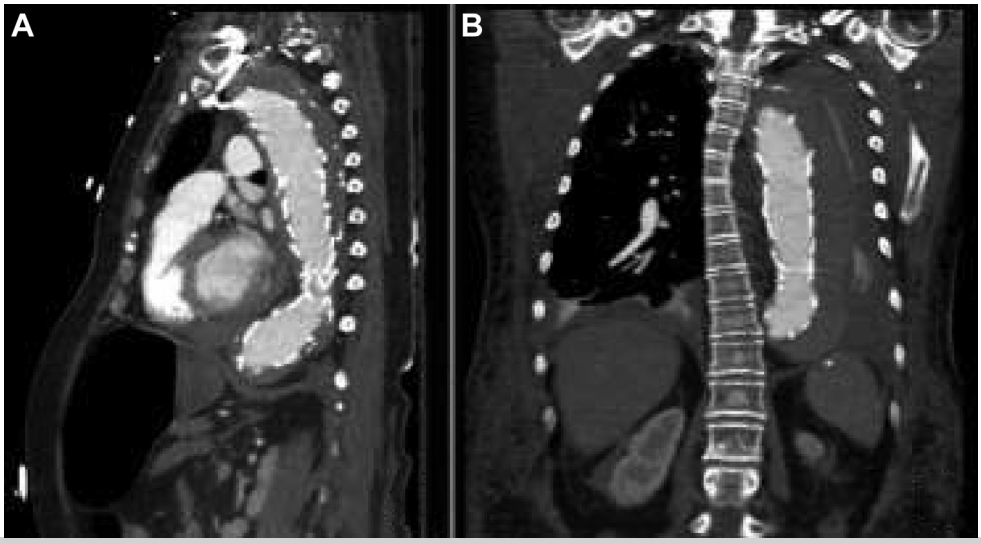

Fig 2. Endovascular treatment of thoracic aortic penetrating ulcer, dissection, and aneurysm. **A,** Sagittal view of aortic stent placement. **B,** Coronal view of patent graft.

Cook Medical (ZTA-P-34-209; Bloomington, Ind) device was initially deployed in the distal aspect of the aorta, because there was no reentry tear, landing at the level of the diaphragmatic hiatus with distal fixation at the level of the celiac axis. A second Cook Medical graft (TZA-P-38-217) was deployed proximally thereby covering zone 2 of the aorta, and downstream the proximal end of the distal graft was overlapped. Subsequent angiograms showed that the aneurysm was completely excluded (Fig 2 and Fig 3).

Pathology. A total length of 8.5 cm of ascending aorta was previously submitted to pathology after repair of the patient's ascending aortic aneurysm in 2010. Gross pathologic examination of tissue from the ascending aorta was reviewed from 2010. The presence of patchy intimal fatty streaks and areas of atherosclerotic plaque were noted. Microscopic examination revealed severe fibrointimal thickening and marked destruction of the media. The usual organization of medial elastic fibers was lost, and there was both medial collapse and foci of necrosis. The

