Coronary Artery Vasculitis and Encasement: Multimodality Imaging Findings and Mimics

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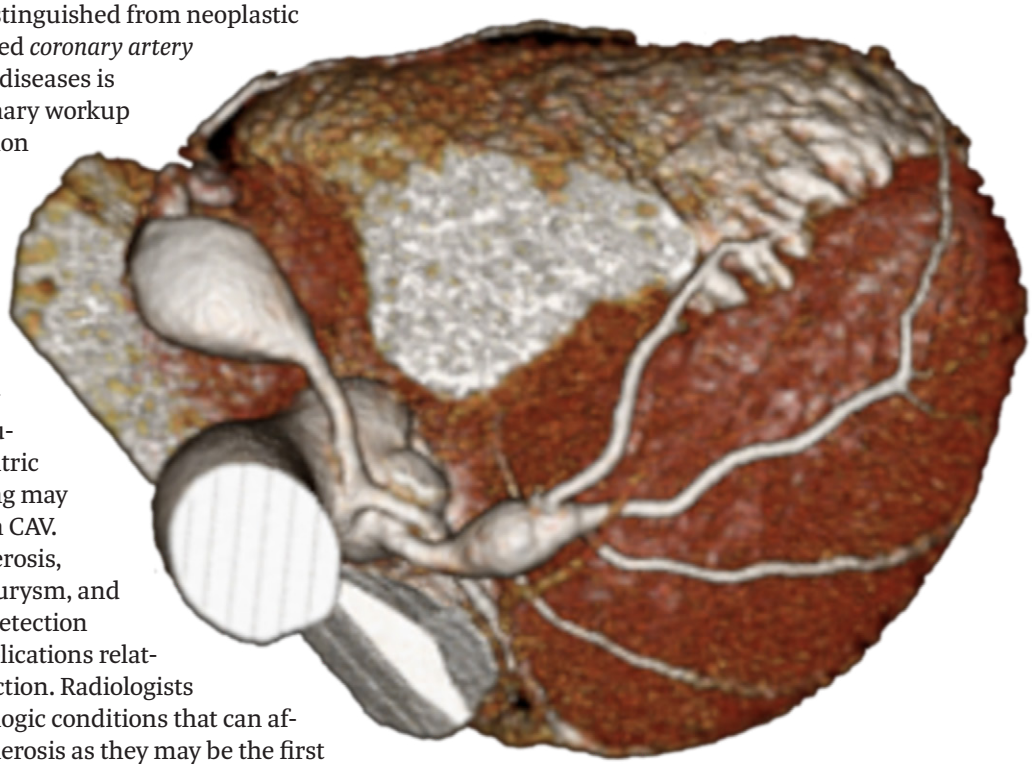
Coronary artery vasculitis (CAV) and coronary artery encasement are rarely diagnosed conditions that are important diagnostic considerations, particularly in patients with acute coronary syndrome without traditional cardiovascular risk factors or systemic illness. *Vasculitis* refers to inflammation of the blood vessel walls, which can be primary or secondary. This process should be distinguished from neoplastic involvement of the coronary arteries, termed *coronary artery encasement*. Prospective diagnosis of these diseases is challenging, often requiring multidisciplinary workup with careful attention to clinical presentation and multiorgan findings. While CAV and coronary artery encasement can be indistinguishable at coronary CT angiography, certain imaging features help order the differential diagnosis. CAV should be considered when there is smooth wall thickening that is circumferential and/or continuous. A diagnosis of coronary artery encasement is favored when there is irregular or nodular wall thickening that is eccentric to the vessel lumen. Epicardial fat stranding may also appear more extensive compared with CAV. Potential mimics of CAV include atherosclerosis, acute plaque rupture, coronary artery aneurysm, and spontaneous coronary artery dissection. Detection and diagnosis of CAV may help avoid complications related to accelerated atherosclerosis and infarction. Radiologists should be familiar with the range of pathologic conditions that can affect the coronary arteries beyond atherosclerosis as they may be the first to raise such diagnostic possibilities, guiding next steps in patient workup and management.



Introduction

Coronary artery vasculitis (CAV) and coronary artery encasement (ie, neoplastic involvement of the coronary arteries) are rarely diagnosed conditions but they are important diagnostic considerations, particularly in patients with acute coronary syndrome without traditional cardiovascular risk factors or systemic illness. With the increasing use of coronary CT angiography (CCTA) as a first-line modality for assessing coronary artery stenosis, imagers may unexpectedly encounter CAV and encasement. Likewise, detection and characterization of these entities can also occur at echocardiography, invasive catheter angiography, cardiac MRI, and fluorine 18 (^{18}F)–labeled fluorodeoxyglucose (FDG) PET (1).

There is little data on the overall frequency of CAV and encasement, likely related to underdiagnosis, low incidence rates, and variable detection rates across modalities (2). The prevalence of CAV and encasement at CCTA is thus unknown, although it can be estimated based on disease epidemiology (Table 1). CCTA can be used in the workup of patients with known systemic illness to determine coronary artery involvement, or the radiologist may be the first to raise such diagnostic possibilities if unexpectedly detected at CCTA. Prospective diagnosis of these diseases is challenging, often requiring multimodality and multidisciplinary workup with careful attention to clinical presentation and multiorgan findings.

Supplemental Material TestYour Knowledge CME

RadioGraphics 2024; 44(11):e24009
https://doi.org/10.1148/rg.240009

Content Codes: CA, CT

Abbreviations: ANCA = antineutrophil cytoplasmic antibody, CAA = coronary artery aneurysm, CAV = coronary artery vasculitis, CCTA = coronary CT angiography, FDG = fluorodeoxyglucose, GCA = giant cell arteritis, IgG4 = immunoglobulin G4, LAD = left anterior descending artery, RCA = right coronary artery, SCAD = spontaneous coronary artery dissection

TEACHING POINTS

- CAV should be considered when there is smooth wall thickening that is circumferential and/or continuous.
- Coronary artery encasement can be favored when there is masslike periarterial thickening that is eccentric relative to the vessel lumen. These lesions can have an irregular or nodular soft-tissue core with edematous epicardial fat stranding that can appear more exuberant compared with CAV.
- A helpful hint for vasculitis is the involvement of other variably sized vessels as well as other extravascular findings such as cavitary pulmonary lesions or renal disease. Likewise, given the heterogeneous causes of coronary artery encasement, unexpected findings in almost any organ can help suggest the diagnosis.
- The presence of calcified plaques and relatively well-defined eccentric arterial wall thickening favor atherosclerosis over vasculitis, which would typically appear as smooth concentric thickening without calcification. Extraluminal soft-tissue attenuation and periarterial stranding are atypical of atherosclerosis and should prompt consideration of an underlying vasculitic or encasement disorder.
- The appearances of most inflammatory and noninflammatory causes of CAAs are less predictable and are best assessed in combination with patient history and other systemic findings.

Classification

Although overlap exists in the literature, the authors of this article recommend that imagers classify cases based on whether they suspect a vasculitic process or an encasement disorder involving the coronary arteries (Table 2).

Vasculitis

Vasculitis refers to inflammation of the blood vessel walls, which can be primary or secondary (3). Noninfectious vasculitis can be categorized based on the size of the vessel involved (large, medium, and small) with the caveat that entities from each category may affect any vessel size. Systemic vasculitis can involve any cardiac tissue, which is associated with increased mortality (4). Overall, cardiac involvement likely occurs in less than 10% of patients with systemic vasculitis, but the rate is highly variable based on disease entity (eg, cardiac complications may occur in up to 60% of patients with Takayasu arteritis) (4,5). Cardiac involvement of systemic vasculitis includes pericarditis, myocarditis or cardiomyopathy, CAV (with associated complications such as stenosis and occlusion, aneurysm, and rupture), valvulopathy, and intracavitary thrombosis (4–7).

CAV should be considered when there is smooth wall thickening that is circumferential and/or continuous. There is generally periarteritis as well, although at CCTA this can be indistinguishable from wall thickening. Together, wall thickening and periarteritis manifest as soft-tissue thickening along a vessel, with sharper margins adjacent to the contrast material–filled lumen and ill-defined hazy borders along the epicardial fat, which can be edematous. Stenosis may be focal or multifocal, or there can be smooth segmental tapering of the vessel lumen. In some cases, inflammation may directly extend from the aorta to affect the coronary artery ostia and proximal vessels. Involvement can also be multifocal, resulting in mural thickening along multiple discrete segments, also called skip lesions. Stenosis or occlusion can be from the inflammation itself or from accelerated atherosclerosis secondary to the underlying vasculitis (Fig 1) (5). Patient presentation can thus be variable and is not always related to active uncontrolled coronary artery inflammation (6).

Encasement

While there are histopathologic similarities between some causes of CAV and encasement, the authors suggest that encasement refers to secondary involvement of the coronary arteries from benign and malignant neoplasms. This is a heterogeneous group of disorders that can have an array of multiorgan findings. Coronary artery encasement can be favored when there is masslike periarterial thickening that is eccentric relative to the vessel lumen. These lesions can have an irregular or nodular soft-tissue core with edematous epicardial fat stranding that can appear more exuberant compared with CAV. Again, skip lesions can occur with possible focal or multifocal stenosis. Additional multiorgan findings are the most suggestive of this diagnosis (Figs 2, S1). CCTA appearances of encasement are also nonspecific and can be indistinguishable from suspected CAV, such as when immunoglobulin G4 (IgG4) encasement causes circumferential smooth soft-tissue encasement along the proximal coronary arteries (Fig 3) (7–10).

Workup and Additional Imaging

Most often, imaging features of CAV and encasement are nonspecific and additional workup is required. These cases demand careful attention to clinical presentation and a multisystem review. Patients can have chronic nonspecific symptoms that go unrecognized or misdiagnosed. A comprehensive history should be obtained, including constitutional symptoms as well as single and multiorgan dysfunction. At physical examination, clinicians must be attuned to possible mucocutaneous manifestations, sensory and motor neuropathy, and circulatory issues. Radiologists should first review the patient's prior imaging examinations as isolated involvement of the coronary arteries is considered extremely rare (2). A helpful hint for vasculitis is the involvement of other variably sized vessels as well as other extravascular findings such as cavitary pulmonary lesions or renal disease. Likewise, given the heterogeneous causes of coronary artery encasement, unexpected findings in almost any organ can help suggest the diagnosis.

Next diagnostic steps may include blood work, biopsy, and/or additional imaging. Inflammatory marker levels (erythrocyte sedimentation rate and C-reactive protein), complete

Table 1: Estimated Prevalence of Some Causes of CAV and Encasement

Disease	Estimated Prevalence	Frequency of Coronary Artery Involvement
Takayasu arteritis	1–2 per 1 000 000 individuals	10%–45% of patients
GCA	10–30 per 100 000 individuals	Rare
Kawasaki disease	9–20 per 100 000 individuals	25%–30% of patients
ANCA-associated vasculitis	1–3 per 100 000 individuals	Rare
IgG4-related disease	0.3–1.0 per 100 000 individuals	1%–3% of patients
Erdheim-Chester disease	Rare	25%–55% of patients

Note.—ANCA = antineutrophil cytoplasmic antibody, GCA = giant cell arteritis, IgG4 = immunoglobulin G4.

