Diagn Interv Radiol 2008; 13:159-162

Angiographic signs in specific vasculitides

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ABSTRACT
Vasculitis, defined by inflammation in the blood vessel wall and reactive mural damage, may present with various clinical signs, depending on the organs involved. Infection, collagen vascular disease, and granulomatous disease or idiopathic inflammation of the vessels may result in various degrees of stenosis, obstruction, aneurysmal dilatation, or rupture. The definitive pathologic diagnosis is established by biopsy; however, the biopsy may pose significant risk to the patient; and the biopsy sample may not be adequate. Knowledge of the angiographic signs of various vasculitides should help in interpreting the clinical picture and reducing the need for biopsy. In this article, we describe the angiographic signs of the most common vasculitides, and review their clinical signs.

Key words: • *vasculitis* • *digital subtraction angiography*

Vasculitis is inflammation and necrosis of the blood vessels, sometimes with granuloma formation. It may result in obstruction, aneurysm formation, and even rupture, leading to ischemia of the tissues supplied by the involved artery. It may be associated with diseases presenting with very different clinicopathological pictures. Laboratory findings are often non-specific. Although the definitive diagnosis is established by biopsy, biopsy samples are not always adequate, and the biopsy procedure may be risky. Hence, the presumptive diagnosis relies on clinical and angiographic findings and on the response to treatment. Knowledge of the angiographic signs of specific vasculitides would assist in interpreting the clinical picture to establish a diagnosis.

Takayasu's arteritis

A disease of the young adults in their second and third decades, Takayasu's arteritis is a panarteritis. This disease involves large arteries such as the aorta and its main branches, as well as the coronary arteries and pulmonary arteries. Diffuse involvement of the artery wall and irregular thickening result in stenosis or occlusion; the loss of the elastic fibers in the media results in dilatation and aneurysm. Vascular lesions may cause malignant hypertension, retinopathy, aortic regurgitation, cerebral ischemia, and claudication (1, 2).

Stenosis and occlusion of the aortic arch are rare in Takayasu's arteritis. Among the supra-aortic branches, the subclavian and the left carotid branches are the most commonly involved (Fig. 1). Lesions are generally located proximally in a long segment, and have smooth contours (Figs. 2, 3). Abrupt termination and flame-shaped terminations are sometimes noted. Severe stenosis in the origin of the subclavian artery or proximal to the orifice of the vertebral artery may result in subclavian steal syndrome. In long-segment occlusion of the subclavian artery beyond the orifice of the vertebral artery, collaterals develop from the thyrocervical trunk and lateral thoracic arteries (Fig. 4).

Involvement of the descending thoracic aorta is generally in the form of diffuse stenosis accompanied by localized stenosis. Focal stenosis at the border of the thoracic descending aorta and abdominal aorta is observed rarely. Collaterals develop from the intercostal arteries and internal mammary arteries when there is stenosis or occlusion of the descending thoracic aorta. Circulation is supplied by the lower intercostals, lumbar arteries, internal mammary arteries, and superior inferior epigastric arteries in cases of occlusion of the proximal abdominal aorta. However, with occlusion of the infrarenal aorta, collaterals develop from the inferior mesenteric and iliac arteries.

The most commonly involved branches of the abdominal aorta include the renal arteries (especially the proximal segments) and their ostia (Figs. 5, 6). Although the stenosis is often localized, it may also

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Received 9 January 2007; revision requested 19 March 2007; revision received 4 April 2007; accepted 13 April 2007.

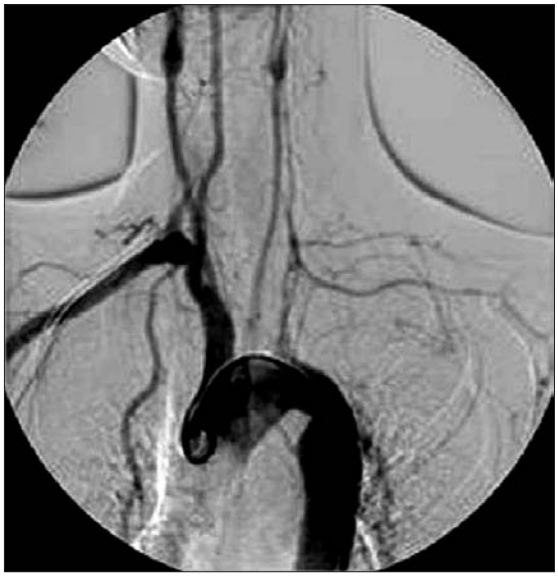

Figure 1. Takayasu's arteritis. Preocclusive stenosis in the left subclavian artery, and a long segment stenosis in the left carotid artery.

