A T2-Weighty Discovery: Aortitis on Cardiac MRI with Histopathologic Correlation

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CASE PRESENTATION

A 39-year-old man of South Asian descent with a history of exertional dyspnea and gradual reduction in functional capacity presented with an early diastolic precordial murmur consistent with aortic regurgitation. He had no medical history of note, denied constitutional symptoms, and did not have signs or symptoms supporting polymyalgia rheumatica or a systemic large-vessel vasculitis. Diminished pulses and bruits were absent on physical examination. Transthoracic echocardiography revealed a dilated aortic root and severe aortic regurgitation with preserved right and left ventricular (LV) systolic function. Cardiac magnetic resonance (CMR) imaging revealed a tricuspid aortic valve with severe aortic regurgitation (regurgitant volume 69 mL, regurgitant fraction 51%), a dilated aortic root (5.1 cm at the sinuses of Valsalva), a dilated ascending thoracic aorta (4.5 cm orthogonal dimension), and a dilated eccentrically hypertrophied LV (end-diastolic and systolic dimensions of 6.5 cm and 5.0 cm, respectively). His quantitative LV ejection fraction was 53%. The ascending aorta and aortic arch had increased wall thickness and were hyperenhanced after gadolinium-based contrast administration, suggesting aortitis (Figure 1). Magnetic resonance angiography revealed normal aortic arch branch vessel anatomy with no evidence of stenoses, occlusions, or aneurysms (Figure 2).

The patient was referred to cardiothoracic surgery and underwent a mechanical aortic valve replacement and ascending thoracic aorta replacement with a Dacron tube graft. Intraoperatively, the ascending aorta was noted to be diffusely thickened down to the aortic sinuses and the coronary ostia appeared friable. Anatomic histopathology revealed extensive chronic and focal acute inflammatory changes in the ascending aortic wall and the presence of giant cells (Figure 3). Gram stain and acid-fast bacilli stain were negative, as were aerobic, anaerobic, fungal, and mycobacterial cultures. A definitive diagnosis had not been determined, with noninfectious etiologies such as idiopathic aortitis or an inflammatory vasculitis remaining on the differential. While nonspecific, the patient had a preoperative microcytic anemia (hemoglobin 12.3 g/dL, mean corpuscular volume 75 fL).

Noninfectious causes of aortitis represent a heterogeneous group of conditions that include idiopathic or clinically isolated aortitis,

