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Case Report

Three-Dimensional Multi-Detector Computed Tomography of IgG4 Coronary (Peri) Arteritis: Expanding the Differential for Giant Coronary Artery Aneurysms

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Received: April 02, 2018; **Published:** June 06, 2018

Abstract

IgG4 coronary (peri) arteritis is a rare manifestation of IgG4 related systemic disease. Given the advances in clinical differentiation of disease, it has emerged as a new entity of autoimmune systemic disease. It has been known to affect multiple organs systems including the cardiovascular system. In this case, it can cause a vasculitis like picture in both the peripheral arterial system, but in the coronary arteries as well with aneurysmal dilation with circumferential arterial wall thickening best seen on cardiac computed tomography. We report a case in which a patient who had a history of biopsy proven IgG4 systemic disease presented with a non-ST-elevation myocardial infarction.

Keywords: *IgG4 Systemic Disease; MDCT; 3D-MDCT*

Abbreviations

IVUS: Intravascular Ultrasound; OCT: Optical Coherence Tomography; 3D-MDCT: 3D-Multidetector Computed Tomography

Introduction

IgG4 related systemic disease, has been identified as a new clinicopathologic entity is in the focus of great interest recently [1]. The disease can affect multiple organs (pancreas, salivary glands, retroperitoneum) and also has cardiovascular manifestations [2-4]. In the recent years there is growing evidence that IgG4 systemic disease can cause coronary arteritis and multiple coronary aneurysms [2-5]. We report a case in which a patient who had a history of biopsy proven IgG4 systemic disease presented with a non-ST-elevation myocardial infarction.

Case Report

A 56 year old male with biopsy-proven IgG4 colitis, which was well controlled on methotrexate and vedolizumab, presented with findings consistent with non-ST-elevation myocardial infarction (new onset chest pain and exertional shortness of breath with new onset

left bundle branch block without ST elevation with a peak Troponin-I of 17.6 ng/mL). Coronary angiography was performed and demonstrated giant fusiform aneurysms intermixed with stenoses of the left main, proximal left anterior descending, proximal left circumflex, and proximal right coronary arteries (Figure 1A-1D). At the time of angiography, intravascular ultrasound (IVUS) and optical computed tomographic (OCT) imaging were unavailable. To better delineate the aneurysms, 3D multi-detector computed tomographic (3D-MDCT) angiography was performed using a 256 slice CT scanner (Brilliance iCT, Philips Healthcare, Cleveland, OH, USA). Collimation was 0.625-mm, field-of-view 200 mm, scan voltage 120 kV, tube current 500 mA, and gantry rotation 270ms. Prospective ECG-gating was used, targeting a diastolic phase at 75% of the R-R cycle (Figures 2-5). As seen with the 3D-MDCTA assessment of coronary morphology, one can see both circumferential thickening of the arterial wall as well as luminal expansion. Given the three-vessel aneurysmal disease, the patient was referred for bypass surgery that resulted in a left internal mammary artery graft to the left anterior descending artery, right internal mammary artery graft to the obtuse marginal, and saphenous vein graft to the posterior descending artery. Intraoperative echocardiography was performed and aneurysms of the left circumflex can be visualized (Figure 6). Post-operatively the patient continues to do well with resolution of all cardiac symptoms and is maintained on chronic immunosuppressants and an appropriate post-operative cardiac regimen.

