Nonatherosclerotic Coronary Vasculitis: Assessment with Cardiac MDCT

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Learning objectives

**Introduce** the various causes of coronary and cardiac vasculitis.

**Understand** the pathophysiology and the etiology of various coronary and cardiac vasculitis.

**Show** the potential of cardiac MDCT imaging to verify coronary and cardiac vasculitis on the basis of our cardiac MDCT registry with 45,327 cases.

*This exhibit aims to illustrate various imaging findings of coronary and cardiac vasculitis revealed by cardiac MDCT.*

Background

Previous articles that approximately 5% of patients with acute myocardial infarction do not have arterosclerotic CAD as demonstrated by invasive coronary angiography or necropsy.

Invasive coronary angiography has been considered as reference standard of CAD. However, it simply represents a *luminogram*, so it has limited to clearly performed the causes of CAD.

Currently, **multidetector-row computed tomography (MDCT)** coronary angiography is widely used as a powerful non-invasive tool for detection of CAD due to increased temporal and spatial resolution.

On the basis of our registry in 45,327 patients who underwent 64-slice MDCT between January 2006 and December 2011, we retrospectively reviewed the medical records and CT images and selected the patients with **coronary vasculitis**.

Imaging findings OR Procedure details

**Takayasu’s Arteritis** (Fig. 1-3)
Takayasu arteritis is an idiopathic vascular disorder characterized by the involvement of large vessels such as the aorta and its major branches, and it mainly affects young and middle-aged women.

Contrast-enhanced CT shows the characteristic "double-ring" sign, consisting of a poorly enhanced inside ring, which represents intimal swelling, and a well-enhanced outside ring, which represents active medial and adventitial inflammatory thickening, during the early stage of disease.

In approximately 10-30% of patients, the coronary artery exhibits one of the following pathologic features: type 1, stenosis or occlusion of the coronary ostia and the proximal segments of the coronary arteries; type 2, diffuse or focal coronary arteritis, which can extend diffusely to all epicardial branches or can involve focal segments, so-called skip lesions; and type 3, coronary aneurysms.

Among them, type 1 lesions are most frequent.

Coronary artery disease as a consequence of Takayasu arteritis, is relatively rare, resulting however in severe life-threatening complications in young patients.

**Kawasaki Vasculitis(Fig. 4-5)**

Cardiac sequelae of Kawasaki vasculitis are the most important manifestations, and the coronary arteries are involved in approximately 20% of patients with Kawasaki disease and subsequently can lead to myocardial infarction and/or sudden death.

In Kawasaki disease, panarteritis occurs in the acute phase and coronary artery aneurysms develop in the subacute phase.

In the chronic phase, aneurysms undergo regression or remodeling. Intramural collateral arteries might develop in occluded segments of the coronary arteries, and they appear as "arteries within the artery" and show a braided appearance on cardiac MDCT.

Despite treatment of acute phase with intravenous immunoglobulin and aspirin, up to 5% of Kawasaki disease patients, still continued to develop serious cardiac life-threatening complications, mainly giant coronary aneurysms and thrombotic stenoses, resulted in myocardial infarction and/or death.

**Behçet’s Disease(Fig. 6)**

Recently, rare cases of cardiac manifestations in Behçet disease have been reported (ie, aneurysm of the sinus of Valsalva, aortic or mitral valve involvement, proximal aortic dilatation, thrombus on right atrium, interatrial septal aneurysm, and myocardial infarction from coronary artery occlusion, endocarditis, myocarditis, pericarditis, or congestive cardiomyopathy).
Although the incidence and natural history of cardiac involvement in Behçet disease have not been clearly documented, the cardiac involvement in Behçet disease is associated with serious morbidity and mortality.

After literature review, the most common cardiac manifestations in Behçet disease were aneurysm of sinus of Valsalva and intracardiac thrombus.

In the almost reported cases of vascular-involved Behçet disease, coronary artery involvement by Behçet disease is extremely rare.