Table 2: Classification of CAV versus Encasement

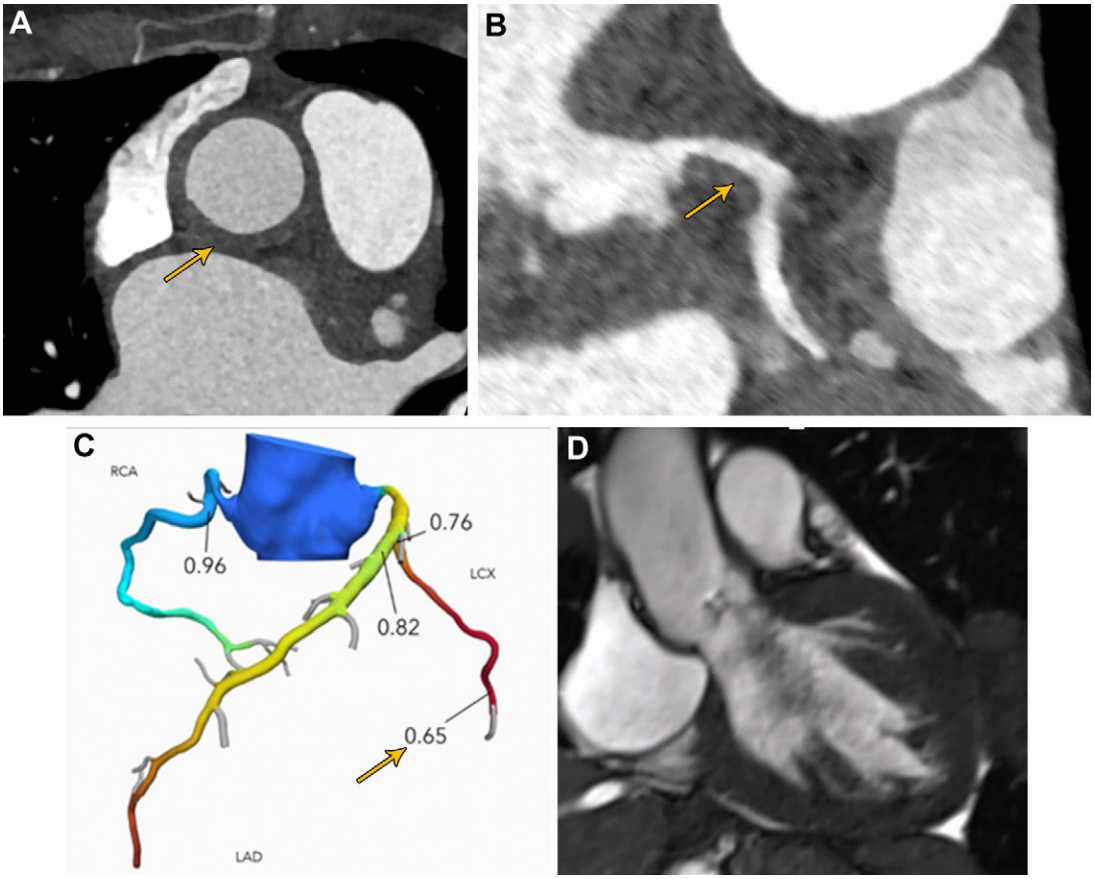
Type	Coronary Imaging Features	Systemic Findings
Vasculitis		
Takayasu arteritis	Osteal stenosis, smooth circumferential wall thickening ± skip lesions, aneurysm formation	Smooth wall thickening of the aorta, proximal aortic branches, and pulmonary artery
GCA	Like those of Takayasu arteritis but less frequent coronary artery involvement	Involvement of the supra-aortic vessels, particularly the temporal artery
Kawasaki disease	Acute: panarteritis ± skip lesions Subacute and chronic: multiple coronary artery aneurysms and stenosis	Fever, conjunctivitis, rash, strawberry tongue, extremity swelling, unilateral cervical lymphadenopathy
ANCA-associated vasculitis	Periarterial soft-tissue thickening that may be nodular, possible aneurysm	Multiple small subendocardial infarcts, asthma, renal disease, sinonasal disease
HIV vasculitis	Focal or diffuse stenosis, mural thickening and irregularity, aneurysmal dilatation	History of HIV infection, dilated cardiomyopathy, pulmonary hypertension
Encasement		
IgG4-related disease	Periarterial thickening ± pseudotumor, arterial stenosis, aneurysm	Autoimmune pancreatitis, sclerosing cholangitis, thyroiditis, Küttner tumor, lymphadenopathy
Histiocytic disorders	Perivascular infiltration and encasement	Perinephric fibrosis, metadiaphyseal sclerosis, painless cervical lymphadenopathy
Amyloid	Soft-tissue mass (amyloidoma) encasing adjacent coronary artery with limited stenosis	Pleural effusions, pericardial effusion, soft-tissue masses with calcification, carpal tunnel syndrome
Sarcoid	Hazy soft-tissue attenuation around vessels without stenosis	Hilar lymphadenopathy, upper lobe perilymphatic nodules ± fibrosis, erythema nodosum
Angiosarcoma	Long-segment stenosis of RCA with displacement, feeding arteries may be present	Metastatic disease often present at diagnosis
Lymphoma	Infiltrative mass encasing the coronary arteries without stenosis	Systemic lymphadenopathy, history of HIV or Epstein-Barr infection
Metastases	Extrinsic encasement with stenosis or infiltration and occlusion	Known primary malignancy, particularly melanoma and pleural mesothelioma

Note.—RCA = right coronary artery.

blood count, renal function tests, liver function tests, indices of collagen disorder (eg, lupus erythematosus cell and antinuclear antibody levels), serum IgG4 levels, blood cultures, and viral serology tests can potentially be diagnostic depending on the pathologic condition (7). Electrocardiography can also be helpful in determining if there is myocyte dysfunction.

Diagnostic imaging modalities can be considered based on the clinical scenario (Table 3). The high temporal and spatial resolution of modern CCTA provides evaluation of luminal stenosis, vessel walls, and the extent of disease activity, thus offering a fast, available, and comprehensive modality for assessing CAV and encasement (5,9). The epicardial fat can become edematous in both CAV and encasement to variable degrees, which can limit distinct characterization of the vessel wall. Accordingly, at CCTA these cases necessitate that cardiac imagers dynamically window images for full interpretation. A delayed phase image acquisition may also help to better characterize the epicardial soft tissues. Retrospective gating

Figure 1. Large-vessel vasculitis (presumed to be Takayasu arteritis) in a 54-year-old woman with a complex constellation of findings (cavitary pulmonary nodules, progressive aortic regurgitation, ileitis, and upper extremity arterial occlusion). **(A, B)** Double-oblique CCTA images at the sinotubular junction **(A)** and along the left main coronary artery and left circumflex artery *(LCX)* origin **(B)** show diffuse mural thickening (arrow) of the thoracic aorta extending to the left main artery and LCX origin, causing moderate stenosis. **(C)** Fractional flow reserve image (Heartflow; Heartflow) derived from CT shows a high likelihood of significant flow limitation in the dominant first obtuse marginal branch (arrow). *LAD* = left anterior descending artery, *RCA* = right coronary artery. **(D)** MR steady-state free-precession image along the left ventricular outflow tract shows moderate aortic valve regurgitation (calculated 40% regurgitant fraction).



can add qualitative or quantitative cardiac functional assessment. This type of gating is also useful when the patient has tachycardia with a low systolic blood pressure, which can contraindicate the use of β-blockers.

Given the severity of these pathologic conditions and the utility of accurate diagnosis, retrospective gating may also be considered, including in pediatric patients. In younger patients, low kilovolts and limited z-axis coverage can be used with premonitoring; images can be reconstructed at 0.5 mm with 0.2-mm increments and a sharper kernel, as well as maximum iterative reconstruction to optimize disease extent characterization. Cardiac MRI and ^{18}F-FDG PET can both be used to detect disease activity, with the latter being a useful modality for monitoring response to therapy (5,9,11,12).

Coronary Artery Vasculitis

Takayasu Arteritis

Takayasu arteritis is a predominantly large-vessel vasculitis characterized by inflammation of the aortic arch, proximal aortic branches, and pulmonary arteries. Young and middle-aged women are most commonly affected (6,7). Symptoms are nonspecific and depend on the territory of vascular involvement. Fever, chest discomfort, limb claudication, and headache are common. Coronary artery involvement is estimated in the range of 10%–30% but has been reported in up to 60% of patients at conventional coronary angiography (2). Other cardiac manifestations of Takayasu arteritis include aortic root or valvular involvement (with regurgitation being more common than stenosis) and heart failure, potentially related to systemic hypertension, left ventricular failure, or pulmonary arteritis and associated hypertension (13).

Takayasu arteritis can be favored when there is a suggestive pattern of large-vessel vasculitis with smooth circumferential wall thickening along the coronary arteries, especially when aortitis causes stenosis of the coronary ostia and proximal vessels (Fig 4). During the acute inflammatory phase of the disease, vessels may demonstrate the double-ring sign on CT images, which represents a hypoenhancing edematous intimal layer with inflammatory thickening and hyperenhancement of the medial and adventitial layers (14). In the chronic phase, the presence of stenoses and aneurysm formation interspersed

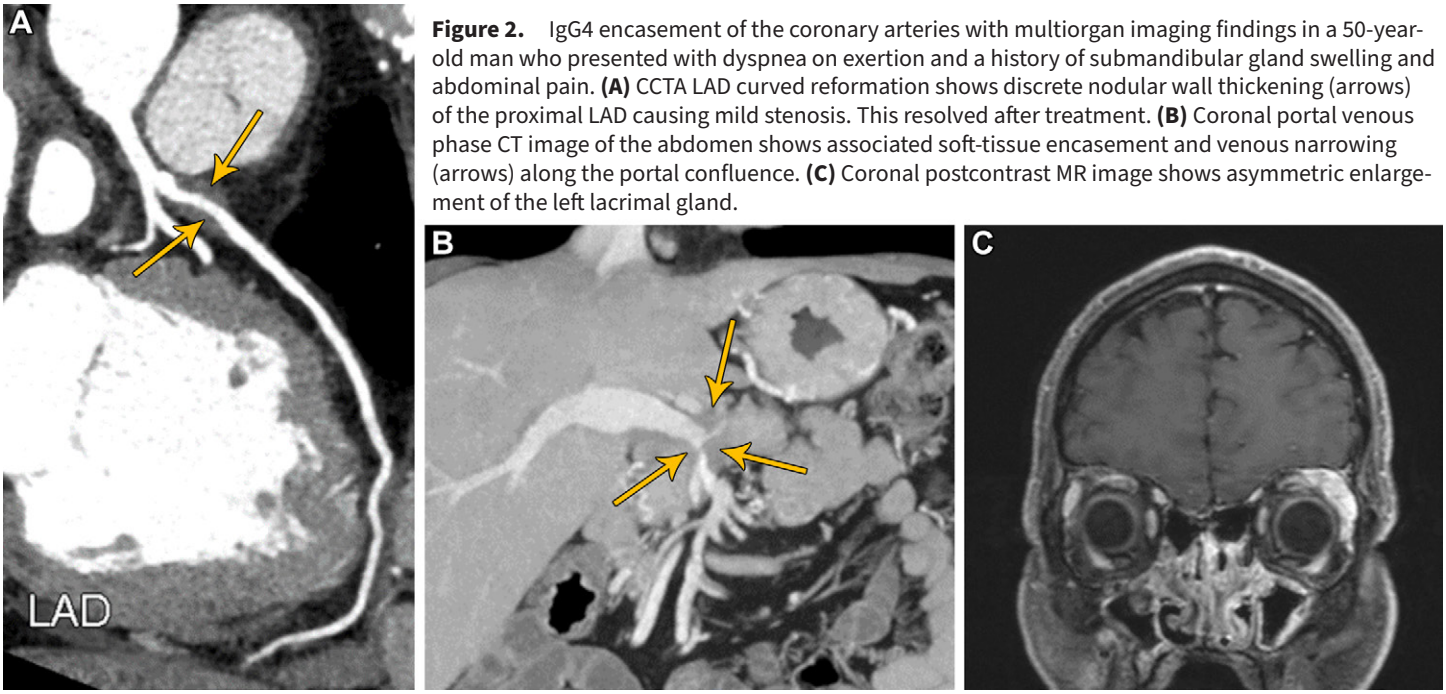


Figure 2. IgG4 encasement of the coronary arteries with multiorgan imaging findings in a 50-year-old man who presented with dyspnea on exertion and a history of submandibular gland swelling and abdominal pain. **(A)** CCTA LAD curved reformation shows discrete nodular wall thickening (arrows) of the proximal LAD causing mild stenosis. This resolved after treatment. **(B)** Coronal portal venous phase CT image of the abdomen shows associated soft-tissue encasement and venous narrowing (arrows) along the portal confluence. **(C)** Coronal postcontrast MR image shows asymmetric enlargement of the left lacrimal gland.