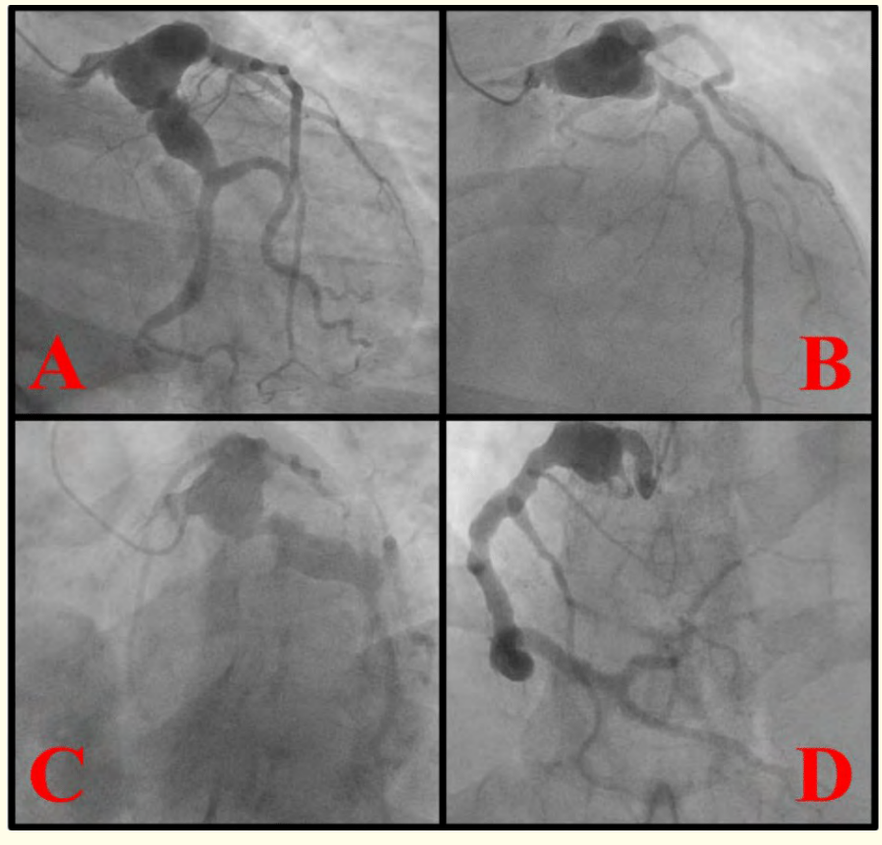

Figure 1: *Representative coronary angiogram of the patient. RAO Caudal projection (A) demonstrated a giant coronary artery aneurysm involving the left main, left circumflex, and proximal left anterior descending with relatively normal vessels beyond the aneurysm and no obvious atherosclerotic disease. RAO Cranial projection (B) demosntrates that the let anterior descending is involved proximal to the first diagonal. LAO Caudal projection (C) of the left coronary system demonstrates aneurysmal dilations with intervening stenosis in the proximal left circumflex. LAO Cranial projection (D) of the right coronary artery illustrates the large proximal aneurysm with mild distal tortuosity without significant stenosis.*