Coronary artery involvement occurs in <1% of cases and coronary artery bypass grafting is difficult because of tissue fragility and there is also a risk of pseudoaneurysm formation.

Hence, a rather conservative approach is proposed in case of symptomatic coronary artery disease.

IgG4 related Vasculitis(Fig. 7)

IgG4-related disease has not been well documented in the cardiovascular system.

Recently, Matsumoto et al reported a case of IgG4-related periarteritis in the coronary artery and abdominal aorta.

The case suggested that IgG4-related periarteritis could occur in the cardiovascular system and might manifest aneurysm or a tumorous lesion along the artery.

Interestingly, that case also had a 90% stenosis of the first diagonal coronary artery, so we should examine whether IgG4-related disease can manifest vascular lesions other than aneurismal or tumorous lesions.

Lately, Maturen et al reported two cases of coronary periarteritis in idiopathic retroperitoneal fibrosis (IRPF), both metachronous to primary IRPF presentation, and suggest attention to this anatomy in imaging assessment of patients with sclerosing diseases.

The case showed abnormal soft tissue encasing normal diameter left main and proximal left anterior descending coronary arteries.

Churg-Stauss Syndrome(Fig. 8)

Cardiac diseases related to Churg-Strauss syndrome include coronary vasculitis, myocarditis, valvular heart abnormalities, congestive heart failure and pericarditis.

Myocarditis and coronary arteritis account for approximately 50% of deaths if the disease remains unrecognized.
Pericardial effusion was detected at the time of diagnosis in 21 of 96 patients (22%), heart failure in nine, and transient heart block in three cases.

In another study, 35% of 112 patients had cardiac manifestations.

Pericarditis was present in 28 cases (25%) with tamponade in seven cases, cardiomyopathy in 27 cases (24%) of whom 20 (18%) had heart failure.

Coronary involvement has rarely been found pre-mortem in contrast to necropsy studies.

Coronary artery disease is significantly more common in ANCA-negative cases and is the leading cause of mortality with a frequency ranging from 16 to 92% in Churg-Strauss syndrome.

**Moyamoya Disease**(Fig. 9-10)

The moyamoya syndrome is a cerebrovascular condition that predisposes affected patients to stroke in association with progressive stenosis of the intracranial internal carotid arteries and their proximal branches.

The steno-occlusive changes in moyamoya disease are believed to be confined to the intracranial arteries and rarely occur in the extracranial arteries, including the renal arteries and coronary arteries.

Only 12 cases of moyamoya disease associated with coronary artery disease have been reported.

There are several findings that support that the case represents cardiac moyamoya pathophysiology: the bilateral ostial stenoses and the intracoronary ultrasound evaluation.

The likelihood of isolated ostial left main and right coronary artery stenosis with sparing of the distal coronary tree in a young patient with no coronary artery disease risk factors is extremely rare, whereas moyamoya of the cerebral circulation is by definition a bilateral focal finding.

The intracoronary ultrasound images obtained from the patient demonstrated homogenous soft, concentric, intimal proliferation with no evidence of hard, fibrotic or calcific build-up, or lipid pooling.

These are findings that are consistent with moyamoya pathophysiology and different from typical atherosclerotic disease.

**Images for this section:**
**Fig. 1:** 59-year-old woman with Takayasu arteritis involving left main coronary artery (LMCA). Curved multiplanar reformation (MPR) (A), volume-rendering (VR) (B), and axial MDCT (C) images show severe luminal narrowing (red, yellow, blue arrows) at ostium of LMCA.

**Fig. 2:** 71-year-old man with Takayasu arteritis involving LMCA and RCA. VR (A), curved MPR (B), (C) MDCT images show multifocal stenoses by soft tissue thickening at LMCA, proximal and mid LAD and proximal to distal RCA; so-called skip lesions (red, yellow, blue arrows).
**Fig. 3:** 35-year-old woman with Takayasu arteritis. VR (A), curved MPR (B) MDCT images show focal aneurysmal dilatation of diagonal branch (red arrows). Thoracic CT with axial scan (C) shows pseudoaneurysm at upper intercostal artery (yellow arrow). MRI with gadolinum-enhanced fat suppressed T1 weighted image (T1WI) (D) shows wall thickening with slight enhancement (blue arrow) at descending thoracic aorta.