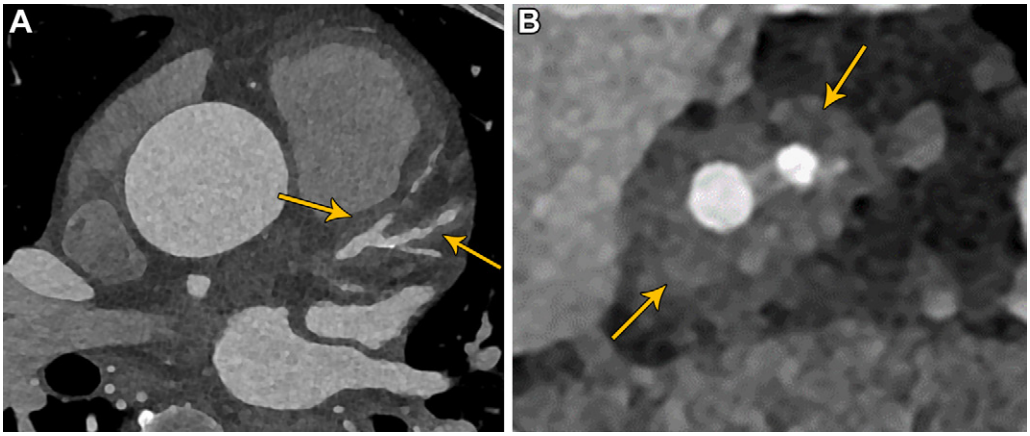


Figure 3. IgG4 encasement of the coronary arteries in a 54-year-old man with a 6-month history of severe daily diarrhea, profound weight loss, and significant fatigue. Axial **(A)** and double-oblique **(B)** photon-counting CT images show circumferential soft-tissue thickening (arrows) along the proximal/mid LAD and proximal first diagonal branch.

with normal segments (skip lesions) is known as the string of pearls sign. Involvement of the aortic arch and its branching vessels can lead to cerebrovascular symptoms (1).

Angiographic findings of coronary artery involvement tend to fall into three patterns, which include stenosis or occlusion of the coronary ostia (type 1), diffuse or focal coronary arteritis that may involve epicardial branches with or without skip lesions (type 2), and coronary artery aneurysms (CAAs)(type 3) (6,1). Type 1 is most common and is seen in over 70% of cases of Takayasu arteritis with coronary artery involvement (4). Involvement of the aortic root may also result in valvular dysfunction (Fig 1).

Imaging can guide treatment planning as invasive coronary angiography and CCTA can grade vasculitis-induced stenosis. CCTA, MR angiography, and ^{18}F-FDG PET are used in diagnosis and monitoring of large-vessel Takayasu arteritis because inflammatory biochemical markers are nonspecific and can even be normal during active disease (11). There is no established algorithm for imaging follow-up of these patients, and monitoring is decided on an individual basis (15).

Giant Cell Arteritis

Giant cell arteritis (GCA) is histopathologically indistinguishable from Takayasu arteritis, sharing many of the same imaging features. Both exhibit a predilection for large- to medium-sized vessels (7). However, GCA more frequently involves the supra-aortic vessels, resulting in headache, jaw claudication, and vision changes, particularly when the external carotid artery and its branches are involved (2,5). Additionally, GCA rarely involves the coronary arteries (2). In equivocal cases, GCA is favored over Takayasu arteritis in patients greater than 50 years of age.

Coronary artery involvement can manifest as inflammation along the proximal vessels. The pattern of involvement of the great vessels is again most helpful for this diagnosis as GCA, similar to Takayasu arteritis, also causes coronary artery ostial stenosis, smooth wall thickening along the proximal vessels, and/or skip lesions (Fig 5). Other cardiac manifestations of GCA include myocarditis (most often ischemic) and pericarditis (16).

Table 3: Comparison of Imaging Modalities for Coronary Vasculitis

Modality	Vessel Wall Imaging*	Stenosis Measurement*	Disease Activity*	Advantages	Disadvantages
TEE	+	+	+	Fast, accessible	Limited visualization of mid and distal vessels and potential for artifacts
CCTA	+++	+++	+++	High temporal and spatial resolution for comprehensive anatomic and functional assessment	Radiation
Cardiac MRA	+	+	++++	High sensitivity for disease activity and ischemia	Long acquisition time and low spatial resolution
ICA	NA	++++	NA	Therapeutic benefits (stent, angioplasty)	Unable to assess vessel wall and epicardial disease
IVUS	++++	++	+++	Excellent characterization of vessel wall	Limited assessment of small and stenosed vessels
FDG PET	NA	NA	++++	Detects sites of disease activity and monitors response to therapy	Low spatial resolution

* Number of + signs indicates the level of modality efficacy.
Note.—ICA = invasive catheter angiography, IVUS = intravascular US, MRA = MR angiography, NA = not applicable, TEE = transesophageal echocardiography.

Kawasaki Disease

Kawasaki disease is a systemic inflammatory disorder characterized by widespread inflammation of small- and medium-sized vessels. While its exact cause remains elusive, it is one of the most common causes of acquired heart disease in the developed world, occurring in children under 5 years of age with an estimated annual incidence of approximately 25 per 100 000 children in this age group (5). Hallmark clinical features of Kawasaki disease include prolonged fever, conjunctivitis, polymorphous rash, oral mucosal changes (strawberry tongue), extremity swelling, and unilateral cervical lymphadenopathy. CAV is present in up to 30% of cases and can lead to myocardial ischemia and sudden death (17,18).

The acute phase of disease is typically self limiting and may appear as diffuse inflammation of the coronary arteries or multifocal segmental stricturing (6). CAAs develop in the subacute phase of the disease and can chronically remodel and thrombose, leading to acute coronary syndromes. The presence of multiple CAAs, particularly if "giant" (defined as >4 times the normal diameter), suggest a diagnosis of Kawasaki disease (19).

If left untreated, CAAs can develop in 15%–25% of patients (5). Giant CAAs and thrombosis complications may still occur in up to 5% of patients with treated Kawasaki disease (Fig 6) (5,17). It should be noted that multisystem inflammatory syndrome in children (MIS-C), an emerging inflammatory disorder associated with prior SARS-CoV-2 infection, mimics the clinical presentation of Kawasaki disease and can cause similar cardiac findings (20).

Antineutrophil Cytoplasmic Antibody–associated Vasculitis

Antineutrophil cytoplasmic antibody (ANCA)–associated vasculitis refers to a family of autoimmune diseases characterized by small- to medium-vessel vasculitis and positive ANCA titers. The three main diseases are granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis (EGPA), and microscopic polyangiitis. Asthma, renal disease, and sinonasal disease are highly associated with ANCA-associated vasculitides. Eosinophilic granulomatosis with polyangiitis carries the highest prevalence of coronary artery involvement and is most common in middle-aged women (7). CAV is reported in up to 3% of patients with EGPA, although cardiac involvement in general is present in up to 60% and is the leading cause of mortality in these patients (4,6).

The coronary arteries may demonstrate periarterial soft-tissue thickening with stranding, which can be somewhat masslike and mimic coronary artery encasement (Fig 7). This contrasts with other vasculitic processes, which tend to produce smooth concentric vessel wall thickening. Aneurysm formation is also possible (6). A characteristic feature of EGPA at MRI is multiple small subendocardial infarcts (Fig 8). Associated cardiac findings also include pericarditis, myocarditis, valvular abnormalities, and restrictive or dilated cardiomyopathy (5,7).

HIV Vasculitis

CAV in the context of HIV infection is a rare but serious complication that can lead to accelerated atherosclerosis and an increased risk of cardiovascular events. The prevalence of coronary atherosclerosis in men who are HIV positive is 59%, compared with 34% in non–HIV-infected individuals (21). The presence of cytokines and other inflammatory biomarkers is thought to facilitate a complex interplay between inflammatory cells and endothelial cells to promote atherosclerosis (22). The SMART (Strategies for Management of Anti-Retroviral Therapy) study group demonstrated that in patients who are HIV positive, higher levels of interleukin-6, high-sensitivity C-reactive protein, and D dimer are associated with various cardiovascular diseases including myocardial infarction and coronary artery disease (23). Even when adjusted for

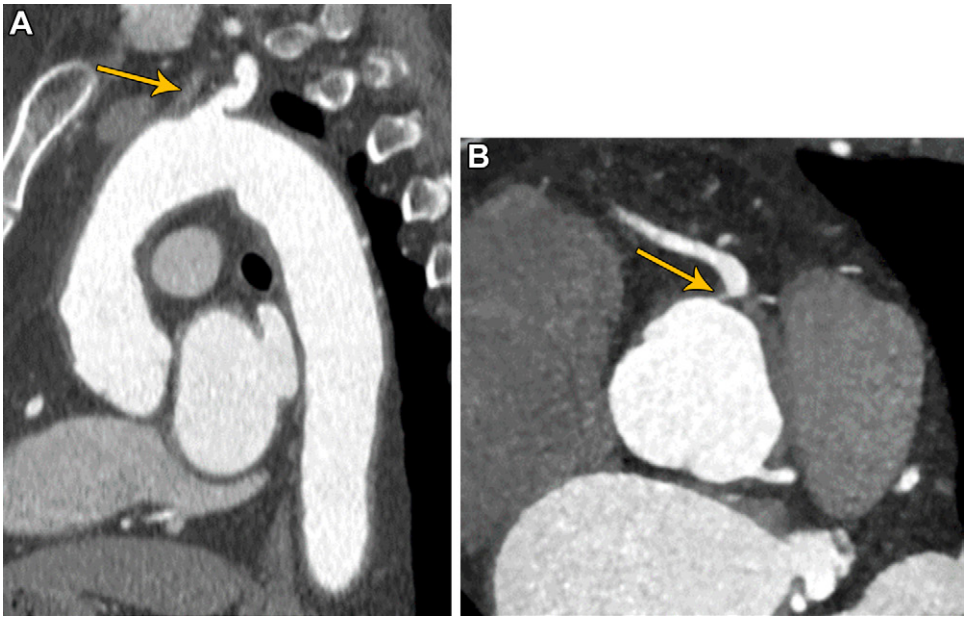


Figure 4. Takayasu arteritis in a 49-year-old man with a history of chronic chest pain. **(A)** Candy cane thoracic CT angiogram shows a typical pattern of large-vessel vasculitis with circumferential smooth wall thickening along the thoracic aorta and occlusion (arrow) of the left common carotid artery. **(B)** Double-oblique CCTA image at the coronary artery origins shows aortitis with severe stenosis (arrow) of the RCA ostium and minimal left main coronary artery stenosis.

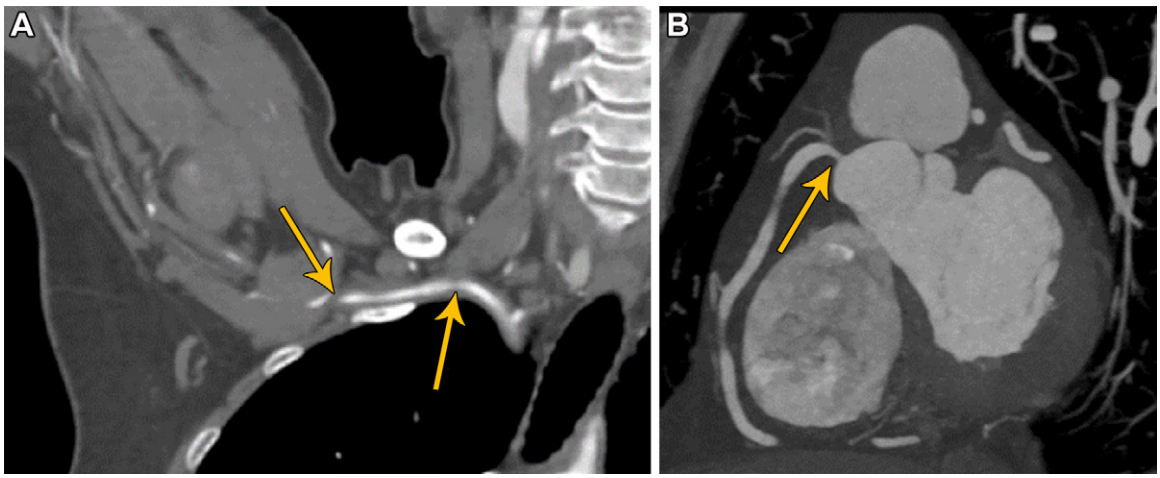


Figure 5. GCA in a 65-year-old woman with right hand numbness and upper extremity claudication. **(A)** Double- oblique right upper extremity CT angiogram shows circumferential wall thickening along the right subclavian and axillary arteries (arrows) with downstream occlusion. **(B)** Short-axis coronary CT angiogram shows moderate stenosis (arrow) of the RCA origin due to large-vessel vasculitis.

traditional cardiovascular risk factors, patients who are HIV positive are at higher risk of developing more severe forms of coronary artery disease when compared with individuals who are not HIV infected (24). Additionally, patients who are HIV positive still seem to develop subclinical atherosclerosis whether on or off active antiretroviral therapy (21).