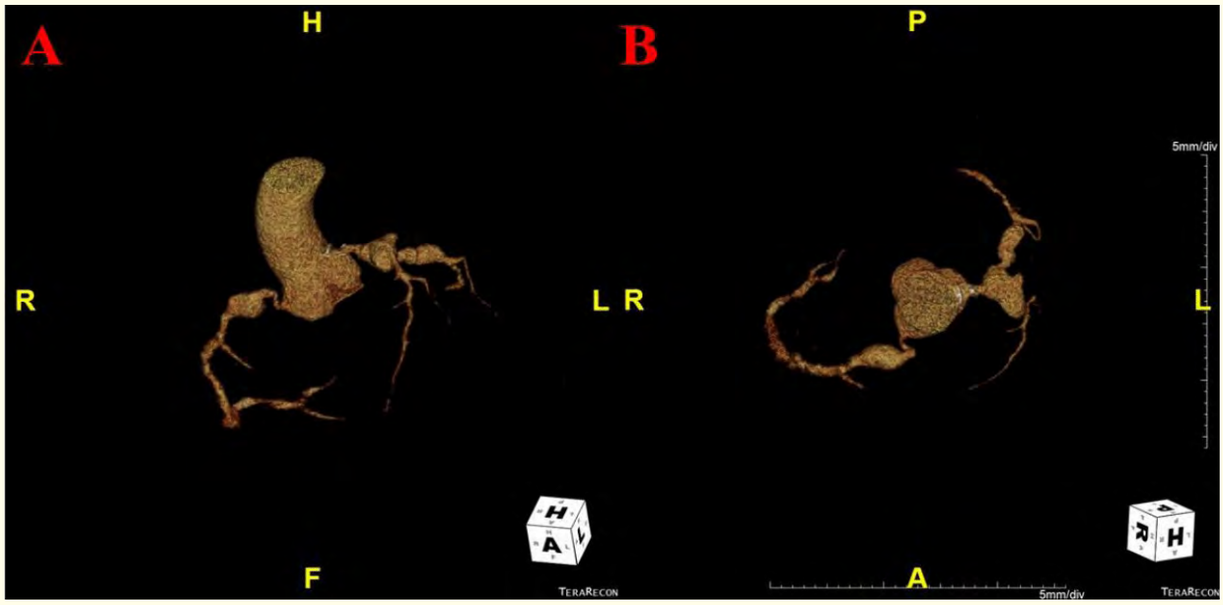

Figure 2: *Three-dimensional reconstruction of the entire coronary arterial tree demonstrating giant aneurysms involving the proximal coronary arteries. (A) represents an antero-posterior projection and (B) represents a superior-inferior projection. These reconstructions aid in delineating the relationship of the aneurysm to the aorta and the involvement of the ostium of the vessels. In the cases of the LCA and RCA, it appear that the aneurysm spares the aorta and the ostia. Lack of aortic involvement may aid in delineating large and medium-sized vessel vasculidities.*