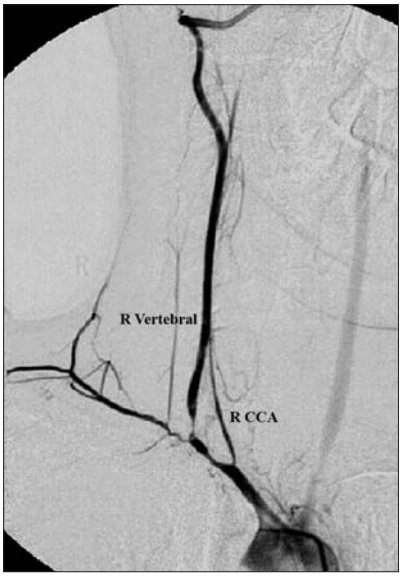

Figure 2. Takayasu's arteritis. Occlusion of the right common carotid artery and postvertebral segment of the right subclavian artery, and severe stenosis in the proximal vertebral artery (R, right; CCA, common carotid artery).

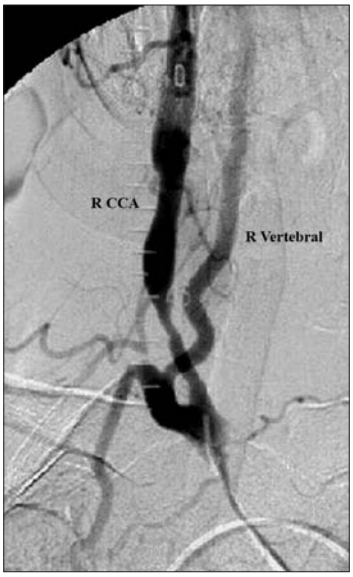

Figure 3. Takayasu's arteritis. Severe stenosis in the proximal right common carotid artery, with post-stenotic dilatation (R, right;, CCA, common carotid artery).

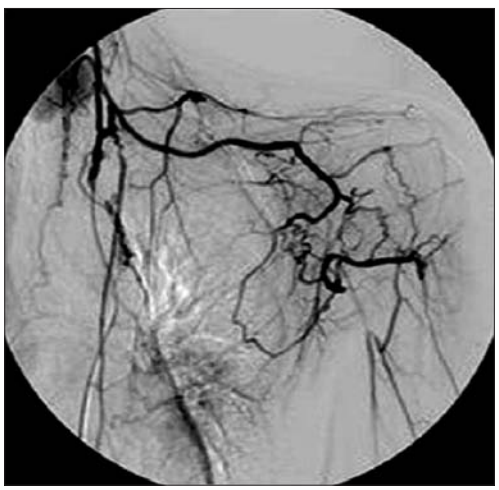

Figure 4. Takayasu's arteritis. Occlusion of the left subclavian artery, and collateral arteries.

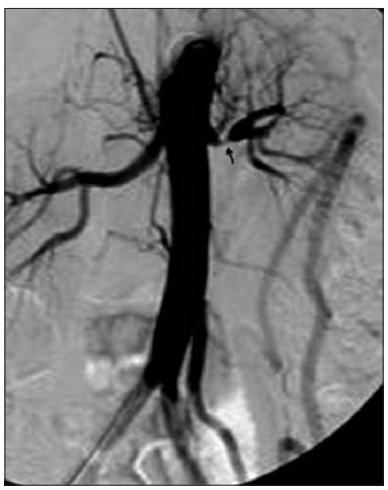

Figure 5. Takayasu's arteritis. A short-segment stenosis is seen in the proximal part of the left main renal artery.

