Systemic Vasculopathies
Imaging and Management

Ankaj Khosla, MD[a], Brice Andring, MD[a], Benjamin Atchie, MD[a], Joseph Zerr, MD[a], Benjamin White, MD[a], Jarrod MacFarlane, MD[a], Sanjeeva P. Kalva, MD[a,b,*]

KEYWORDS

• Vasculitis • Vasculopathy • Arteritis • Connective tissue disease

KEY POINTS

- Systemic vasculopathies represent a wide variety of vascular disorders characterized by vessel wall thickening, luminal irregularities, stenosis, occlusion, dissection, and aneurysm formation.
- Imaging plays an important role in the management of systemic vasculopathies by identifying the involved vessels and vascular abnormalities, assessing disease progression and the risk of complications, localizing active areas of vascular inflammation, and recognizing end organ damage.
- This article discusses various common systemic vasculopathies emphasizing the salient vascular imaging findings and the role of endovascular therapy for management of these conditions.

INTRODUCTION

Systemic vasculopathies represent multifocal vascular disorders that occur either in isolation (of unknown etiology) or are associated with systemic inflammatory/infective/neoplastic conditions, connective tissue disorders, or chemical toxicity (**Box 1**).[1] These vasculopathies are often classified according to the size of the vessels involved (**Box 2**) to facilitate practical approach for diagnosis and management.[2] Despite varied etiologies and vessels involved, the pathologic manifestations of various systemic vasculopathies remain similar, making it difficult to diagnose these conditions solely on imaging morphology. Associated clinical manifestations, involvement of other organs, laboratory findings, and other radiologic tests help distinguish these entities. Some conditions require histopathologic confirmation for definitive diagnosis. Imaging plays an important role in the management of systemic vasculopathies by identifying the involved vessels and vascular abnormalities, assessing disease progression and the risk of complications, localizing active areas of vascular inflammation, and recognizing end organ damage. This review briefly discusses various common systemic vasculopathies emphasizing the salient vascular imaging findings and the role of endovascular therapy for management of these conditions.

LARGE-VESSEL VASCULITIS

Takayasu Arteritis

Also known as *pulseless disease* and *aortic arch syndrome*, Takayasu arteritis is an idiopathic large-vessel arteritis that involves the aorta and its major branches, pulmonary arteries, and coronary arteries. The disease often manifests before the age of 40 years with a distinct female preponderance. Patients typically present with nonspecific constitutional symptoms during initial phase of the disease. Active inflammatory phase is associated with elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) levels. Chronic granulomatous and lymphocytic

Disclosure: No relevant financial disclosures by any authors.
[a] Department of Radiology, University of Texas Southwestern Medical Center, 5393 Harry Hines Boulevard, Dallas, TX 75390, USA; [b] Interventional Radiology, UT Southwestern Medical Center, 5393 Harry Hines Boulevard, Dallas, TX 75390-8834, USA
* Corresponding author. Interventional Radiology, UT Southwestern Medical Center, 5393 Harry Hines Boulevard, Dallas, TX 75390-8834.
E-mail address: Sanjeeva.Kalva@UTSouthwestern.edu

Radiol Clin N Am 54 (2016) 613–628
http://dx.doi.org/10.1016/j.rcl.2015.12.011
0033-8389/16/$ – see front matter

Box 1
Systemic vasculopathies

Inflammatory/immune complex conditions
Takayasu arteritis
GCA
Systemic lupus erythematosus
Sjögren's syndrome and scleroderma
Rheumatoid arthritis
Seronegative arthritis
Behçet disease–associated vasculitis
PAN
Kawasaki disease
Thromboangiitis obliterans
Churg Strauss syndrome
Leukocytoclastic vasculitis
Henoch-Schonlein purpura
Wegener granulomatosis (granulomatosis with polyangiitis)

Connective tissue disorders
Marfan syndrome
LDS
EDS

Infective
Septic emboli
Viral infections (eg, herpes, hepatitis B and C, human immunodeficiency virus)
Bacterial infections

Neoplastic
Hematologic malignancies
Solid organ malignancies

Unknown etiology
Fibromuscular dysplasia
Segmental arterial mediolysis

Chemical toxicity
Antimicrobials (penicillins, cephalosporins, tetracyclines, gentamicin, sulfasalazine, quinolones)
Antithyroid medications (Propylthiouracil, methimazole)
Drugs of abuse (cocaine, heroin, methamphetamine, ecstasy)
Psychotropic medications (olanzapine, trazodone, clozapine)
Analgesics (naproxen, ketorolac, sulindac, indomethacin)
Cardiovascular drugs (Digitalis, Hydralazine, Methyldopa, Thiazides)
Anticonvulsants (Phenytoin, Carbamazepine)
Anti–tumor necrosis factor–α agents (infliximab, etanercept, adalimumab)
Others (phenylpropanolamine, sulfonamides, leukotriene inhibitors)