Common to other vasculitic processes, the coronary arteries may demonstrate circumferential mural thickening with variable degrees of stenosis and skip lesions. However, noncalcified and high-risk low-attenuation plaques are seen more frequently in patients with HIV (21). Thus, it can be challenging to differentiate regions of active vascular inflammation from areas of atherosclerosis, as both can cause stenosis, mural thickening and irregularity, and aneurysmal dilatation. In such situations, ^{18}F-FDG PET can be helpful to identify areas of active vascular inflammation (Fig 9). Other cardiovascular manifestations include dilated cardiomyopathy, pericardial disease, and pulmonary artery hypertension.

Coronary Artery Encasement

IgG4-related Disease

IgG4-related disease is a multiorgan disorder characterized by tissue infiltration of IgG4-positive plasma cells with subsequent fibrosis (25). It is more common in men and patients over 60 years of age. Systemic manifestations include autoimmune pancreatitis, sclerosing cholangitis, thyroiditis, chronic sclerosing sialadenitis (Küttner tumor), and lymphadenopathy. Indeed, given the wide variety of manifestations, organ-specific diagnostic criteria are used with integration of clinical symptoms, radiographic findings, and serologic and pathologic abnormalities (26). Typically, large vessels are involved, with abdominal aortic aneurysms being one of the

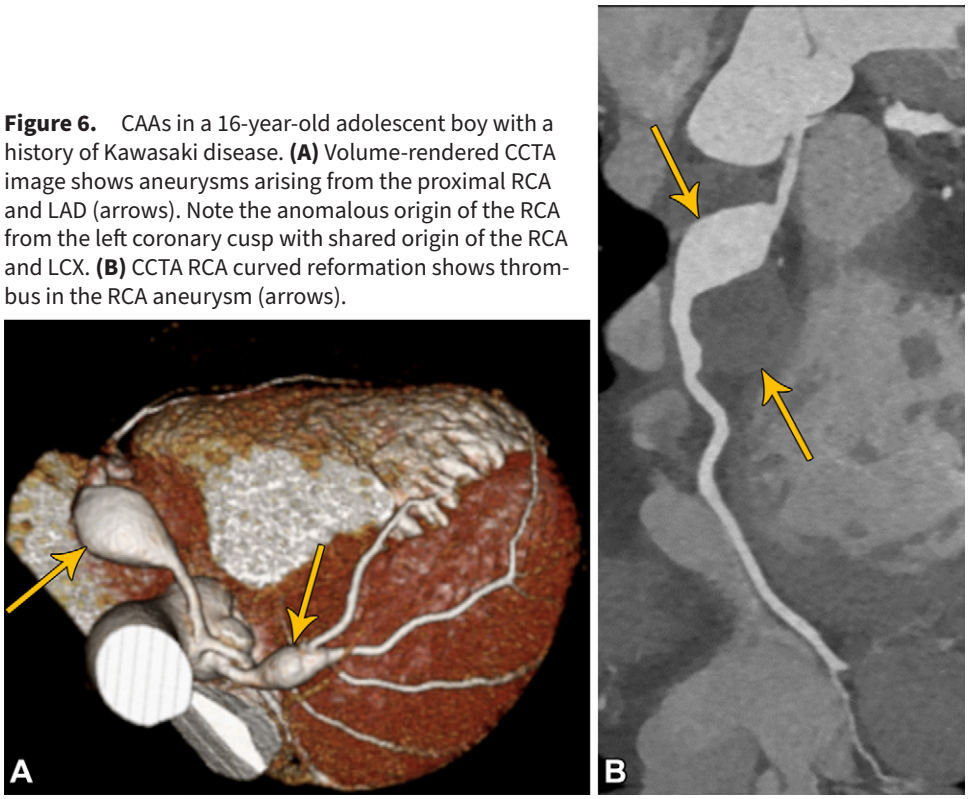


Figure 6. CAAs in a 16-year-old adolescent boy with a history of Kawasaki disease. **(A)** Volume-rendered CCTA image shows aneurysms arising from the proximal RCA and LAD (arrows). Note the anomalous origin of the RCA from the left coronary cusp with shared origin of the RCA and LCX. **(B)** CCTA RCA curved reformation shows thrombus in the RCA aneurysm (arrows).

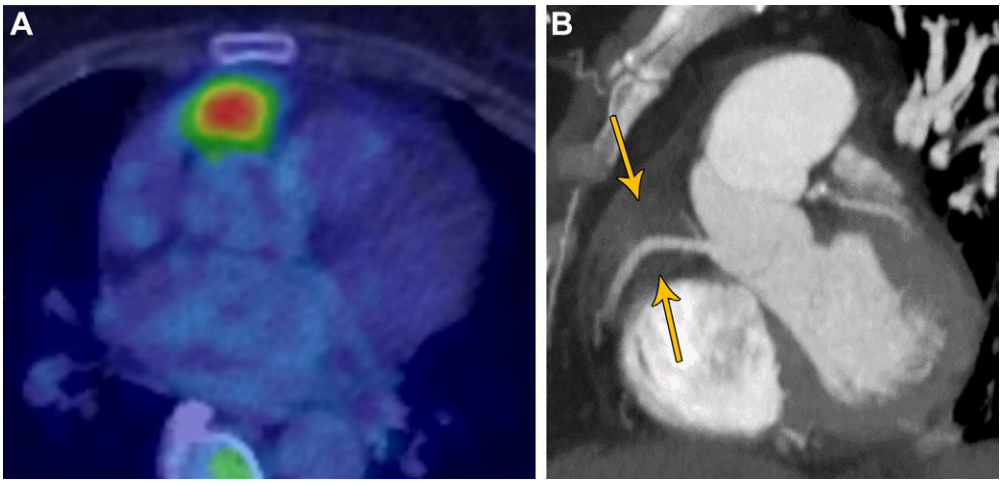


Figure 7. ANCA-associated vasculitis in a 66-year-old woman with multiple FDG-avid lesions in the lungs, kidneys, spleen, and pericardium. **(A)** Axial FDG PET/CT image shows an FDG-avid soft-tissue lesion within the pericardium in the region of the proximal RCA. **(B)** Double-oblique ungated high-pitch CT angiogram along the proximal RCA shows the underlying nodular soft-tissue thickening (arrows), which is eccentric to and encases the proximal and mid vessel, causing no underlying stenosis. This patient had elevated cytoplasmic ANCA levels, and subsequent renal biopsy confirmed granulomatosis with polyangiitis.

most common findings (27). Coronary artery involvement is relatively uncommon, with a reported incidence of 1%–5% (5,27). Of note, coronary artery involvement, when present, is almost never seen in isolation (27).

Imaging findings that can be seen with coronary artery involvement of IgG4 include periarterial thickening, arterial stenosis, and aneurysm, which can ultimately lead to thrombosis and myocardial ischemia (9). Periarterial thickening can be more exuberant when compared with vasculitis, although this is not necessarily a distinguishing feature. In some cases, the coronary arteries may simply demonstrate circumferential mural thickening that can be easily confused with vasculitides or soft atherosclerotic plaque (Fig 3).

More advanced cases may demonstrate perivascular inflammatory masses, or pseudotumor formation, which represent masslike fibrosclerotic thickening of the arterial wall (10). This can demonstrate the "pigs-in-a-blanket" sign on CT images, a specific finding of IgG4-related coronary arteritis referring to the appearance of soft-tissue masses encasing a coronary artery in the context of multiple coronary pseudotumors. These will exhibit increased uptake at ^{18}F-FDG PET and increased late gadolinium enhancement (LGE) at MRI when active periarterial inflammation is present (12,28). Both ^{18}F-FDG PET and MRI can be used to assess the degree of active inflammation following treatment. Persistent FDG uptake after treatment may indicate increased risk of future relapse (12).

Non-Langerhans Cell Histiocytosis

Non-Langerhans cell histiocytosis encompasses a group of rare diseases characterized by tissue accumulation of

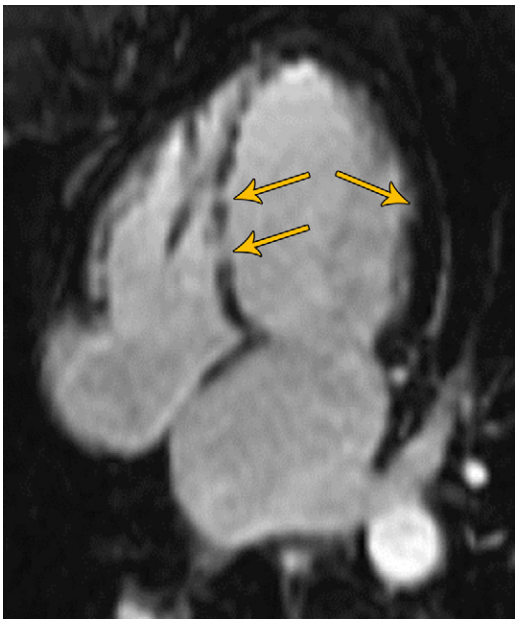


Figure 8. Eosinophilic granulomatosis with polyangiitis in a 56-year-old woman. Four-chamber late gadolinum-enhanced MR image shows multiple subendocardial and transmural infarctions (arrows).

CD68-positive and CD1a-negative histiocytes, a type of immune cell derived from macrophages (29,30). Several subtypes exist, the two most well-known being Erdheim-Chester disease and Rosai-Dorfman disease (31).

Erdheim-Chester disease is a multisystem disorder in patients of middle age and has a slight male predominance. It frequently affects large- and medium-sized vessels (up to 82% of cases involve the aorta), with coronary artery involvement in up to 55% of cases (32,33). Myocardial infiltration and thickening at the atrioventricular groove is present in over one-third of patients, which can lead to masslike encasement and stenosis of the coronary arteries, particularly the right coronary artery (RCA) (Fig 10) (7,34). At catheter angiography, isolated smooth concentric stenosis of the RCA should prompt consideration of this disease entity, the main differential diagnosis being angiosarcoma, which also has a predilection for encasing the RCA. Other classic findings of Erdheim-Chester disease include perinephric fibrosis demonstrating the characteristic hairy kidney sign at cross-sectional imaging and bilateral symmetric metadiaphyseal sclerosis (7,33).

On the other hand, Rosai-Dorfman disease is more common in children and young adults and classically presents with painless cervical lymphadenopathy (29,35). Cardiac involvement is exceedingly rare, with an estimated incidence of 0.1%–0.2% of cases (35). A case report by Heidarian et al (36) describes a left atrial mass extending into the interatrial septum and anterior leaflet of the mitral valve. Soft-tissue encasement of the coronary arteries is also possible (Fig 11).

Amyloidosis

A known manifestation of amyloidosis is the development of amyloidomas, soft-tissue masses representing collections of amyloid protein that can be found in various organ systems. Cardiac amyloidomas while rare may occur in the absence of classic myocardial infiltration and can have varied involvement of adjacent structures.

A case report by Iyer et al (37) demonstrates an isolated epicardial amyloidoma in the right atrioventricular groove without involvement of other structures, while another by Cicco et al (38) depicts a similarly positioned epicardial amyloidoma causing right ventricular compression with associated pericardial effusion. Neither case had myocardial involvement. If an amyloidoma occurs in close enough proximity to a coronary artery, then encasement can occur (Fig 12). This may be occult at catheter angiography when there is no resultant stenosis, although displacement of structures may provide a subtle clue suggesting an underlying mass. At CCTA, the appearance of an isolated amyloidoma is nonspecific. The more common manifestation of cardiac amyloidosis is myocardial infiltration with associated heart failure from restrictive cardiomyopathy.

