Utility of multimodality imaging in diagnosis and follow-up of aortitis

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CASE REPORT

A 47-year-old male of European descent without any cardiac risk factors initially presented in 2005 with chest discomfort precipitated by emotional stress. Figure 1 illustrates work-up with multiple non-invasive cardiac imaging and invasive angiography studies prior to referral with suspected aortitis in 2013. At the time of his evaluation in 2013, he continued to have fleeting episodes of chest discomfort with emotional stress. Review of symptoms included recurrent aphthous ulcers and acne. Serologic work-up for relevant immunologic and infectious etiologies was negative, except for mildly elevated high sensitivity-C reactive protein (2.8 mg/L). Based on the findings of multimodality imaging in 2013, which included magnetic resonance angiogram (MRA) (limited due to motion), computed tomography angiogram (CTA) and Fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG-PET/CT) (Figure 2) and chronic symptoms with serologic work-up, a diagnosis of aortitis of undetermined etiology vs Behcet's disease was considered. Follow-up imaging on corticosteroid therapy (initial high dose of prednisone 50 mg daily followed by taper to 20 mg) (Figure 3) and after termination of steroid therapy (Figure 4) demonstrated initial improvement followed by recrudescence.

DISCUSSION

Aortitis is an inflammation of the aortic wall, and the most common etiologies include infectious, immunologic, and idiopathic.[1] While clinical presentation is often non-specific, accurate and early diagnosis is critical as it can be life threatening.[1] Invasive angiogram (IA), ultrasonography, MRA, CTA, and ^{18}F-FDG-PET have been used for imaging aortitis. Although early imaging is essential for confirmation of diagnosis, no specific protocol exists (Figure 5).[1-3]

Our case illustrates the challenges in diagnosing aortitis even in this era of multimodality imaging and highlights the key differences between anatomic and functional methods. While MRA/CTA may be performed as the initial study, metabolic imaging with ^{18}F-FDG-PET helps determining the disease activity. Although serum biomarkers are routinely used for monitoring the disease activity and guide therapy, ^{18}F-FDG-PET may be superior in identifying relapse as in this case.[3] Hybrid imaging using ^{18}F-FDG-PET with CTA/MRA may allow for simultaneous anatomic localization as well as determine the inflammatory activity, providing a complimentary and complete assessment.

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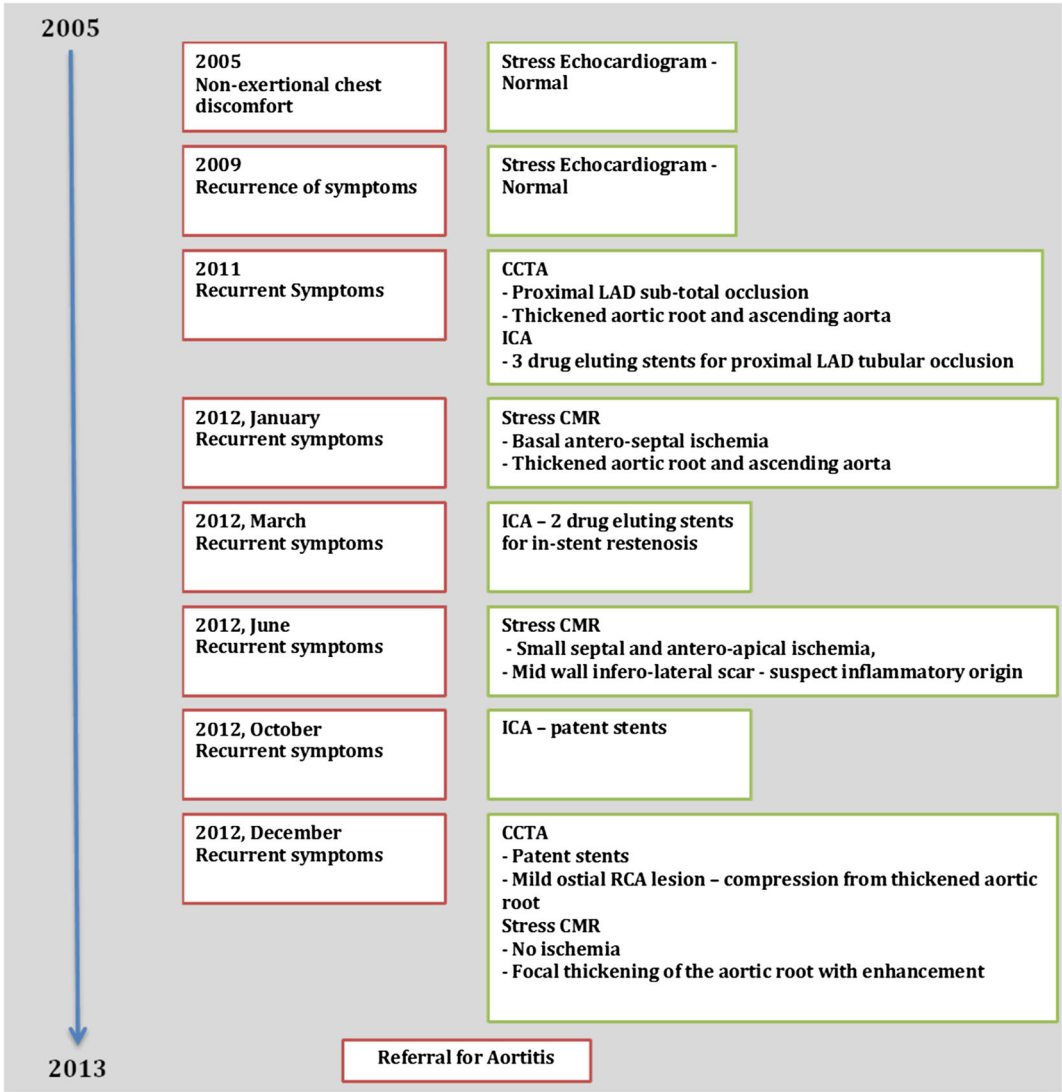