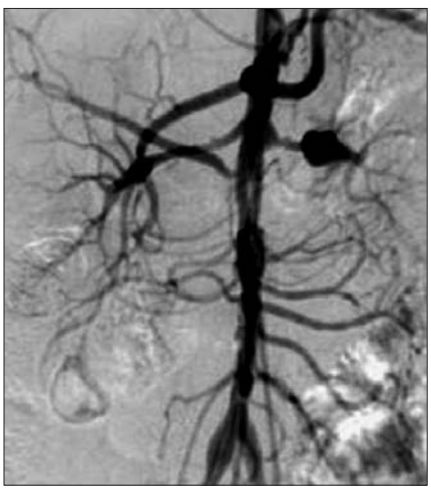

Figure 6. Takayasu's arteritis. A long-segment stenosis and contour irregularity in the infrarenal abdominal aorta, and proximal stenosis and post-stenotic aneurysmal dilatation in the left main renal artery.

involve the entire artery. Capsular and periureteric collaterals are observed in occlusions of the renal artery.

Among the visceral arteries, the ostium of the superior mesenteric artery is the one that most commonly presents with stenosis or occlusion. The inferior mesenteric artery is often preserved; however, it may be highly dilated as a collateral in cases of superior mesenteric artery involvement. Stenoocclusive lesions of the iliac artery are very rare.

In Takayasu's arteritis, dilatation and aneurysms are observed less commonly than are stenosis and occlusion. Dilatation is seen most commonly in the ascending aorta and aortic arch. Aneurysms, which may be fusiform, saccular, or dissecting, are more common in the thoracic descending and abdominal aorta.

The current angiographic classification of Takayasu's arteritis (by location) has been defined at the Takayasu's Congress in 1994:

Type I : Main branches of the aortic arch
Type IIa : Ascending aorta, aortic arch and its branches
Type IIb : Ascending aorta, aortic arch and its branches, and thoracic descending aorta
Type III : Thoracic descending aorta, abdominal aorta, and/or renal arteries
Type IV : Abdominal aorta and/or renal arteries
Type V : Type IIb and IV occurring together

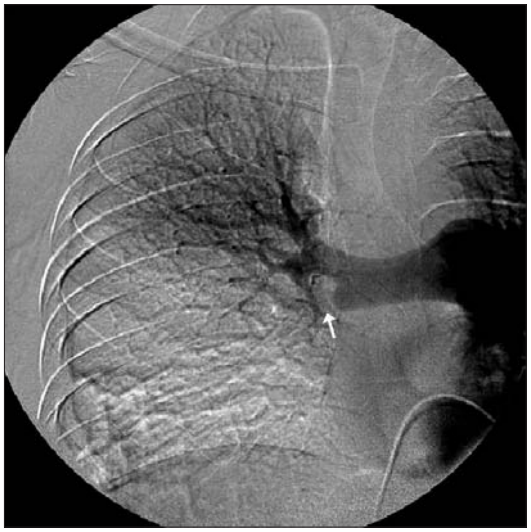

Figure 7. Behçet's disease. Occlusion in the interlobar artery *(arrow)*.