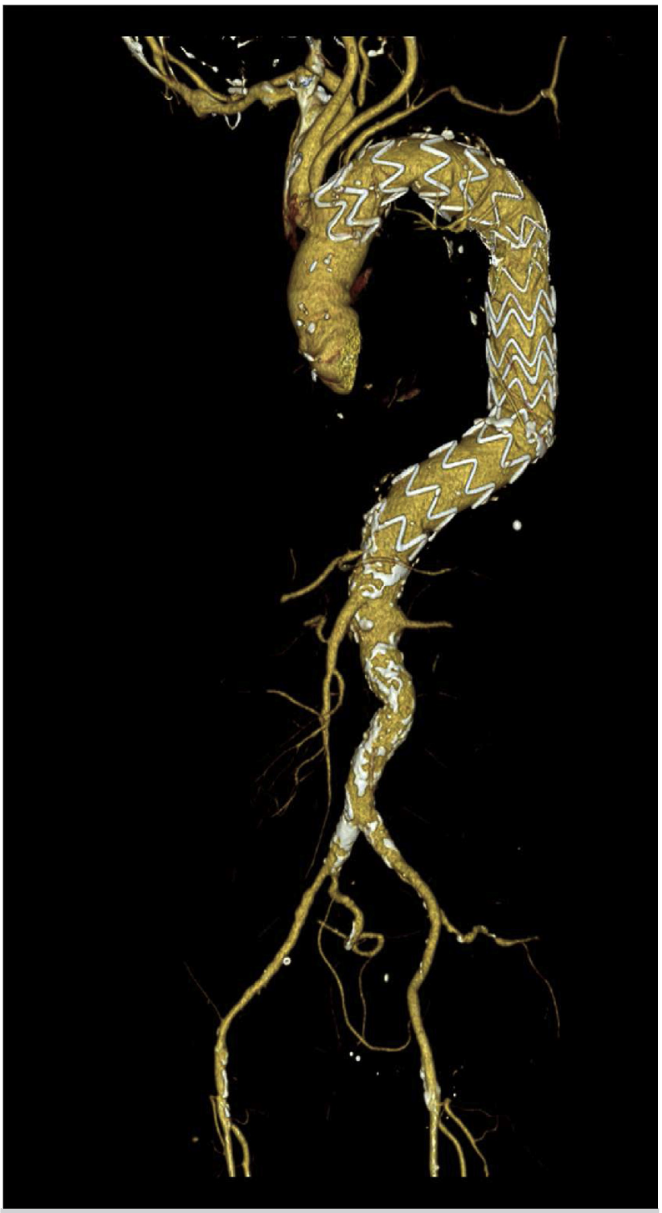

Fig 3. Three-dimensional rendition of the aorta with endovascular repair of the aortic dissection and aneurysm in the descending aorta on postoperative day 1. The proximal fixation of the stent graft is at the origin of the innominate artery and distal fixation is at the level of the celiac axis.

extensive medial disruption was appreciated on hematoxylin and eosin staining (Fig 4). The chronic medial inflammation was composed predominantly of plasma cells, lymphocytes, and giant cells.

Follow-up. The patient was discharged in stable condition with no spinal cord ischemic events. She was sent home with a tapering dose of prednisone over a 4-month period, with an eventual long-term dose of 5 mg/d. During follow-up, the patient underwent CT angiography imaging at 1, 6, and 12 months postoperatively; the stent graft was patent with no migration or endoleaks noted.

DISCUSSION

Aortitis is commonly classified as either infectious or noninfectious aortitis.[1] GCA is the most common cause of noninfectious aortic wall inflammation and it is involved in the development of TAAs[1] and has a preference for the thoracic aorta.[4] In particular, GCA is implicated in aneurysmal formation secondary to infiltration of giant cells, lymphocytes, and macrophages,[5] thus leading to necrosis of the medial layer of the aorta causing structural degeneration.[6] The triggers of this inflammatory process are not entirely known and the pathogenesis of GCA is not well-understood.

Advanced aortitis may be complicated by the presence of saccular aneurysms, pseudoaneurysms, and thrombus.[7] For those with GCA, aortic aneurysms occur in approximately 18 per 1000 patient-years.[8,9] TAAs, which develop secondary to GCA, have a greater risk of development and rupture in females as compared with males.[10-12] In a study by Kermani et al,[13] active aortic inflammation was noted in patients with dissection/rupture but not with isolated aneurysmal degeneration, suggesting that active aortitis may be a determinant of dissection/rupture in the setting of GCA. Other risk factors associated with TAAs include hypertension, atherosclerosis, smoking, older age, and those with Ehlers-Danlos or Marfan syndrome.[14]

Patients with GCA are 2.4 times more likely to develop abdominal aneurysms and 17.3 times more likely to develop TAAs.[4] In a retrospective cohort study, Nuenninghoff et al[8] found that GCA patients over the age of 50 developed aortic dissection (with no aneurysm development), which was fatal in 77% of cases. Other complications of TAAs secondary to GCA include ischemic stroke, upper extremity stenosis, and death.[15,16] In a rare case, Rynio et al[17] reported that secondary to the inflammatory process within the aortic wall, the integrity of the wall is compromised leading to increased fragility as compared with noninflammed aortic aneurysms.