Figure 1. Flow chart of the work-up prior to referral. A stress echocardiogram was normal in 2005, as was a repeat study performed 4 years later. In early 2011, computed tomography (CT) coronary angiogram was performed for similar symptoms and showed sub-total proximal left anterior descending (LAD) occlusion. Subsequent invasive coronary angiogram (ICA) led to revascularization of an occluded non-calcified and tubular LAD lesion with placement of three drug-eluting stents. Due to continued non-exertional symptoms, patient underwent multiple non-invasive and invasive studies as detailed raising the suspicion of aortitis as the potential etiology. (*CCTA*, coronary computed tomography angiogram; *LAD*, left anterior descending coronary artery; *ICA*, invasive coronary angiogram; *CMR*, cardiac magnetic resonance; *RCA*, right coronary artery).

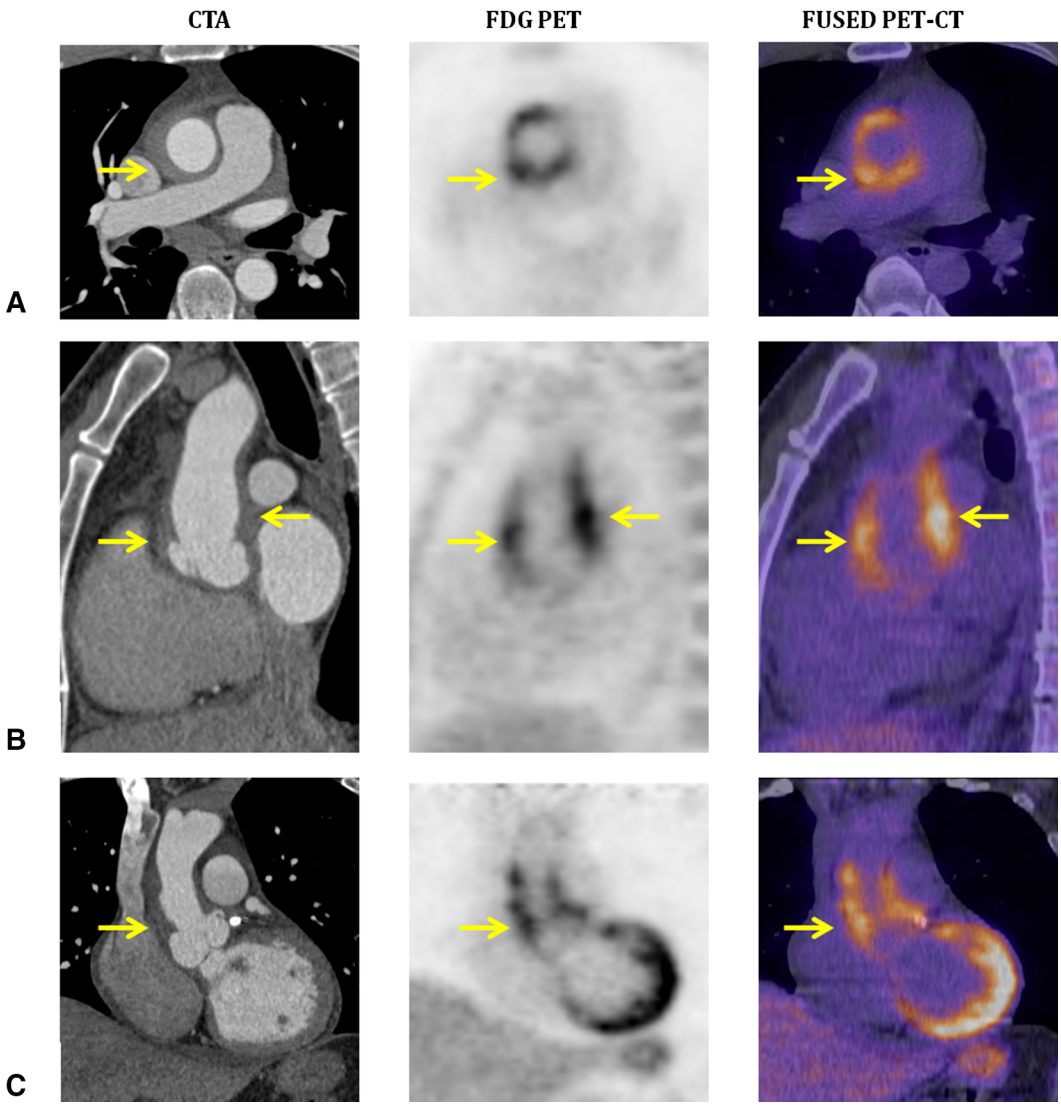

Figure 2. Pre-treatment Imaging. Axial (**A**), Sagittal (**B**), and Coronal (**C**) computed tomography angiogram (CTA) images demonstrating marked concentric thickening (maximal wall thickness of 10 mm) of the ascending aortic wall to the level of the brachiocephalic origin, along with involvement of the ostia of right and left coronary arteries, and a small pericardial effusion. Fluorine-18-fluorodeoxyglucose positron emission tomography (FDG-PET)/CT showed intense FDG uptake with a standard uptake value (SUV) of 4.9, corresponding to the areas of aortic wall thickening seen on CTA and consistent with active inflammation. Mildly elevated high sensitivity-C reactive protein (hs-CRP) of 2.8 mg/L. Based on the clinical presentation and imaging findings, a diagnosis of aortitis of undetermined etiology vs Behcet's disease was considered.