Sarcoidosis

Cardiac sarcoidosis is a rare but potentially serious manifestation of sarcoidosis, a multisystem inflammatory disorder characterized by the formation of noncaseating granulomas in various organs, most commonly the lungs and lymph nodes. Subclinical cardiac involvement is thought to be present in as many as 20%–30% of cases. However, clinically significant cardiac involvement is only seen in about 5% of cases (11,29).

Coronary artery encasement is occasionally present during the inflammatory phase of disease and appears as hazy soft-tissue attenuation surrounding the involved vessels (Fig 13). These changes tend to remain extrinsic to the vessel lumen, making stenosis less common when compared with other causes of encasement. Thus, catheter angiography is typically normal and only used to exclude underlying coronary artery stenosis in patients presenting with cardiac symptoms (40,41). The life-threatening complications of cardiac sarcoidosis such as heart failure, atrioventricular block, and ventricular tachyarrhythmias are largely due to chronic myocardial infiltration and scarring, which has variable appearances with LGE imaging at MRI, described elsewhere in the literature (11). Clinicians can also use ^{18}F-FDG PET in combination with single-photon emission CT perfusion imaging to categorize disease status into early-stage disease, progressive disease, or fibrous disease (41,41). Generally, both the degree of FDG uptake and perfusion anomalies progress with the disease until it reaches end-stage fibrosis, at which point there is minimal FDG uptake and severe perfusion defects (41). After treatment, the degree of FDG uptake may regress more substantially than the degree of LGE at cardiac MRI, making ^{18}F-FDG PET more useful for treatment monitoring (11).

Angiosarcoma

Cardiac angiosarcoma is a rare aggressive primary cardiac tumor originating from the vascular or lymphatic endothelium of the heart. Primary cardiac tumors are extremely rare

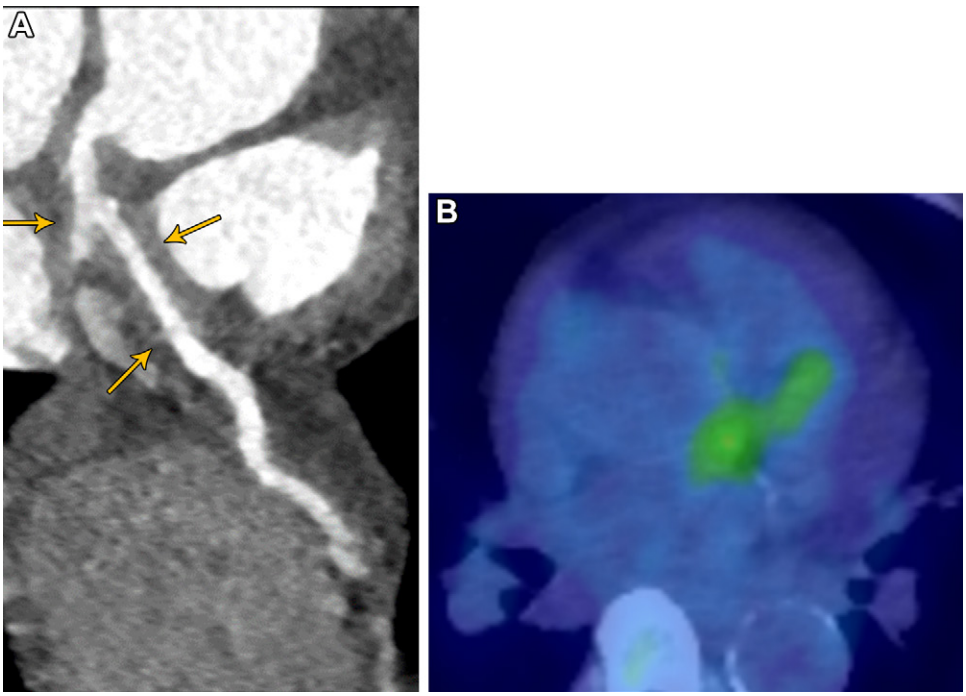


Figure 9. HIV vasculitis in a 49-year-old woman with a history of HIV infection who was on antiretroviral therapy and had an adequate CD4 count. The patient presented with chest pain and was found to have positive troponin measurement and electrocardiographic findings consistent with non-ST-segment elevation myocardial infarction (NSTEMI). **(A)** CCTA LAD curved reformation shows mural thickening (arrows) along the left main and proximal LAD with no luminal stenosis. **(B)** Axial PET/CT image shows moderate FDG avidity consistent with active vasculitis, most intense along the left main coronary artery and proximal LAD.

and carry an incidence of about 0.001%–0.03% at autopsy, with angiosarcoma being the most common type, accounting for about 25%–30% of these cases (42,43). It tends to occur in middle age and has a male predominance, and metastases are often present at the time of diagnosis (44,45). Initial diagnosis can be challenging due to the nonspecific clinical presentation, which depends on the size and location of the tumor within the heart and can include symptoms such as dyspnea, chest pain, palpitations, and syncope (42).

Cardiac angiosarcoma has a predilection for the right side of the heart, particularly the right atrium, and commonly encases the RCA and other surrounding structures (Fig 14) (45,46). It is unlikely to be mistaken for other vasculitic disorders at cross-sectional imaging owing to its characteristic heterogeneous and infiltrative appearance. At CCTA, for example, angiosarcoma is most likely to be confused with a pericardial condition or other cardiac tumors. However, conventional angiography may only demonstrate nonspecific narrowing of the coronary arteries that can mimic atherosclerosis or vasculitis. A long segment of narrowing involving a single vessel, particularly the RCA, should raise suspicion for an encasement syndrome. Certainly, many cases of cardiac angiosarcoma are relatively sizable at diagnosis. Displacement of the artery away from its expected course may also suggest an underlying mass. The most suspicious feature is the presence of feeding arteries arising from the encased coronary artery to the tumor (47). Characteristic features on MR images include a cauliflower-like appearance from internal hemorrhage and necrosis, as well as a sunray appearance when there is diffuse pericardial infiltration (48).

Lymphoma

Cardiac lymphoma rarely occurs as a primary malignancy but is a common manifestation of disseminated disease. Accounting for less than 2% of primary cardiac tumors, nearly all cardiac lymphomas are a diffuse large B-cell variant of non-Hodgkin lymphoma (49). Primary cardiac lymphomas usually occur in immunocompetent adults, and there is a subset of disease related to Epstein-Barr virus and HIV infection (50).

Imaging typically demonstrates an infiltrative mass with a predilection for the right atrium but can involve any part of the heart and encase the coronary arteries (Fig 15). Vascular stenosis is rare. Contrast enhancement is variable, ranging from homogeneous to heterogeneous, and there is no specific LGE pattern (50). The appearance on MR images can be variable, but masses are usually relatively hypointense on T1-weighted images and hyperintense on T2-weighted images (50,51). Because of its dense cellularity, lymphoma has characteristic restricted diffusion. Epicardial and pericardial infiltration manifest as pericardial thickening or effusion and are often an early feature of disease (51). ^{18}F-FDG PET is used for staging and monitoring systemic treatment response.

Metastasis

While still uncommon, cardiac metastases occur much more frequently than primary cardiac malignancies. In patients with known malignancies, cardiac metastases have been reported in up to 9.1% of patients at autopsy (52). Melanoma and pleural mesothelioma have a particular predilection for involving the heart, although most cases are from lung, breast, and hematologic primary malignancies due to their high prevalence in the general population (53).

The presence of encasement again depends on the proximity between the metastatic deposit and the coronary artery. Extrinsic encasement with stenosis or frank vascular infiltration by the tumor may occur (Fig 16). Metastases more often extend to encase the vessels than appear as infiltrative processes along the coronary arteries. In the specific case of carcinoid heart disease, the release of vasoactive neuroendocrine substances

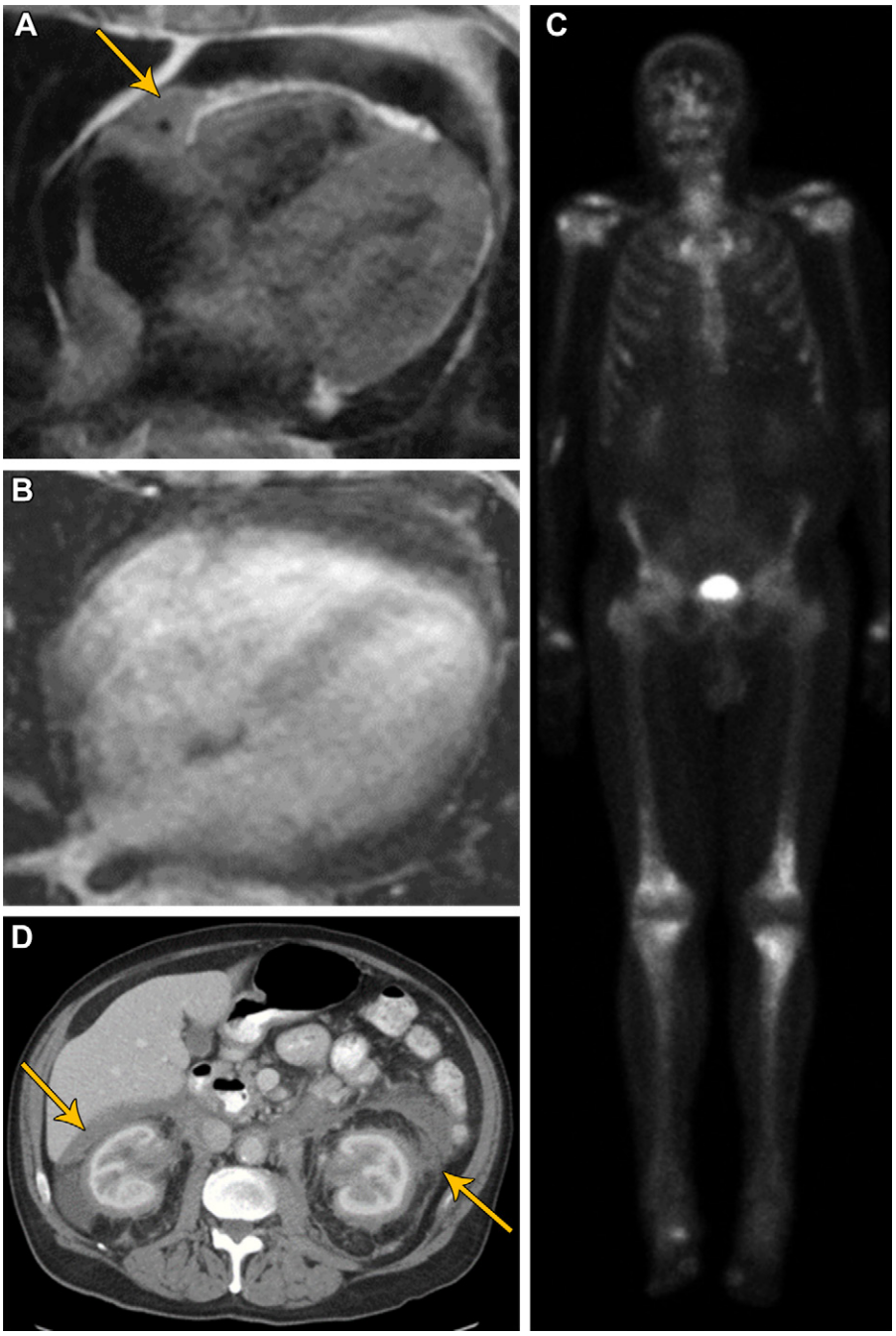


Figure 10. Encasement of the RCA in a 74-year-old man with Erdheim-Chester disease. **(A, B)** Four-chamber cardiac MR images show T2 intermediate signal intensity with a preserved RCA flow void (arrow in **A**) on the double inversion-recovery image **(A)**, with homogeneous enhancement on the delayed gadolinium-enhanced image **(B)**. **(C)** Methylene diphosphonate bone scan shows symmetric radiotracer uptake in the metadiaphyses of the bilateral femurs and tibias. **(D)** Axial contrast-enhanced CT image shows characteristic perirenal and retroperitoneal soft-tissue thickening (arrows).

can cause coronary vasospasm, mimicking primary vasculitis at conventional catheter-based coronary angiography (54). Similar to other causes of encasement, displacement of normal structures may provide a clue to the diagnosis. The pericardium is the most common site of cardiac metastasis and may manifest as pericardial thickening, pericardial effusion, or enhancing pericardial nodules (55). Intracardiac metastasis can occur anywhere in the heart but most commonly involves the right atrium (52).