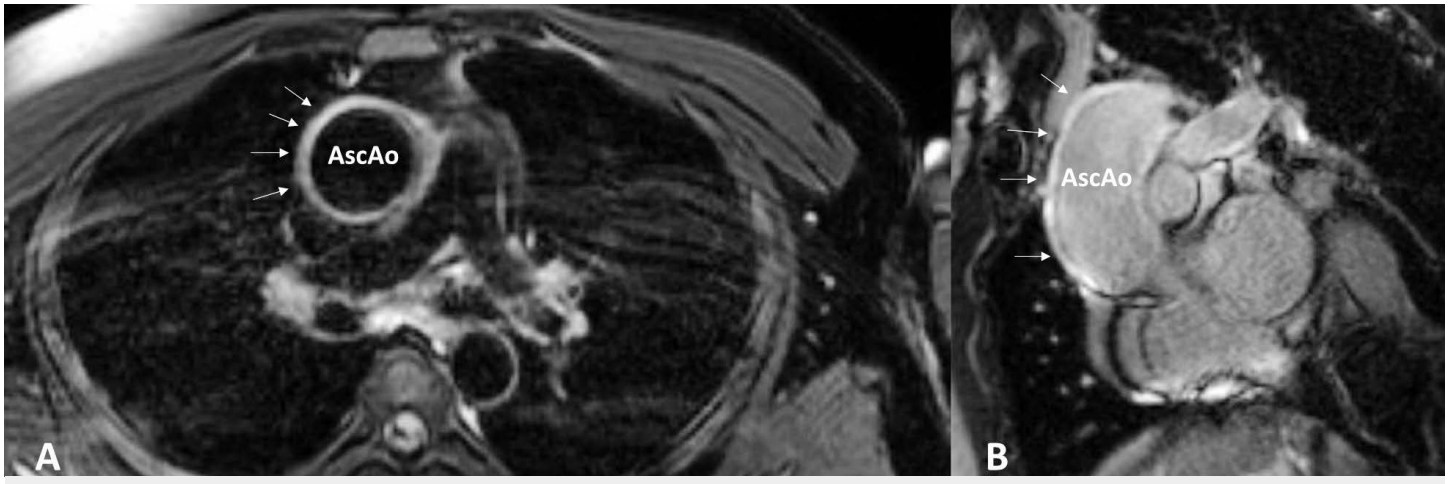

Figure 1.
(A) Axial cardiac magnetic resonance slice demonstrating diffuse, circumferential ascending aortic wall thickening (arrows) on T2-weighted fat saturation imaging. (B) Late gadolinium enhancement imaging reveals hyperenhancement (arrows) of the ascending aorta and proximal aortic arch. AscAo: ascending aorta

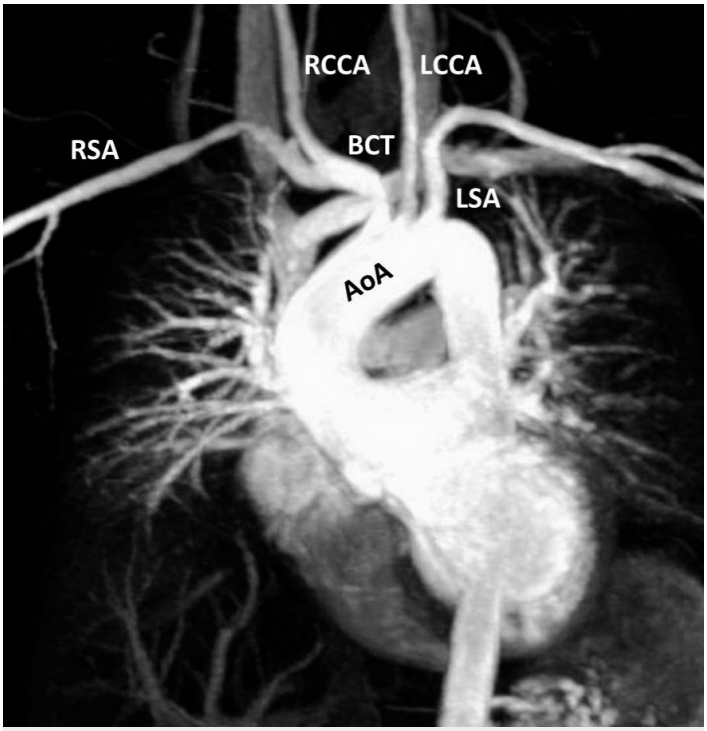

Figure 2.
Three-dimensional gadolinium-enhanced maximum intensity projection rendering of aortic arch and major branch vessels. No significant stenoses, occlusions, or aneurysms are seen. AoA: aortic arch; RSA: right subclavian artery; LSA: left subclavian artery; RCCA: right common carotid artery; LCCA: left common carotid artery; BCT: brachiocephalic trunk

large-vessel inflammatory vasculitides (eg, giant cell arteritis, Takayasu arteritis), autoimmune conditions (eg, Behçet's disease, sarcoidosis, Sjögren's syndrome), and other inflammatory conditions such as ankylosing spondylitis or antineutrophil cytoplasmic antibody associated vasculitides.[1] Characteristic CMR findings across a spectrum of underlying causes include wall thickening and increased wall edema as evidenced by increased T2-weighted signal intensity and hyperenhancement after gadolinium-based contrast injection.[2-4] Histopathologic findings, including the presence of giant cells, often overlap among different etiologies,[5] highlighting the importance of a carefully revisited history and physical examination as well as directed laboratory testing and multimodality aortic imaging.