**Fig. 4:** 43-year-old man with Kawasaki vasculitis involving all coronary arteries. VR (A), (B) MDCT images show multifocal ectasia at RCA (red arrow), LAD (yellow arrow), and left circumplex artery (LCX) (blue arrow) without intraluminal thrombus.

**Fig. 5:** 71-year-old man with Takayasu arteritis involving LMCA and RCA. VR (A), curved MPR (B), (C) MDCT images show multifocal stenoses by soft tissue thickening at LMCA, proximal and mid LAD and proximal to distal RCA; so-called skip lesions (red, yellow, blue arrows).
Fig. 6: 60-year-old man with Behçet’s disease involving ascending aorta and aortic arch vessels. Left (A), (D): VR images showed multifocal stenosis at all coronary arteries due to calcified plaque (red arrows). Mid (B), (E): borderline fusiform dilatation of ascending aorta (maximum diameter: 4cm, yellow arrows). Mid and Right (B), (C), (E), (F): aortic wall thickening with enhancement at ascending aorta and aortic arch vessels (blue arrow).
**Fig. 7:** 68-year-old woman with IgG4 related sclerosing disease involving LAD. Curved MPR (A), VR (B) MDCT images clearly show luminal narrowing at proximal LAD (red, yellow arrows).

**Fig. 8:** 69-year-old male with Churg-Strauss syndrome involving RCA and LCX. Oblique coronal MDCT (A), (B) images show aneurysmal dilatation and luminal narrowing at proximal RCA (red arrows) and luminal stenosis by concentric wall thickening at proximal LCX (yellow arrows). After 1-year, oblique coronal MDCT (C), (D) images show progression of concentric wall thickening at proximal LCX (blue arrows).

**Fig. 9:** 20-year-old man with Moyamoya disease involving LAD. VR MDCT (A), conventional angiography (B), and SA view MDCT (E) images show discrete moderate luminal stenosis at proximal LAD (red, yellow arrows). Intravascular ultrasonograph (IVUS) images (C), (D) show luminal stenosis at proximal LAD due to wall thickening with fibrous (green) or fibrous-fatty (light green) tissue (blue arrows).
Fig. 10: 53-year-old man with Moyamoya disease involving LMCA. 4C view (A), VR (B), curved MPR MDCT (C), and conventional angiography (D) images show moderate stenosis at the LMCA (red arrows). Conventional angiography (E) image shows patent flow after LMCA stenting (yellow arrow). Other that this lesion, he also has multifocal calcified plaques along the three coronary arteries. IVUS images (F) shows luminal stenosis at LMCA due to wall thickening with fibrous (green) or fibrous-fatty (light green) tissue (blue arrows).
Conclusion

The true prevalence and clinical importance of coronary vasculitis, both at the time of presentation and during evolution of the disease, is difficult to delineate from the existing literature data.

Technical advances of cardiac MDCT allow for the noninvasive assessment of coronary artery vasculitis.

This sophisticated technological assessment reveals frequencies of cardiac abnormalities similar to and even higher than the older autopsy studies for coronary artery vasculitis.

However, the clinical significance of these findings is not yet established. Moreover, cardiac MDCT enables us to detect various nonatherosclerotic causes of CAD as well as atherosclerotic lesions.

Recognition of the various imaging findings of nonatherosclerotic, nonanomalous coronary artery vasculitis revealed by cardiac MDCT is important for optimizing patient care.

In conclusion, radiologists should be familiar with various findings and their clinical significance, as assessed through MDCT for accurate diagnosis and proper management in patients with coronary artery vasculitis.

References


Personal Information