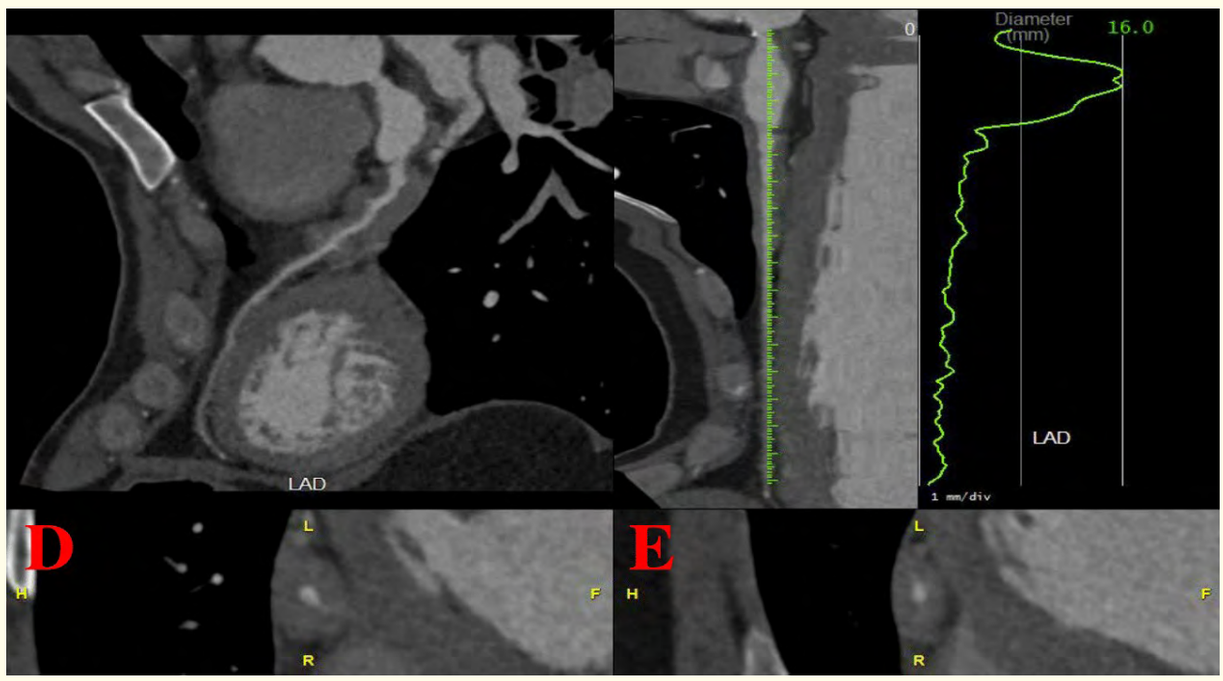

Figure 3: *Left anterior descending reconstruction. (A) demonstrates the course of the LAD from aorta to left ventricular apex. Clear soft-tissue thickening (tumefactive lesions) are noted encasing the proximal and mid LAD. (B and C) demonstrate a vessel diameter map showing that the largest portion of the LAD aneurysm is approximately 16.0 mm in diameter. (D and E) are cross-sectional images showing the luminal narrowing and circumferential adventitial thickening.*

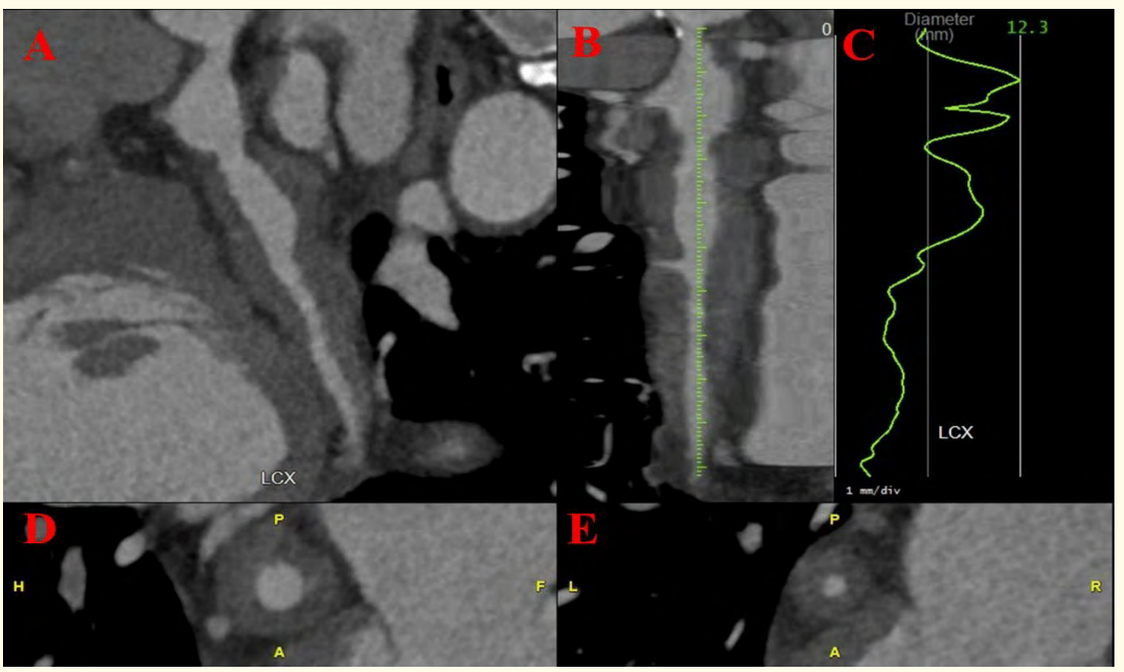

Figure 4: *Left circumflex reconstruction. (A) demonstrates the course of the LCx from aorta to left ventricle. Clear soft-tissue thickening are noted encasing entire length of the vessel. (B and C) demonstrate a straight-multiplanar reconstruction and correlative vessel diameter map showing that the largest portion of the aneurysm is approximately 12.3 mm in diameter. (D and E) are cross-sectional images showing the circumferential adventitial thickening combined with dilation (D) or stenosis (E).*