MR images usually depict metastases as having low T1 signal intensity and high T2 signal intensity, with the exception of metastatic melanoma, which can demonstrate high intrinsic T1 signal intensity due to the paramagnetic effects of melanin (53). CT is better at depicting coronary artery involvement, which may appear as vascular encasement or infiltration by the tumor (Fig 16). The appearance at ^{18}F-FDG PET is variable due to the high degree of radiotracer uptake in normal myocardium. Although maximum standard uptake values tend to be higher in malignant cardiac tumors when compared with benign tumors, this has not been shown to reliably distinguish cardiac metastasis from primary cardiac malignancies (56).

Complications and Mimics

Potential mimics of CAV at imaging, namely atherosclerosis and CAA, are overall more common than the vasculitides themselves, as both represent separate pathologic conditions as well as common complications of vascular inflammation.

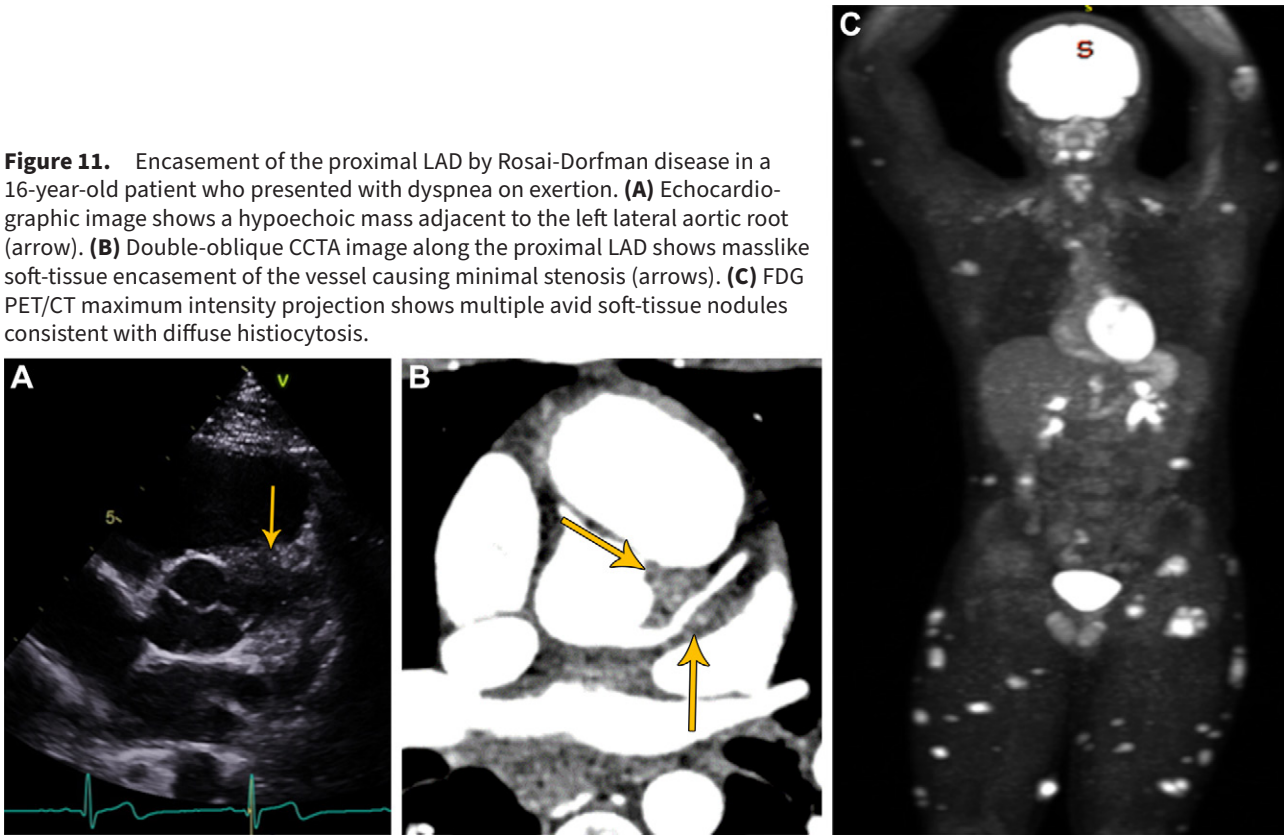


Figure 11. Encasement of the proximal LAD by Rosai-Dorfman disease in a 16-year-old patient who presented with dyspnea on exertion. **(A)** Echocardiographic image shows a hypoechoic mass adjacent to the left lateral aortic root (arrow). **(B)** Double-oblique CCTA image along the proximal LAD shows masslike soft-tissue encasement of the vessel causing minimal stenosis (arrows). **(C)** FDG PET/CT maximum intensity projection shows multiple avid soft-tissue nodules consistent with diffuse histiocytosis.

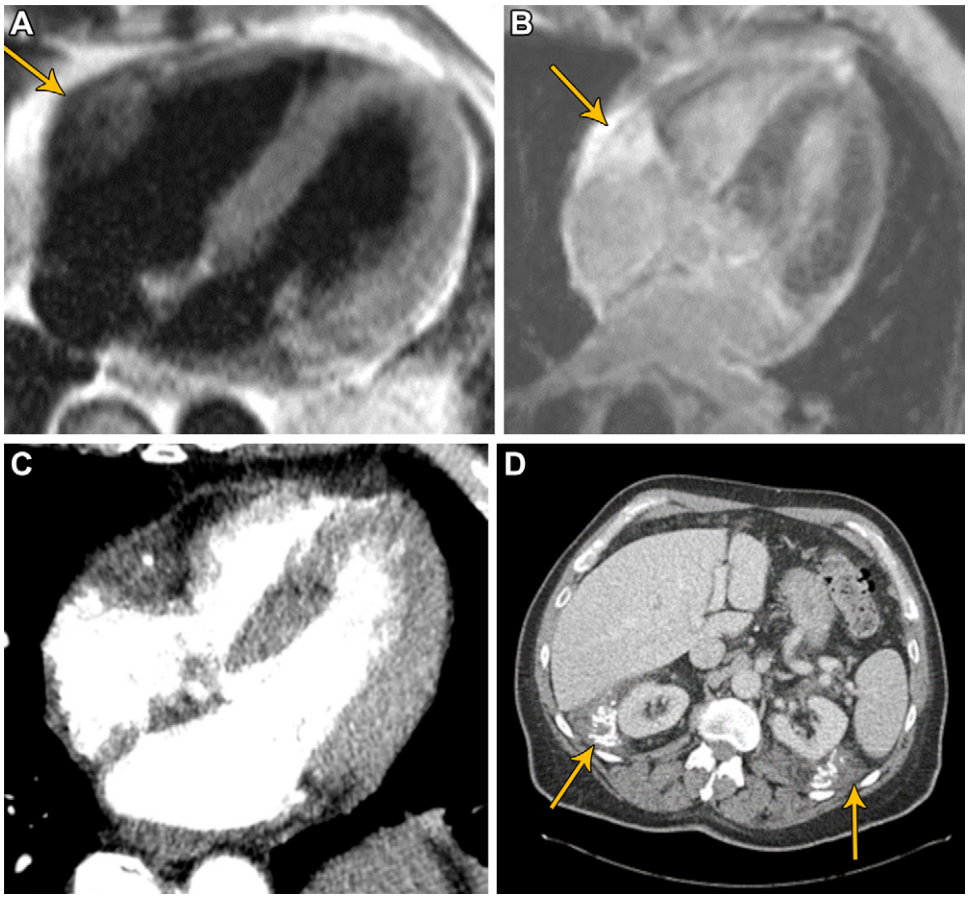
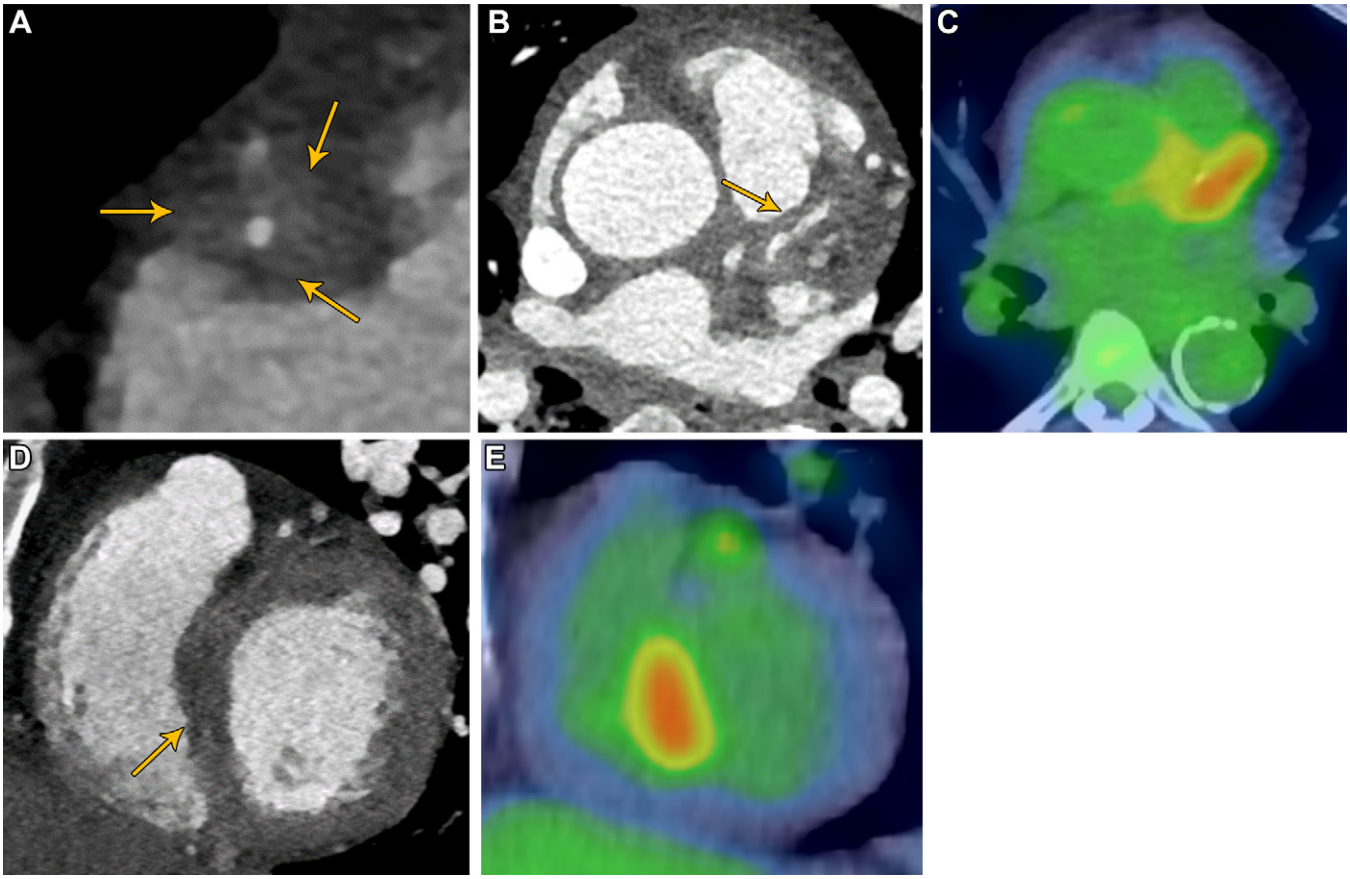


Figure 12. Amyloid encasing the RCA in a 77-year-old man with a history of monoclonal gammopathy of undetermined significance (MGUS). MRI was performed to exclude myocardial involvement. **(A, B)** Axial MR images show a soft-tissue mass (arrow) in the right atrioventricular groove encasing the mid RCA, with intermediate signal intensity on the T2-weighted half-Fourier single-shot turbo spin-echo (HASTE) image **(A)** and avid enhancement on the delayed gadolinium-enhanced image **(B)**. **(C)** Axial CT angiogram shows a soft-tissue mass encasing the mid RCA with no associated RCA stenosis. **(D)** Axial contrast-enhanced CT image of the abdomen shows calcified retroperitoneal soft tissue (arrows). Biopsy results were consistent with light chain amyloid.