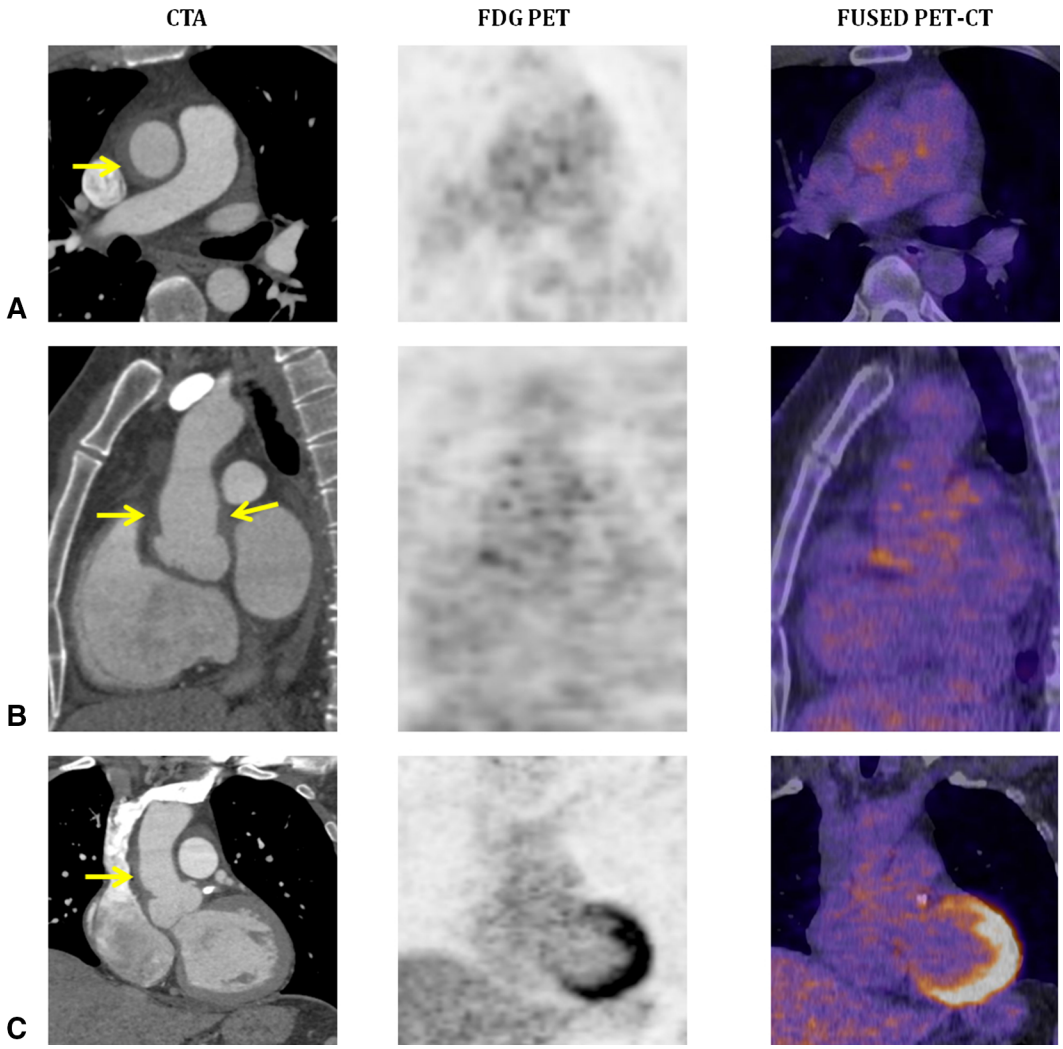

Figure 3. Repeat imaging 6 months on immunosuppressive therapy. Immunosuppressive therapy with prednisone 50 mg daily was initiated, and subsequently reduced to 20 mg daily after 2 months of therapy. Axial (**A**), Sagittal (**B**), and Coronal (**C**) computed tomography angiogram (CTA) still showing increased wall thickness (arrows) with maximal wall thickness of 8 mm, with an interval resolution of pericardial effusion. However, Fluorine-18-fluorodeoxyglucose positron emission tomography (FDG-PET)/CT showed a marked reduction in the focal FDG uptake in the ascending aorta, consistent with significant response to treatment. Reduction in FDG uptake was corresponding to the resolution of clinical symptoms and a reduction in hs-CRP level to 0.9 mg/L. This highlights the key difference between anatomic and functional imaging modalities. Changes in metabolic activity of inflammatory cells precede the anatomic changes and also correlate with the changes in clinical symptoms and biochemical markers of inflammation, such as hs-CRP.