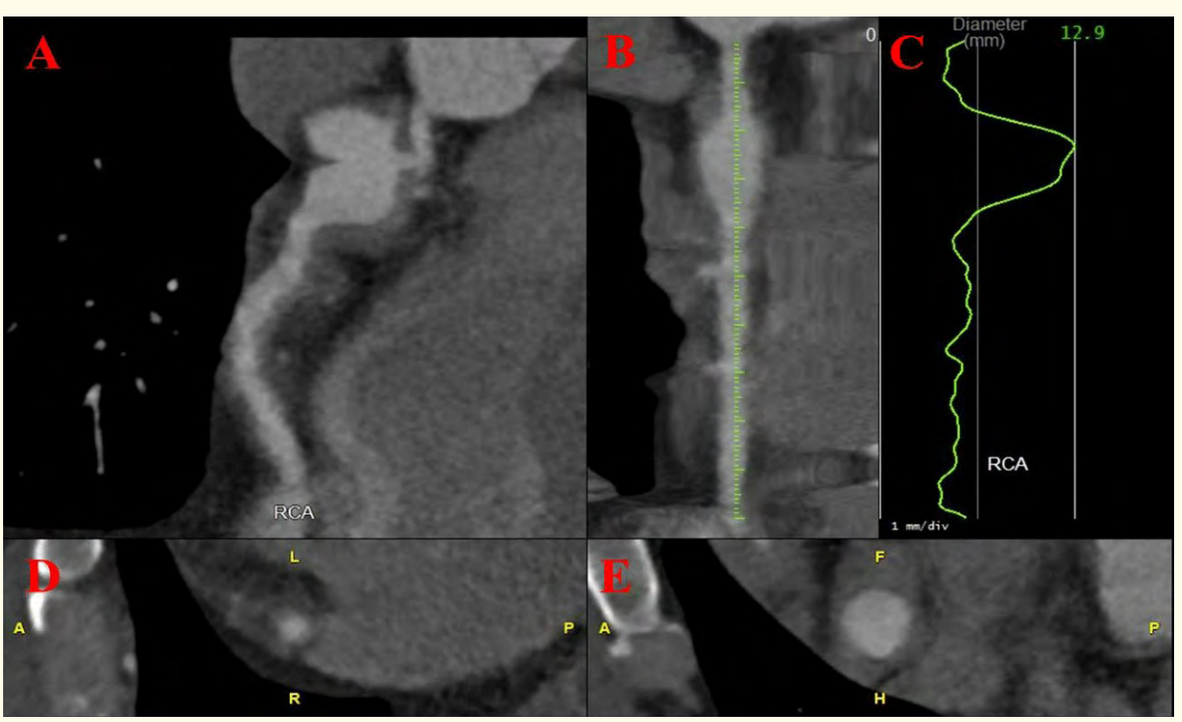

Figure 5: *Right coronary artery reconstruction. (A) demonstrates the course of the RCA originating from the aorta. The ostium of the coronary is spared, however, there is a large, multi-pedunculated aneurysmal dilation and adventitial thickening. (B and C) demonstrate a straight-multiplanar reconstruction and correlative vessel diameter map showing that the largest portion of the aneurysm is approximately 12.9 mm in diameter. (D and E) are cross-sectional images showing the circumferential adventitial thickening combined with stenosis (D) and dilation (E).*