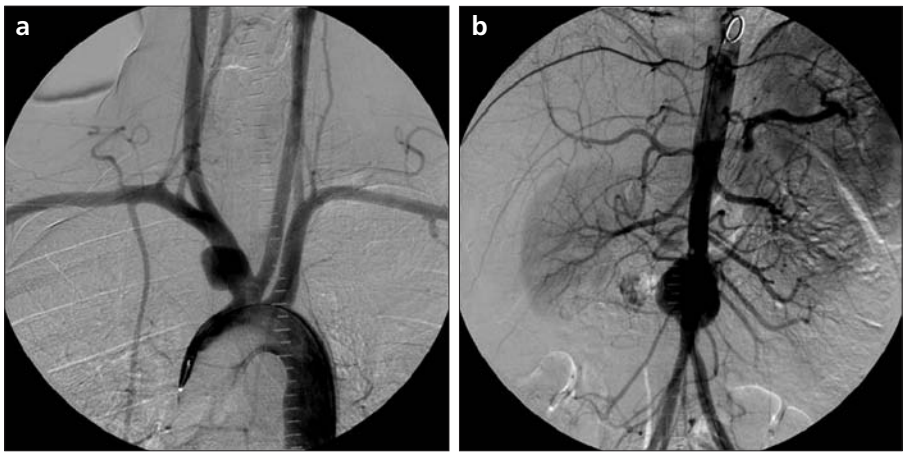

Figure 8. a, b. Behçet's disease. Brachiocephalic truncal aneurysm **(a)** and infrarenal abdominal aortic saccular aneurysm **(b)**.

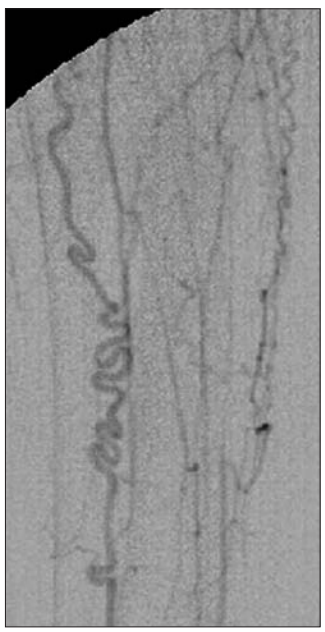

Figure 9. Buerger's disease. Corkscrew appearance in the leg collateral arteries.

In addition, coronary artery involvement and pulmonary artery involvement are designated K+ and P+, respectively.

Behçet's disease

Behçet's disease is a rare, multisystem chronic inflammatory disease, characterized by recurrent oral ulcers, and any of several other manifestations including genital ulcers, ocular disease, skin lesions, and a positive pathergy test. Neurologic, gastrointestinal, and cardiovascular system involvement and arteritis also may be observed.

Arteries and veins of various sizes and locations may be involved. About 20–40% of Behçet patients have vascular complications. Lesions of the venous system are more common than are lesions of the arterial system (3). Occlusion (Fig. 7) and aneurysms (Fig. 8) of the arterial system are seen in the pulmonary artery, aortic arch, and subclavian and coronary arteries. Behçet's disease is the only vasculitis associated with pulmonary artery aneurysm.

Buerger's disease (thromboangiitis obliterans)

Buerger's disease is a non-atherosclerotic, inflammatory, occlusive vascular disease of small and medium-sized arteries and veins of the extremities seen (almost exclusively) in smokers. It rarely involves the visceral and cerebral arteries (4, 5).

Digital substraction angiography (DSA) is the method of radiologic examination that gives the most detailed information in this disease.

Multiple, bilateral, symmetrical segmental stenoses and occlusions are located distally in the arteries of the leg and forearm with inadequate collateral vessels described as "corkscrew collaterals" (Fig. 9). The lack of atherosclerotic changes in the more proximal vessels is a characteristic finding.

Polyarteritis nodosa

Polyarteritis nodosa is a rare necrotizing vasculitis that involves small and medium-sized arteries. Patients present with clinical findings (fever, abdominal pain, fibromyalgia, weight loss, erythema nodosum) and laboratory findings (increased sedimentation rate and C-reactive protein levels, anemia, reduced albumin) suggestive of systemic necrotizing vasculitis.

DSA is the most valuable method for demonstrating the involvement of the vascular system. In a study by Hagspiel et al., the sensitivity and specificity of angiography in the diagnosis of polyarteritis nodosa were 89% and 90%, respectively (6). Visceral arteries, and less commonly, arteries of the extremities and the smaller branches of the aorta are involved (7).

This vasculitis is typically focal and segmental, and is more prominent at arterial bifurcations. Fusiform or saccular aneurysms, irregular stenosis, thinning, and abrupt terminations are observed in small and medium-sized arteries (Fig. 10).

Aneurysms are visualized by angiography in 12.5–94% of the cases (6, 7).