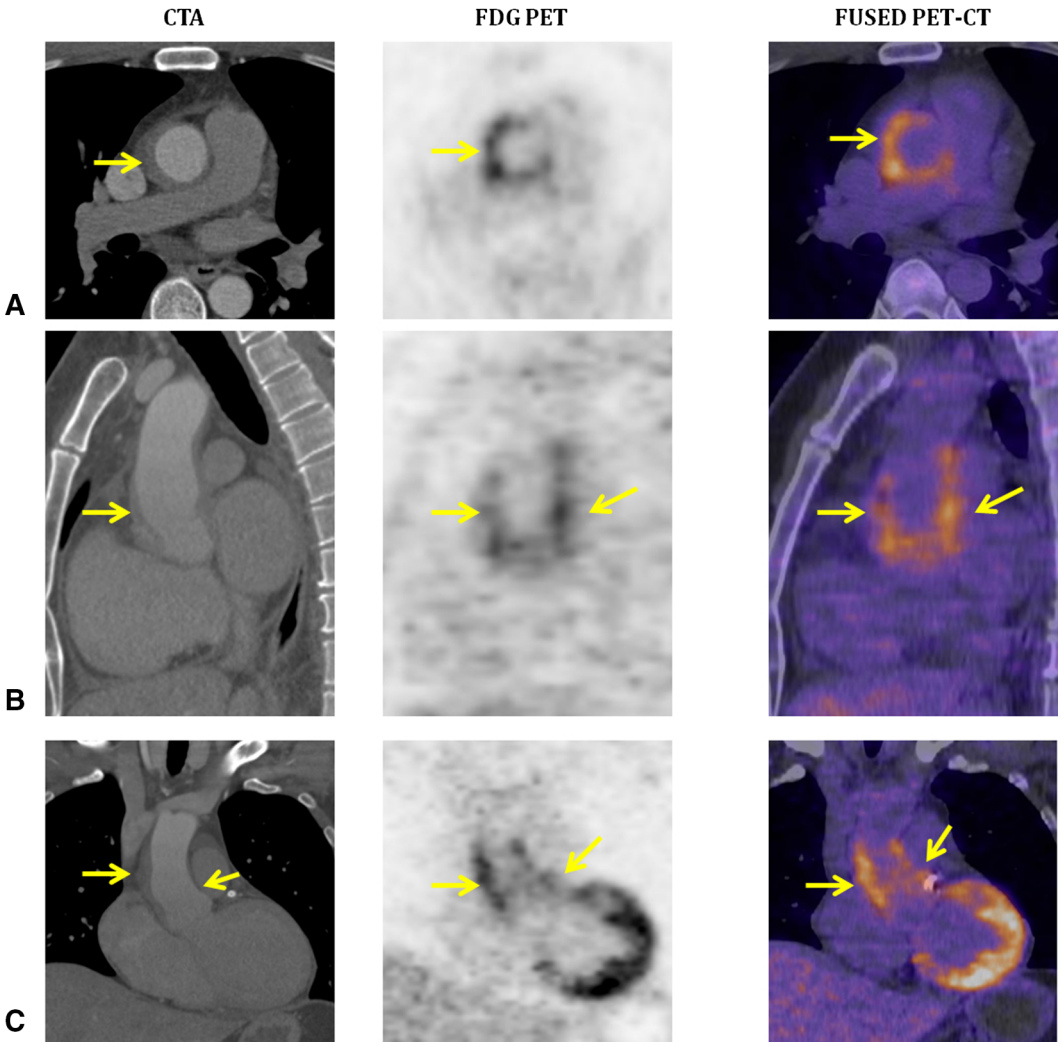

Figure 4. Repeat imaging on no therapy. Axial (**A**), Sagittal (**B**), and Coronal (**C**) computed tomography angiogram (CTA) showing marked concentric thickening of the ascending aortic wall with wall thickness of 10 mm, similar to the initial study. Fluorine-18-fluorodeoxyglucose positron emission tomography (FDG-PET)/CT showing moderately intense FDG uptake involving the wall of the ascending aorta with maximal standard uptake value (SUV) of 4.2. Although he remained asymptomatic, repeat imaging 6 months after the cessation of treatment showed recurrence with corresponding hs-CRP level was 2.6 mg/L. While this again highlights the key difference between anatomic and functional imaging, ^{18}F-FDG-PET imaging is also found to be superior in identifying relapse compared to biochemical markers.

Modality	Aortic Findings	Advantages	Disadvantages
MRA	• Wall thickening, mural edema (T2 weighted images) and fibrosis (GE images) • Detect stenosis, aneurysms and occlusions • Identify other etiologies such as dissection, IMH or penetrating ulcer	• Assess disease activity (T2 weighted and GE images) and soft tissue characterization • No concern of radiation exposure	• Longer scan time and need patient co-operation (breath holds) • Limited in patients with renal dysfunction and with implant/device
CTA	• Wall thickening and contrast enhancement • Detect stenosis, aneurysms and occlusions • Identify other etiologies such as dissection, IMH or penetrating ulcer	• High spatial resolution • Detect structural findings with high sensitivity and specificity • Guide planning for revascularization • Short scan time	• Poor characterization of disease activity and mural inflammation • Ionizing radiation and iodinated contrast (renal dysfunction and allergy)