Our case demonstrates classic CMR findings of aortitis with radiopathologic correlation. While the patient's demographic profile argues against giant cell arteritis or Takayasu arteritis, further investigations are needed before a diagnosis of idiopathic aortitis should be entertained.

Keywords:
aortitis, cardiac magnetic resonance, aortic regurgitation, dilated root

REFERENCES

1. Keser G, Aksu K. Diagnosis and differential diagnosis of large vessel vasculitides. Rheumatol Int. 2019 Feb;39(2):169-185.

2. Narváez J, Narváez A, Nolla JM, Sirvent E, Reina D, Valverde J. Giant cell arteritis and polymyalgia rheumatica: usefulness of vascular magnetic res-

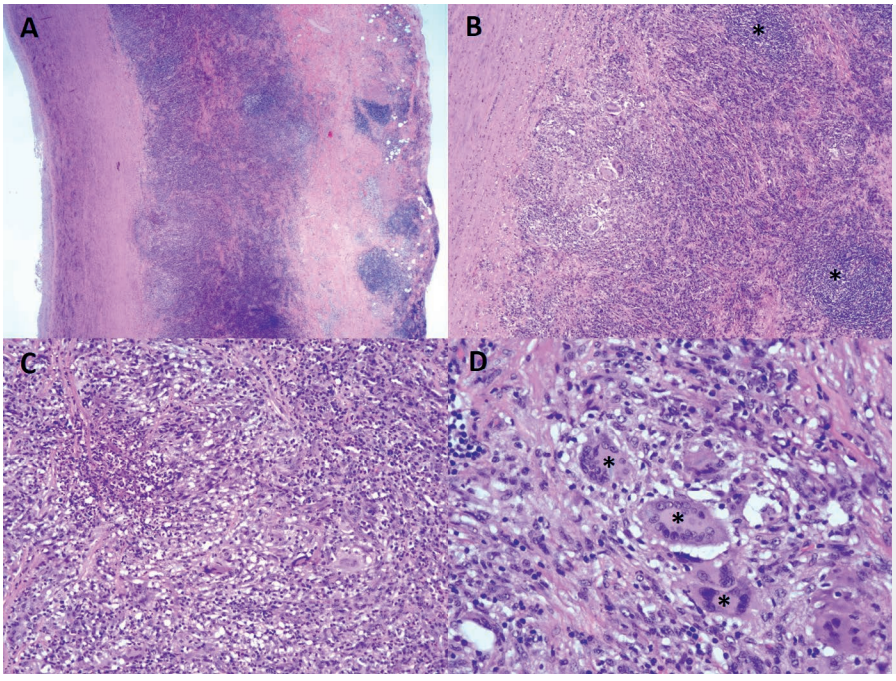

Figure 3.
(A) Histology of the ascending aortic wall demonstrating abnormal wall thickness up to 0.7 cm, (B, asterisks) lymphoid aggregates in the tunica media and adventitia, (C) predominantly lymphoplasmacytic inflammatory cell infiltrates, and (D, asterisks) multinucleated giant cells.

onance imaging studies in the diagnosis of aortitis. Rheumatology (Oxford). 2005 Apr;44(4):479-83.

3. Looi JL, Pui K, Hart H, Edwards C, Christiansen JP. Valvulitis and aortitis associated with ankylosing spondylitis: early detection and monitoring response to therapy using cardiac magnetic resonance imaging. Int J Rheum Dis. 2011 Oct;14(4):e56-8.

4. Roghi A, Pedrotti P, Milazzo A, et al. Acute myocardial infarction and cardiac arrest in atypical Takayasu aortitis in a young girl: unusual diagnostic role of cardiac magnetic resonance imaging in emergency setting. Circulation. 2010 Apr 13;121(14):e370-e375.

5. Cinar I, Wang H, Stone JR. Clinically isolated aortitis: pitfalls, progress, and possibilities. Cardiovasc Pathol. 2017 Jul - Aug;29:23-32.