Figure 13. Sarcoid encasement of the coronary arteries and myocardial infiltration in a 49-year-old woman with a history of sarcoidosis and HIV infection. **(A)** Axial CCTA image shows extensive hazy periarteritis along the mid RCA (arrows). **(B, C)** Axial CT **(B)** and PET **(C)** images show soft-tissue thickening and periarteritis along the LAD (arrow in **B**) with corresponding intense FDG avidity. **(D, E)** Short-axis CT **(D)** and PET **(E)** images show decreased perfusion along the mid septum (arrow in **D**) with corresponding intense FDG avidity. A mediastinal lymph node biopsy showed non-necrotizing granulomatous inflammation consistent with sarcoidosis. Given the associated myocardial infiltration and resolution with subsequent treatment, the coronary artery encasement was also caused by the presence of sarcoid.



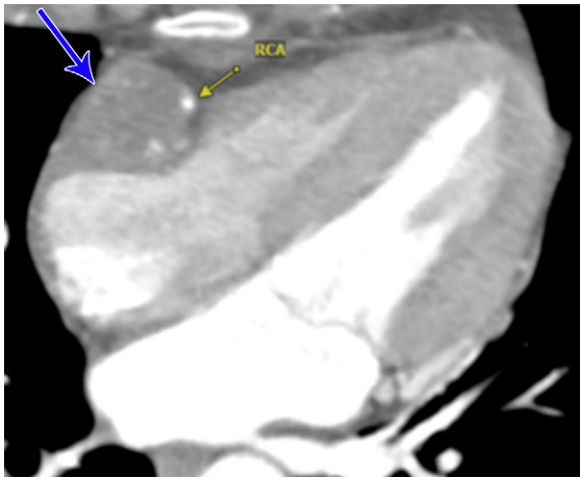


Figure 14. Angiosarcoma encasing the RCA in a 43-year-old woman who presented with shortness of breath. Four-chamber CCTA image shows a mildly heterogeneous soft-tissue mass (blue arrow) involving the right atrium with extension into the right atrioventricular groove and encasement of the RCA, which remains widely patent.

Acute plaque rupture and spontaneous coronary artery dissection (SCAD) are causes of acute coronary syndrome that are also in the differential diagnosis of CAV.

Atherosclerosis

While a separate disease entity from CAV, atherosclerosis is known to be accelerated in the setting of underlying vasculitic processes. Interferon γ, tumor necrosis factor α, and type 1 T helper (Th1) interleukin are inflammatory cytokines that are commonly overproduced in vasculitic disorders and thought to induce atherosclerotic inflammatory changes of the endothelium (1).

The presence of calcified plaques and relatively well-defined eccentric arterial wall thickening favor atherosclerosis over vasculitis, which would typically appear as smooth concentric thickening without calcification (Fig 17). Extraluminal soft-tissue attenuation and periarterial stranding are atypical of atherosclerosis and should prompt consideration of an underlying vasculitic or encasement disorder. Rarely, diffuse noncalcified plaque in the context of exuberant atherosclerosis can mimic the smooth concentric wall thickening seen in vasculitis (Fig 18). In such cases, correlation with clinical, laboratory, and other systemic findings of disease is necessary to exclude underlying inflammation. ^{18}F-FDG PET may help

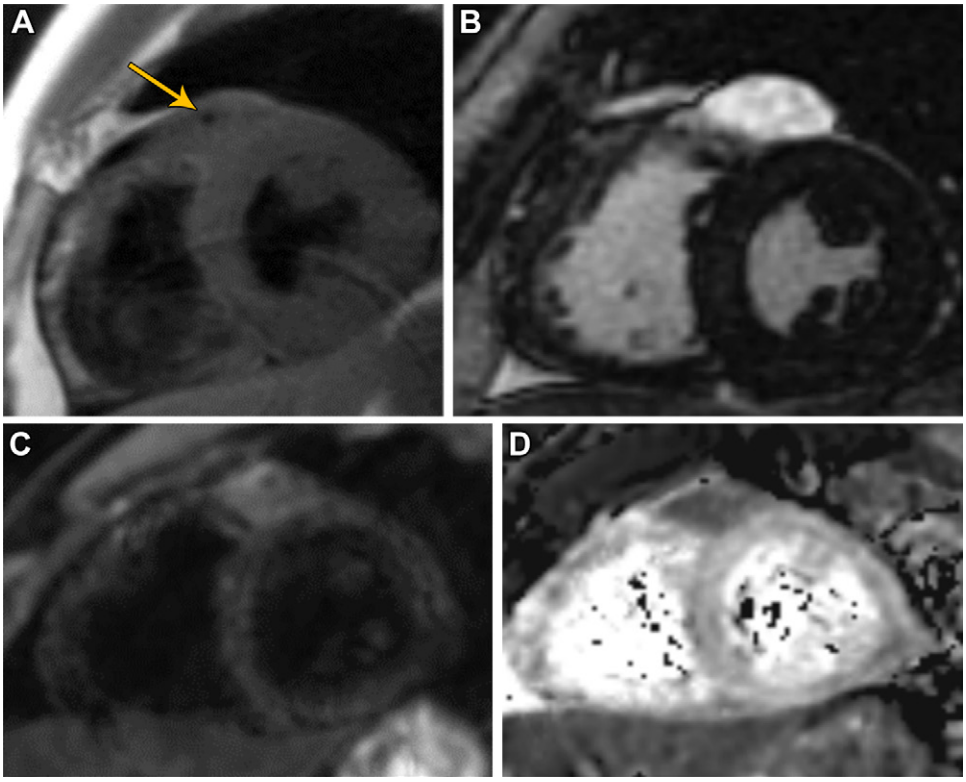


Figure 15. Lymphoma encasing the LAD in a 41-year-old man who presented with dyspnea at rest and on exertion. **(A)** Short-axis double inversion-recovery MR image shows a mass encasing the proximal LAD with intermediate T1-weighted signal intensity. Note the preserved LAD flow void (arrow). There was similar intermediate T2-weighted signal intensity at triple inversion imaging (not shown). **(B)** Delayed gadolinium-enhanced MR image shows the mass with intense homogeneous enhancement at delayed gadolinium imaging. **(C, D)** Short-axis diffusion-weighted (*b* value = 100 sec/mm^2) **(C)** and apparent diffusion coefficient **(D)** MR images show characteristic restricted diffusion. The imaging characteristics strongly suggest lymphoma, which was confirmed at biopsy.

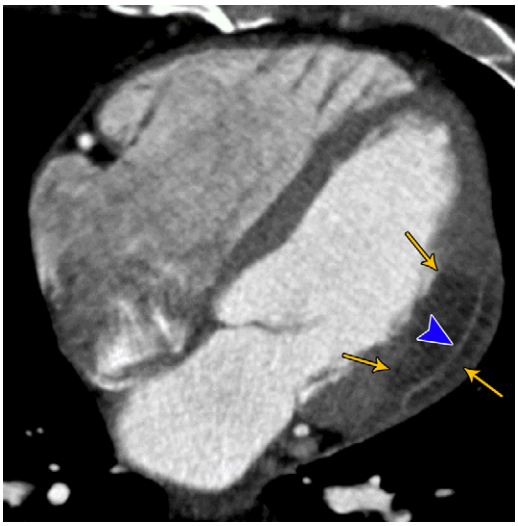


Figure 16. Metastasis encasing an obtuse marginal branch in a 55-year-old man with cutaneous melanoma. Double-oblique CCTA image along an obtuse marginal branch shows an infiltrative hypoattenuating mass in the left lateral wall of the left ventricle (arrows). This mass diffusely encases the vessel without occlusion or significant stenosis (arrowhead).

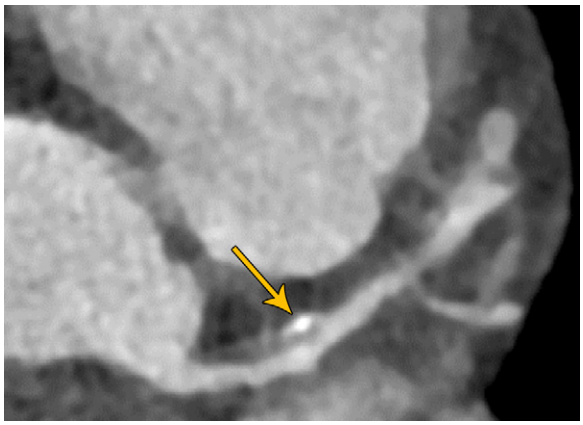


Figure 17. Atherosclerosis in a 35-year-old man who presented with syncope and abdominal pain. Double-oblique CCTA image shows segmental mixed plaque along the left main coronary artery that causes mild stenosis with focal severe stenosis along the mid LAD. Calcification (arrow) and eccentric well-defined wall thickening favor atherosclerosis over vasculitis. This is an extensive burden of plaque in a patient of this age. However, a review of the remainder of the CT images demonstrated muscle hypertrophy, gynecomastia, and gluteal injection sites (not shown), suggesting anabolic steroid use.

to differentiate areas of active inflammation from atherosclerosis, although on occasion noncalcified plaques undergoing remodeling can demonstrate FDG uptake as well (57).

Acute Plaque Rupture

Acute plaque rupture constitutes a pivotal juncture in the pathophysiologic trajectory of atherosclerosis, whereby destabilization of the protective fibrous cap precipitates an acute rupture event or thromboembolism (58). Noncalcified plaque is regarded as the initial precursor of calcified plaque (and thus an early marker of coronary artery atherosclerosis) and is present in high-risk plaques, although the significance of isolated noncalcified plaques in symptomatic patients remains to be seen (59,60). On images, thromboembolism may