FDG-PET	• FDG uptake by metabolically active cells such as inflammatory infiltrate • Normal vascular wall has no FDG uptake, and any uptake is pathologic	• Identify, monitor response to therapy and relapse (better than serum biomarkers) • Quantification of disease activity based on standardized uptake value • Whole body assessment is possible	• Limited availability and ionizing radiation • Non standardized methodologies, interpretations and reference standards • Limited spatial resolution and anatomic information • No differentiation of etiology of FDG uptake – atherosclerotic versus infectious versus immunologic
Ultrasound	• Wall thickening and presence of mural edema (hypo-echoic halo) • Detect stenosis, aneurysms and occlusions – with assessment of functional significance • May identify dissection, IMH or penetrating ulcer	• Easily available with good resolution • No radiation exposure	• Poor tissue characterization • Operator dependent • Limited by body habitus, bony structures and air
IA	• Detect stenosis, aneurysms and occlusions	• Excellent resolution • Guide therapeutic procedures (angioplasty or stent placement)	• No tissue characterization • Invasive, ionizing radiation and iodinated contrast (renal dysfunction and allergy)

MRA – magnetic resonance angiography; CTA – computed tomography angiography; FDG-PET – Fluorine 18- fluorodeoxyglucose positron emission tomography; IA – invasive angiography.

Figure 5. Illustrates the various imaging modalities used in assessment of aortitis.

Disclosure

The authors have no conflicts relevant to this work.

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