Ironically, there is currently no diagnostic criteria that has been accepted for the diagnosis of GCA despite ongoing efforts.[18] Temporal artery biopsy remains the gold standard for diagnosing GCA,[19] with a sensitivity of 15% to 40% and a specificity of 100%.[20] However, other diagnostic criteria can be met to establish a diagnosis of GCA. This includes age greater than 50 years, new headache, temporal artery abnormalities, elevated erythrocyte sedimentation rate of greater than 50 mm/h, and abnormal arterial biopsy results. At present if three of these five criteria are met, patients are diagnosed with GCA.[18]

Radiographic studies show that 67% of patients with a new diagnosis of GCA have involvement of the aorta[21]

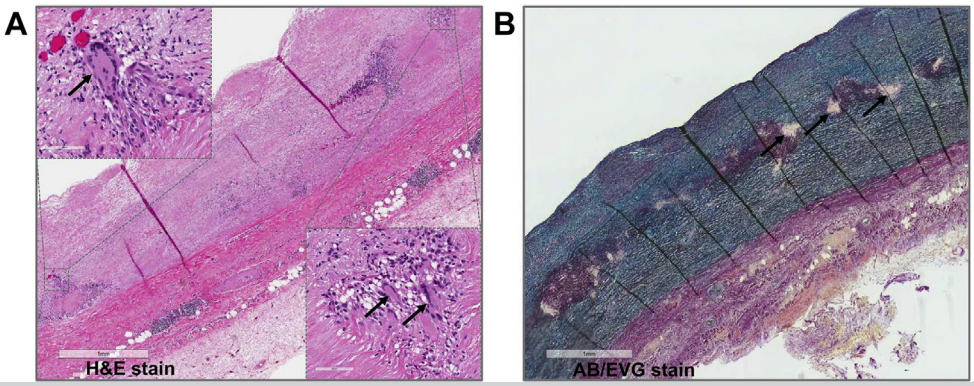

Fig 4. A, Low-power view of the aortic wall. *Insets*: Multinucleated giant cells (*arrows*) and mixed lymphoplasmacytic inflammation of the media. **B,** Aortic wall stained with AB/EVG demonstrating marked disruption of the elastic fibers, medial collapse, and areas of necrosis (*arrows*).

where a mural wall thickness of greater than 2 to 3 mm and periaortic inflammatory signs are diagnostic for aortitis.[7] Aortitis may also be seen as a long segment of aorta with a thickened wall that tapers distally, especially in the descending aorta and subclavian arteries.[7,22]

CONCLUSIONS

GCA is linked with a greater risk of aneurysmal degeneration, specifically aortic aneurysms and dissections. This case provides an example of a patient with previous ascending aortic deterioration with eventual descending aortic involvement through aneurysmal formation and dissection several years later from diagnosis of GCA. Complications can be sudden and life threatening.[23] Both radiologic and historical histologic data supported the presence of inflammatory aortitis with the presence of giant cells seen in the medial layer of the vessel. Our patient had specific risk factors associated with GCA such as age greater than 50 years,[24] female sex,[25] former smoker,[26] and European ethnicity.[27] The greatest of these is her advanced age, which is the strongest risk factor to developing GCA.[28] Also, with patients presenting with active aortic inflammation, physicians should have a high degree of suspicion regarding a risk of dissection. Thus, patients presenting with an acute aortic syndrome while having the aforementioned risk factors, the clinician should thoroughly evaluate the patient for the presence of GCA.

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