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Acute-on-Chronic Aortic Dissection in the Setting of Giant Cell Arteritis: A Case Report

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Abstract

Giant cell arteritis (GCA) is a chronic vasculitis of large- and medium-sized vessels that can lead to acute aortic adverse events. We describe a 74-year-old female patient who presented with acute-on-chronic aortic dissection. Her pathology was significant for aortitis in the setting of giant cell arteritis. The patient ultimately underwent successful repair of her dissection and was discharged home.

Introduction:

Giant cell arteritis (GCA) is a chronic vasculitis of large- and medium-sized vessels that tends to affect women and the elderly. One population study describes a two-fold higher incidence among women with a mean age at the time of diagnosis of about 75 years[1]. The classic presentation of GCA is headache, temporal artery sequelae, and elevated inflammatory markers. While there is a known association between GCA and aortic dissection, no known studies or case reports have described patients who presented with acute-on-chronic thoracic aortic dissection. We describe a patient who presented with thoracic aortic aneurysm with acute-on-chronic dissection secondary to GCA underwent successful repair.

Case Report:

The patient is a 74-year-old female with a past medical history significant for asthma, hypertension, diabetes, and rheumatoid arthritis on etanercept. She was initially admitted for respiratory failure due to pulmonary edema. She did not present with chest pain on admission. Her echocardiogram was significant for a decreased ejection fraction to 36%, and moderate to severe aortic regurgitation. Computed tomography angiogram (CTA) demonstrated an incidental aneurysmal ascending aorta measuring 5.9 cm in diameter with dissection that did not involve the arch vessels, as well as a small penetrating atherosclerotic ulcer. The patient was taken to the operating room for open repair of her ascending aortic dissection. No blood was encountered when the pericardium was opened. Upon entry, the walls of the dissected aorta were noted to be thickened and densely adherent to the pulmonary trunk and the right pulmonary artery, consistent with an acute-on-chronic aortic dissection. The dissection was limited to the ascending aorta that did not extend proximal to the sinotubular junction. She underwent open repair with a 34mm Gelweave supra-coronary graft and her aortic valve was repaired using a sub-comissural annuloplasty. Postoperative echocardiogram demonstrated near complete resolution of her aortic insufficiency. Her postoperative course was uneventful, and she was discharged home after her stay in rehabilitation. Her pathology was significant for aortitis with poorly formed granuloma, giant cells, and medial necrosis, consistent with giant cell arteritis (Figure 1).

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Comment:

For GCA in large vessels, the incidence of aortic dissection appears to increase over time, likely due to aortic inflammation seen in patients with GCA[2]. When the aorta is affected in giant cell arteritis, it commonly results in thoracic aortic aneurysm, with almost a 17-fold increase compared to age and gender-matched controls[3,4]. One study showed that 6% of patients with GCA with aneurysm developed a dissection. All of these dissections involved the thoracic aorta, mostly the ascending portion (72%), with the size of the aneurysm ranging from 29-80 mm[5]. Another study showed that in patients with GCA with aortic aneurysm and dissection, aneurysm size was not a predictor of dissection[6].

In this case, our patient presented with shortness of breath which was initially attributed to aortic insufficiency. On workup for her respiratory failure, she was found to have an acute-on-chronic ascending aortic dissection. One study noted similar findings in patients who presented with aortic dissection. Most patients were asymptomatic, but 7 out of 37 patients in the study presented with shortness of breath due to aortic insufficiency, and 6 out of 37 patients had congestive heart failure on presentation[7]. Patients with underlying autoimmune diseases, such as rheumatoid arthritis, should be followed closely. Currently, there are no guidelines that suggest the amount of time for follow up. We recommend that these patients undergo a thorough clinical exam and imaging with CTA yearly. Additionally, those with any aortic dilation should be followed closely for worsening aneurysm and possible dissection. Maintaining a close relationship with the patient's rheumatologist and primary care physician can help ensure control of the patient's rheumatological disease, enabling early detection and treatment and preventing mortality.

Disclosures:

The authors do not have any disclosures to report.

References:

1. Salvarani C, Crowson CS, O'Fallon WM, Hunder GG, Gabriel SE. Reappraisal of the epidemiology of giant cell arteritis in Olmsted County, Minnesota, over a fifty-year period. *Arthritis Rheum.*2004;51(2):264-268.

2. Nuenninghoff DM, Hunder GG, Christianson TJ, McClelland RL, Matteson EL. Incidence and predictors of large-artery complication (aortic aneurysm, aortic dissection, and/or large-artery stenosis) in patients with giant cell arteritis: a population-based study over 50 years.*Arthritis Rheum.* 2003;48(12):3522-3531.

3. Erbel R, Aboyans V, Boileau C, et al. 2014 ESC Guidelines on the diagnosis and treatment of aortic diseases: Document covering acute and chronic aortic diseases of the thoracic and abdominal aorta of the adult. The Task Force for the Diagnosis and Treatment of Aortic Diseases of the European Society of Cardiology (ESC). *Eur Heart J.*2014;35(41):2873-2926.

4. Gornik HL, Creager MA. Aortitis. *Circulation.*2008;117(23):3039-3051.

5. Kebed DT, Bois JP, Connolly HM, et al. Spectrum of Aortic Disease in the Giant Cell Arteritis Population. *Am J Cardiol.*2018;121(4):501-508.

6. Kermani TA, Warrington KJ, Crowson CS, et al. Predictors of Dissection in Aortic Aneurysms From Giant Cell Arteritis. *J Clin Rheumatol.* 2016;22(4):184-187.

7. Zehr KJ, Mathur A, Orszulak TA, Mullany CJ, Schaff HV. Surgical treatment of ascending aortic aneurysms in patients with giant cell aortitis. *Ann Thorac Surg.* 2005;79(5):1512-1517.

Figure Legend:

Figure 1 - Aortitis with poorly formed granuloma, giant cells, and medial necrosis, consistent with giant cell arteritis

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