Temporal arteritis

Temporal arteritis is the most commonly observed vasculitis in adults older than 50. Systemic symptoms in-

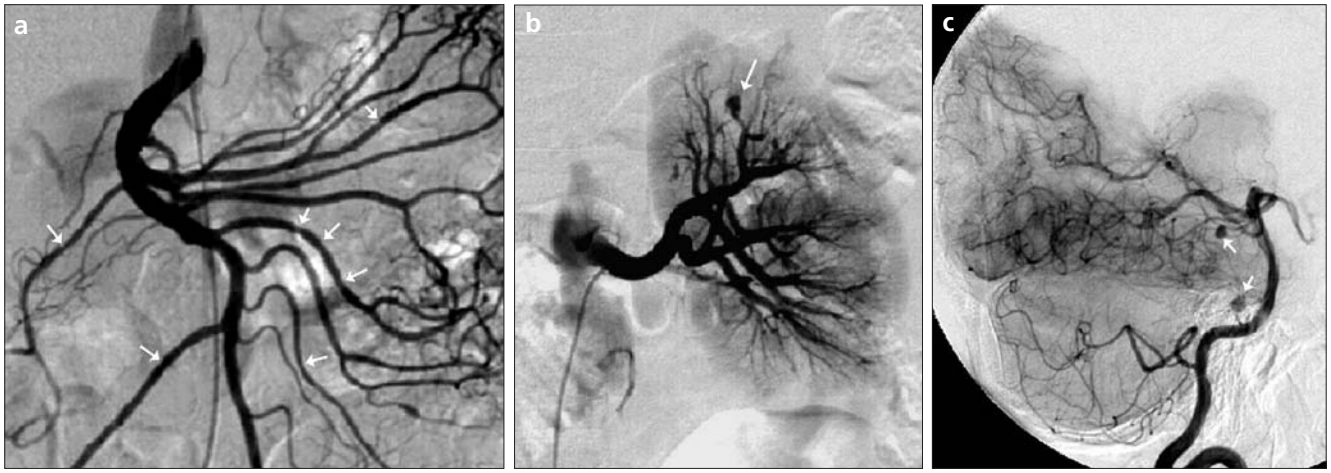

Figure 10. a–c. Polyarteritis nodosa. Small aneurysms in the branches of the superior mesenteric artery, and irregular stenosis, thinning, and abrupt terminations in the arteries **(a,** *arrows***)**. Aneurysms in the distal branches of the left renal artery **(b,** *arrow***)**. Aneurysms in the superior cerebellar artery and anterior inferior cerebellar arteries **(c,** *arrows***)**.

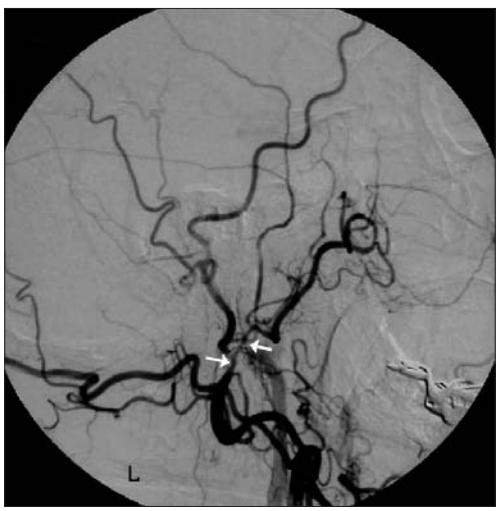

Figure 11. Temporal arteritis. Contour irregularity due to stenotic segments in the superficial temporal arteries *(arrows)*.

clude head and neck pain, sensitivity of the scalp, jaw claudication, transient ischemic attacks, fatigue, weight loss, fever, and ocular symptoms including pain, amaurosis fugax, diplopia, visual hallucinations, and loss of vision (8). Irregularity and stenosis may be observed in the superficial temporal artery (Fig. 11).

In conclusion, DSA is the most commonly utilized method of radiological examination for demonstrating the anatomopathological and hemodynamic changes in vascular structures suggesting vasculitis, for determining the extent of disease, and for assessing the results of interventions. Knowledge of the systemic clinical signs as well as the angiographic signs of specific vasculitides allows a specific diagnosis in the majority of patients, and may obviate the need for biopsy.

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