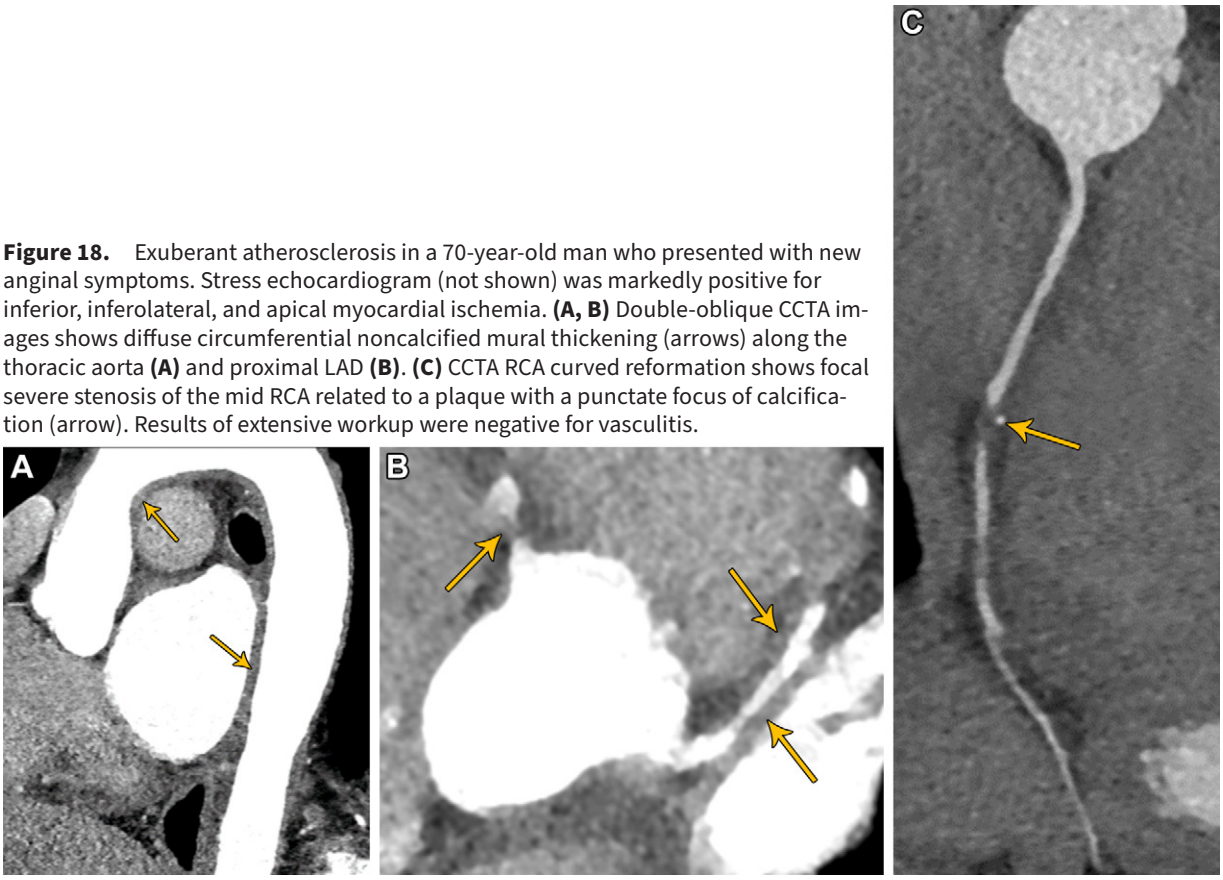


Figure 18. Exuberant atherosclerosis in a 70-year-old man who presented with new anginal symptoms. Stress echocardiogram (not shown) was markedly positive for inferior, inferolateral, and apical myocardial ischemia. **(A, B)** Double-oblique CCTA images shows diffuse circumferential noncalcified mural thickening (arrows) along the thoracic aorta **(A)** and proximal LAD **(B)**. **(C)** CCTA RCA curved reformation shows focal severe stenosis of the mid RCA related to a plaque with a punctate focus of calcification (arrow). Results of extensive workup were negative for vasculitis.

appear as abrupt total or near occlusion of a coronary artery with maintained vessel caliber immediately proximal and distal to the obstructing lesion (Fig 19). Periarteritis may occur in acute plaque rupture, although CAV is more likely to have periarteritis or ill-defined soft-tissue edema extending over a long segment. Diagnostic techniques, including both conventional and CT angiography, play a crucial role in identifying vulnerable plaques before they rupture and cause acute coronary syndromes. The updated Coronary Artery Disease Reporting and Data System (CAD-RADS) 2.0 reporting guidelines contains an additional high-risk plaque modifier to identify vulnerable plaques, which are defined as plaques demonstrating two or more of the following features: low attenuation (<30 HU), positive (outward) vessel remodeling, spotty calcification, and napkin-ring sign (60).

Coronary Artery Aneurysms

CAAs can be a common endpoint of atherosclerotic, vasculitic, and encasement disorders. They are defined as localized dilatation of a coronary artery by at least 1.5 times its normal diameter and are pathologically classified into three categories: atherosclerotic (most common), inflammatory, and noninflammatory (18). Morphologically they may be saccular or fusiform.

It can be difficult to distinguish visually between the underlying causes of CAAs, particularly as many are discovered incidentally. Atherosclerotic aneurysms are characteristically fusiform with mural calcifications, and there is usually evidence of plaque in other vessels. Multiple CAAs, particularly if giant (defined as more than four times the normal diameter), suggest a diagnosis of Kawasaki disease (18). However, the appearances of most inflammatory and noninflammatory causes of CAAs are less predictable and are best assessed in combination with patient history and other systemic findings. The clinical significance of CAAs lies in the risk of thrombus formation within the aneurysm (Fig 6), which can dislodge and cause myocardial infarction. The critical nature of this condition is also highlighted by the uncommon but serious risk of aneurysmal rupture. The potential for complications increases with the size of the CAA.

Spontaneous Coronary Artery Dissection

SCAD is an increasingly recognized cause of acute coronary syndrome in young to middle-aged women without traditional cardiovascular risk factors. Reported in up to 4% of all acute coronary syndromes, it occurs secondary to hematoma in the wall of a coronary vessel that can eventually obstruct coronary blood flow, leading to cardiac ischemia and infarction (61). Patients can present with chest pain, acute myocardial infarction, and even sudden cardiac death. The exact cause is unknown but is hypothesized to be from either blood entering the subintimal space from the true lumen via an intimal flap or disruption of the traversing microvessels of the subintimal space resulting in hematoma formation from within the vessel wall itself (62). Fibromuscular dysplasia, pregnancy, states of extreme emotional distress, and various connective tissue disorders have reported associations with this condition.

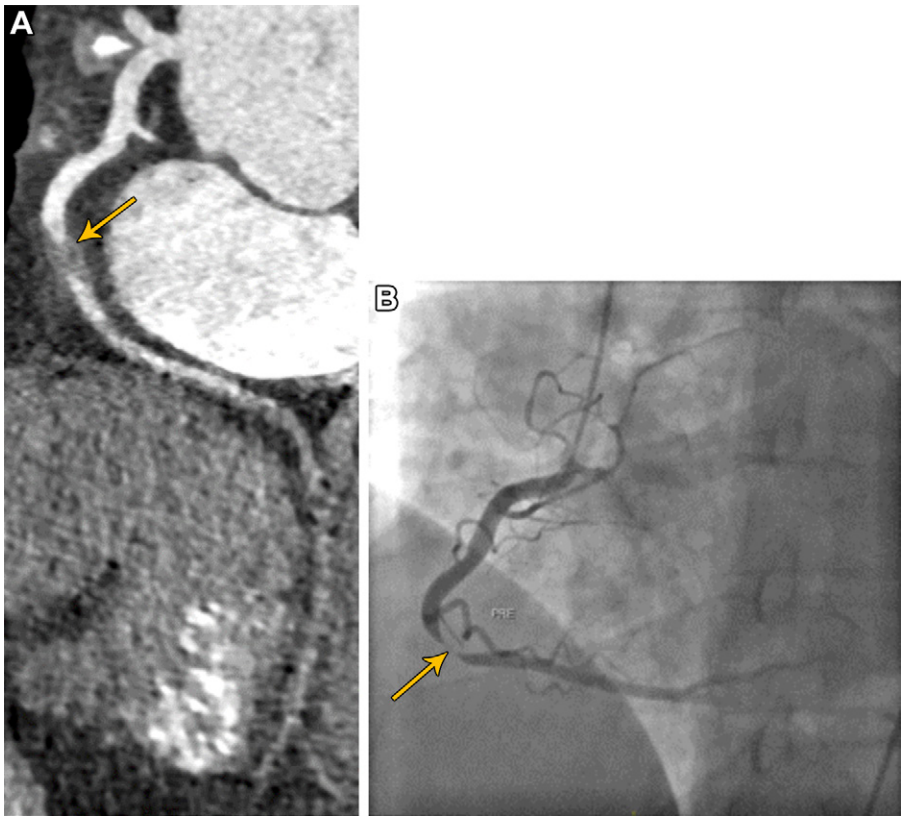


Figure 19. Acute plaque rupture in a 41-year-old man who presented with sudden-onset chest pain and electrocardiographic findings consistent with non-ST-segment elevation myocardial infarction (NSTEMI). **(A)** CCTA RCA curved reformation shows focal severe stenosis of the mid RCA at the level of a noncalcified plaque and/or thrombus with positive remodeling and mild perivascular epicardial fat stranding (arrow). **(B)** RCA catheter angiogram shows focal 99% stenosis along the mid RCA with an otherwise normal-caliber vessel (arrow). The patient had mixed atherosclerotic plaques in other vessels (not shown).

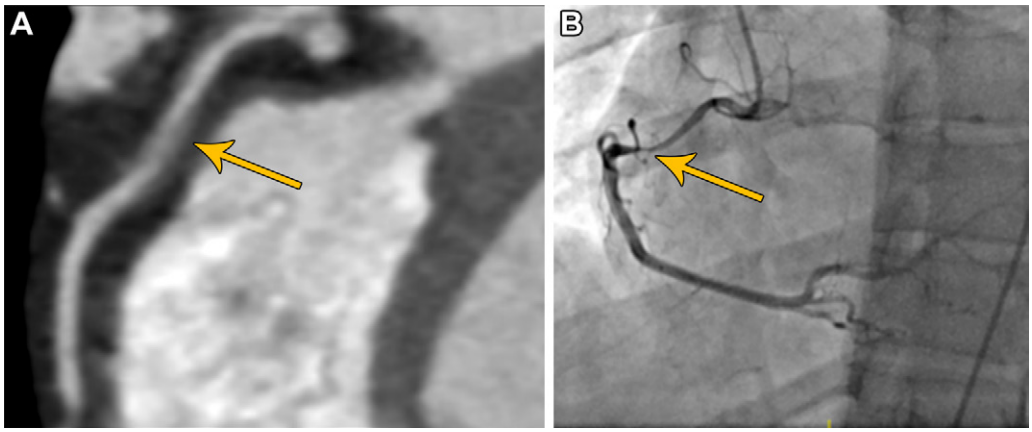


Figure 20. SCAD in a 45-year-old woman with a history of Ehlers-Danlos syndrome who presented with chest pain. **(A)** CCTA curved RCA reformation shows tubular mural thickening of the proximal/mid RCA with associated luminal stenosis and minimal periarteritis (arrows). **(B)** RCA catheter angiogram shows long-segment gradual luminal tapering of the RCA with up to 80% (severe) stenosis (arrow).

Invasive catheter-based coronary angiography has conventionally been the first-line imaging test for SCAD. However, modern CCTA is a viable alternative for diagnosis and monitoring of these patients. The Yip-Saw classification describes three angiographic patterns of SCAD: arterial wall contrast material staining with a radiolucent lumen (type 1), a long segment of smooth narrowing with distal tapering (type 2), and focal or tubular stenosis (type 3) (63). Type 2 is by far the most common pattern, seen in over 50% of cases, and can mimic the appearance of smooth arterial wall thickening seen in vasculitis (64). However, the presence of gradual distal tapering favors a diagnosis of SCAD (Fig 20). It most commonly involves the left anterior descending coronary artery and the mid to distal coronary segments (65). Skip lesions do not exclude the diagnosis. Because of its lower spatial resolution compared with that of conventional angiography and lack of dynamic flow imaging, CCTA was initially reserved for follow-up of these patients. Various appearances of SCAD at CCTA are now recognized in the acute setting (64). Tapered luminal stenosis can be the most useful sign at CCTA, with potential for vessel occlusion and periarterial epicardial fat stranding. In some cases, intramural hematoma or a dissection flap can also be seen. CCTA can be used for surveillance involving the proximal or large-caliber coronary arteries to assess vessel remodeling and preservation of flow over time (66).

Conclusion

Although suspected coronary artery stenosis, most commonly from atherosclerosis, is the most common cause for referral for CCTA, cardiac imagers should remain vigilant to the pattern of coronary stenosis and the configuration and distribution of any associated wall thickening, as well as associated

coronary aneurysms. The classification framework reviewed in this article is helpful in distinguishing coronary artery vasculitides from infiltrative processes mimicking CAV. Cardiac imagers may be the first to suggest a systemic disease as a cause of the patient's clinical presentation. Associated findings in the thoracic aorta and its branches and/or pulmonary artery may be appreciated at CCTA.

Multimodality imaging is crucial in the initial workup of CAV and encasement, and it also adds to prognostication. However, given the rarity of these entities, the benefit and frequency of imaging monitoring is not as well established across all patients with CAV or encasement. Review of other imaging studies and discussion with the patient's care team are important in arriving at the most likely diagnosis and helping guide follow-up.

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Disclosures of conflicts of interest.—**C.F.W.** Travel support for attending conferences from MUN during residency and fellowship. **J.D.C.** Society for Magnetic Resonance Angiography board member, Society for Cardiovascular Magnetic Resonance Board of Trustees member. All other authors, the editor, and the reviewers have disclosed no relevant relationships.

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