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PICTORIAL REVIEW

# Spectrum of angiographic findings in aortoarteritis

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Angiographic appearances are characteristic, distinctive and a major basis of established criteria in the diagnosis of aortoarteritis. We present a pictorial review of digital subtraction angiography imaging in patients with proven aortoarteritis, based upon 16 years' experience in our institution. Understanding of these angiographic appearances is important for definitive diagnosis, and for evaluation of the extent of the disease in order to plan appropriate further management.


## Introduction

Aortoarteritis is recognized as a non-specific inflammation of the aorta and its main branches, and is frequently associated with reactive intimal proliferation.[1] This usually leads to luminal stenosis, occlusion, dilatation and aneurysm formation of involved vessels.[1] This disease is also known as aortitis syndrome, Takayasu's arteritis, middle aortic syndrome, pulseless disease, atypical coarctation of aorta and aortic arch syndrome.[1] It is most common in young populations in the Orient, although it occurs worldwide,[2] being predominantly a disease of young adults in the second and third decades of life.[3] The reported female:male ratio has varied from 9:1 to 1.3:1.[4,5]

The disease is a panarteritis, mainly involving the media. The wall of the artery is diffusely or irregularly thickened and fibrotic. The thickened, whitish-grey intima protrudes into the lumen, causing luminal stenosis and occlusion of the vessel.[1] Microscopically, extensive fragmentation, destruction and fibrosis of the elastic and smooth muscle fibres are found in the media, in combination with inflammatory cell infiltration, predominantly of lymphocytes and monocytes, and granulation tissue proliferation.[1] Traditionally a conventional aortogram, and presently digital substraction angiography, are the main imaging methods to establish the diagnosis and enable disease mapping.

None of the diagnostic criteria is entirely satisfactory, but the clinical diagnosis is seldom difficult. Essentially, the diagnosis depends on the typical angiographic morphology, history or presence of constitutional symptoms suggestive of a systemic illness, and the differential diagnosis from other, similar conditions such as other causes of inflammatory aortitis (e.g., syphilis, tuberculosis, giant cell arteritis, Buerger's, Behçet's, Cogan and Kawasaki diseases, spondyloarthropathies) and developmental abnormalities (e.g., Ehlers–Danlos syndrome, Marfan's syndrome). The criteria for the diagnosis of Takayasu's arteritis as suggested by Ishikawa[6] are shown in Table 1.

Nasu et al.[7] classified angiographic findings into 4 types, comprising:

type I, involvement of branches of the aortic arch only
type II, involvement of the thoracic aorta and its branches
type III, involvement of the abdominal aorta and its branches only

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**Table 1** Ishikawa's criteria for the clinical diagnosis of Takayasu's disease[6]

| Criteria[a] | Definition |
|---|---|
| Obligatory criteria | |
| Age <40 years | Age <40 years at diagnosis or at-onset characteristic signs and symptoms of 1 month duration in patient history |
| Two major criteria | |
| Left mid subclavian artery lesion | The most severe stenosis or occlusion present in the mid portion, from 1 cm proximal to the left vertebral artery orifice to 3 cm distal to the orifice, determined by angiography |
| Right mid subclavian artery lesion | The most severe stenosis or occlusion present in the mid portion, from the right vertebral artery orifice to 3 cm distal to the orifice, determined by angiography |
| Nine minor criteria | |
| High ESR | Unexplained persistent high ESR >20 mm/h (Westergren) at diagnosis, or evidence in patient history |
| Carotid artery tenderness | Unilateral or bilateral tenderness of common carotid arteries by physician palpation |
| Hypertension | Persistent blood pressure 140/90 mmHg brachial or >160/90 mmHg popliteal at age <40years, or history thereof |
| Aortic regurgitation or annuloaortic ectasia | By auscultation or Doppler echocardiography or angiography |
| Pulmonary artery lesions | By angiography, scinitigraphy or 2D echocardiography: lobar or segmental arterial occlusion or stenosis, aneurysm or luminal irregularity in pulmonary trunk or unilateral or bilateral pulmonary arteries |
| Left mid common carotid lesion | Most severe stenosis or occlusion in the mid portion, 5 cm in length from 2 cm distal to its orifice, by angiography |
| Distal brachiocephalic trunk lesion | Most severe stenosis or occlusion in the distal third, by angiography |
| Descending thoracic aorta lesion | Narrowing, dilatation, aneurysm or luminal irregularity by angiography; tortuosity alone unacceptable |
| Abdominal aorta lesion | Narrowing, dilatation, aneurysm, luminal irregularity or any combination, and absence of lesions over 2 cm of terminal aorta and bilateral common iliac arteries, by angiography; tortuosity alone unacceptable |

[a] The proposed criteria consist of one obligatory criterion, two major criteria and nine minor criteria. In addition to the obligatory criterion, the presence of two major criteria, or one major and two or more minor criteria or four more minor criteria, suggests a high probability of Takayasu's disease.

type IV, extensive involvement of the whole length of the aorta, and its branches.

Depending on the location, type and severity of the lesions, aortoarteritis presents with a wide variety of clinical manifestations or syndromes, such as renovascular hypertension, aortic arch syndrome, aortic coarctation syndrome and obstructive disease of the aortoiliac arteries.[1]

## Study

From June 1988 to May 2004, 548 patients, with diagnoses based on the criteria established by the Aortitis Syndrome Research Committee of Japan,[8] were studied by digital substraction angiography. Clinical features included:

symptoms caused by ischaemia of the central nervous system, upper extremities or kidney

fever, absent or decreased pulses, bruits and fundoscopic findings

raised ESR or presence of C-reactive protein

characteristic angiographic findings.

Characteristic angiographic findings involved a spectrum of changes ranging from minimal intimal irregularity to typical rat-tail narrowing, complete occlusion or aneurysm formation in the involved blood vessel. Simultaneous involvement of the aorta and its medium-sized branches, or of at least two medium-sized branches in the absence of aortic involvement, was considered essential for the diagnosis of aortoarteritis. The examination protocol comprised an AP view of abdominal and pelvic vessels and an LAO view of the aortic arch and its branches; further examination, including additional oblique views of the aorta for evaluation of neck and visceral branches, were tailored to the situation.

## Spectrum of angiographic findings

The disease process of aortoarteritis predominantly results in arterial stenosis or occlusion, and also