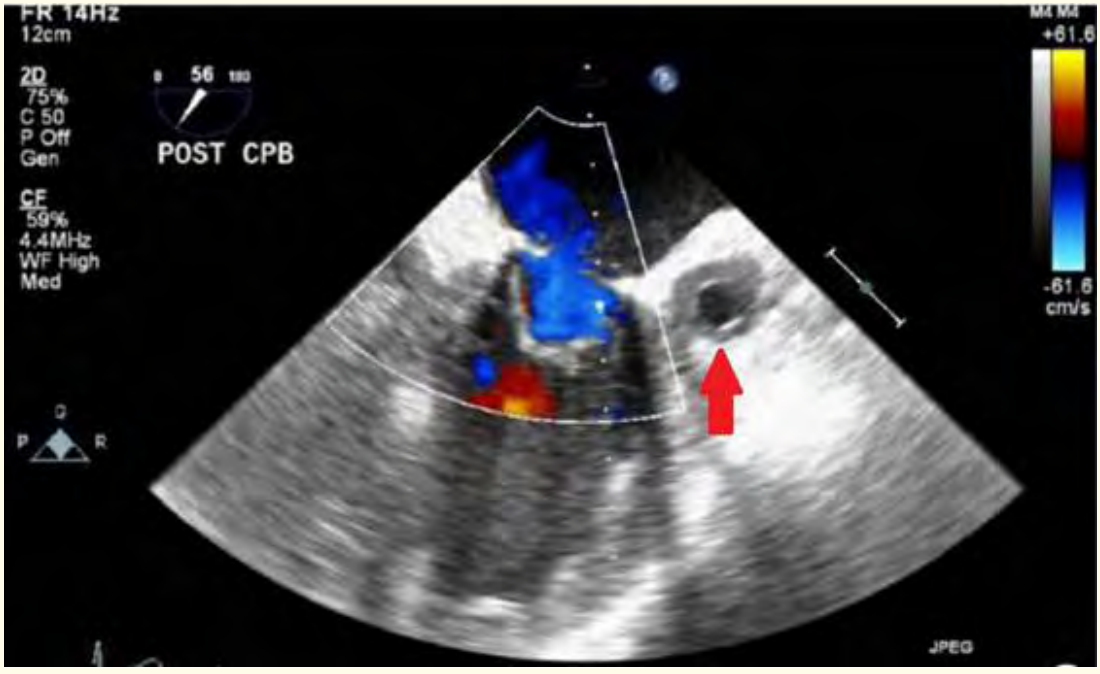

Figure 6: *Intraoperative echocardiography. Demonstration of aneurysmal dilation and arterial wall thickening of the left circumflex (Red Arrow).*

Discussion

IgG4-associated coronary (peri) arteritis is a rare manifestation of a larger multisystem disease with presentation ranging from non-specific cardiac symptoms to sudden death[1]. The differential diagnosis includes Kawasaki disease, Takayasu's arteritis, giant cell arteritis, polyarteritis nodosa, Erdheim-Chester, histiocytosis, sarcoidosis, and lymphoma. Coronary angiography may reveal large aneurysms, long-segment stenoses, or both. Computed tomographic angiography typically reveals circumferential arterial wall thickening with aneurysmal dilatations, tumefactive lesions, and long regions of compressive stenosis. IVUS and OCT may reveal similar features with poor to differentiate circumferential arterial wall thickening versus normal parenchyma using these technologies, as such 3D-MDCT angiography is preferable to intravascular imaging for this reason [4,6]. Fibrosis and infiltration of IgG4-positive plasamacytes into the media and adventitia of the coronaries is pathognomonic. Therapeutic options may include steroids, immunosuppressants, and intravenous monoclonal antibodies. If coronary arterial stenoses are found, treatment should ensue based on standard guidelines for the management of stable and acute coronary syndromes, however, the long term outcomes of drug-eluting stents in the setting of inflammatory arteritis have not been studied [5].

Conclusion

IgG4 Coronary (Peri) Arteritis is a rare manifestation of a rare disease that is seldom seen. IgG4 related disease should be included in the differential whenever giant coronary aneurysms are observed. Currently, in a non-emergent situation, it appears that 3D-MDCT angiography is the best way in which to observe the concentric arterial wall dilation due to its widespread availability versus intravascular imaging which is limited by luminal size and arterial wall thickness.

Conflicts of Interest

None.

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Volume 5 Issue 7 July 2018