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Techniques in Vascular and Interventional Radiology

Diagnostic Approach in Patients With Suspected Vasculitis

Aditya M. Sharma, MBBS,* Shailendra Singh, MBBS,† and Janet E. Lewis, MD†

Vasculitides are a heterogeneous group of disorders that share the common feature of inflammation of the blood vessel wall. Vasculitis can be a systemic or localized process and depending on the disorder can affect large, medium, or small vessels. Vascular physicians including interventional radiologists often may be involved early in these cases before the establishment of a diagnosis as these patients may present with manifestations attributable to occlusive vascular syndromes. In this article, we discuss the presenting signs and symptoms of patients with vasculitis as well as laboratory and imaging studies required to further evaluate these disorders and treatment options, which include interventional as well as noninterventional options.

KEYWORDS vasculitis, vasculitidies, non-atherosclerotic vascular diseases

Introduction

The cardinal feature of vasculitis is inflammation of blood vessel walls, which classically affects the arteries but also can occur in veins. In vasculitis, a key histologic finding is of an inflammatory infiltrate that results in damage to the blood vessel wall. This often leads to narrowing, occlusion, or rupture of the involved vessel leading to end-organ or tissue damage. Scarring and weakness of the vessel wall can occur after the acute stage leading to symptoms of chronic vascular insufficiency and aneurysms.[1-3] Owing to the diverse presentations including varied organ distribution and size of vessel involvement, these disorders can be challenging to diagnose and treat. Interpretation of imaging studies is further complicated by noninflammatory disorders that mimic vasculitis.

Classification

Vasculitis can be classified as a primary process or secondary to an underlying disease and is often due to humoral or cellular immune-related injury.[1-4] Primary vasculitis is typically considered to be idiopathic as the causative mechanisms remain poorly understood. Secondary vasculitides are typically associated with other conditions such as infections, medications, and connective tissue disease (Table 1). Vasculitis is commonly classified based on the size of the involved blood vessel (Table 2). The vasculitides can be further characterized based on the presence of certain antibodies particularly antineutrophil cytoplasmic antibody (ANCA) as well as involvement of certain organ systems.

Large Vessel Vasculitis

The 3 most common causes of large vessel vasculitis are giant cell arteritis (GCA), Takayasu arteritis (TA), and idiopathic aortitis. GCA is a granulomatous arteritis that affects the major branches of the aorta and has a predilection for extracranial branches of the carotid arteries.[1] It also can affect the aorta. Although multinucleated giant cells are a classic finding, they may be absent in biopsy specimens. The incidence of GCA increases with age, and it is uncommon before 50 years of age. It is more common in women and whites particularly of Northern European descent with an incidence of 15-25 per 100,000 people older than 50 years.[5,6] Typically, branches of the external, internal carotid, and subclavian arteries are affected, and luminal narrowing or occlusion causes blindness, headache, scalp tenderness, cervical bruits, pulselessness, and arm and jaw claudication.[1] Less commonly, the aorta and

*Division of Cardiovascular Medicine, Department of Internal Medicine, University of Virginia School of Medicine, Charlottesville, VA.
†Division of Rheumatology, Department of Internal Medicine University of Virginia School of Medicine, Charlottesville, VA.
Address reprint requests to Aditya Sharma, MBBS, PO BOX 800158, Charlottesville, VA 22901. E-mail: asharma@virginia.edu

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Table 1 Secondary Causes for Vasculitis (The List Includes Most of the Disorders; However, It Is Not a Complete List)

Infectious etiology
Virus
Hepatitis B and C
Human immunodeficiency virus
Parvovirus B19
Cytomegalovirus
Herpes simplex virus
Varicella zoster
Bacteria
Salmonella
Streptococcus
Staphylococcus
Clostridium septicum
Chlamydia pneumoniae
Mycobacterium tuberculosis
Treponema pallidum
Borrelia burgdorferi
Mycoplasma
Cryptococcus
Neisseria
Coccidioides
Connective tissue disorders
Relapsing Polychondritis
Cogan Syndrome
Rheumatoid arthritis
Sjögren syndrome
Systemic lupus erythematosus
Scleroderma
Drugs
D-Penicillamine
Penicillin
Propylthiouracil
Hydralazine
Minocycline
Cocaine
Leukotriene inhibitors (association with Churg-Strauss remains controversial)
Sulfasalazine
Ciprofloxacin
Pantoprazole
Phenytoin
Allopurinol
Sulfonamides
Thiazides
Neoplasia
Hematologic malignancies (myeloproliferative and lymphoproliferative disorders)
Solid organ tumors including lung, colon, and GI carcinomas

runoff vessels are affected, which may be manifested by claudication or even rest pain from severe occlusive disease.[7] Over time, vascular scarring and aneurysms may develop. TA is another large vessel vasculitis that typically affects the aorta and branches but can also affect the pulmonary and coronary arteries. Vessel involvement is often patchy resulting in segmental areas of stenosis and sometimes areas of dilation and aneurysm formation. TA is a relatively rare disorder that tends to affect young women particularly of Asian descent. Onset typically occurs before the age of 40 years. The annual incidence is estimated at 2.6 cases of TA per million Americans.[2] Presentation can range from asymptomatic to severe limb ischemia or claudication as well as stroke or transient ischemic attacks.[8] TA typically occurs more frequently in the upper extremities, but lower extremities may be involved. Concomitant constitutional and systemic symptoms are common.[2,8] The tendency for individuals with this disorder to have greatly reduced upper extremity pulses has led to TA being referred to as "pulseless disease."

Patients with GCA will usually have significantly elevated levels of inflammatory markers such as erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). In a meta-analysis of 941 cases of biopsy-proven GCA, the ESR was within reference range in only 4%.[9]

GCA is one of the causes of marked elevations in inflammatory parameters and should be included in the differential diagnosis of an elderly patient presenting with constitutional symptoms and an ESR >100. The ESR and CRP level are often elevated in TA but can be within reference range in up to half of the cases of TA even in

Table 2 Classification of Primary Vasculitis Based on Most Commonly Involved Vessels With Respect to Their Size (The List Includes Most of the Disorders; However, It Is Not a Complete List)

Large vessel vasculitis
Giant cell arteritis
Takayasu arteritis
Cogan syndrome
Aortitis associated with spondyloarthropathies
Idiopathic aortitis
Medium vessel vasculitis
Polyarteritis nodosa
Kawasaki disease
Small vessel vasculitis
Goodpasture disease or anti–glomerular basement membrane disease
Cryoglobulinemic vasculitis
Cutaneous leukocytoclastic angiitis or hypersensitivity vasculitis
Henoch-Schönlein purpura
Drug-induced vasculitis
Vasculitis related to RA, SLE, and Sjögren
Medium and small vessel vasculitis
Primary angiitis of the central nervous system
Wegener granulomatosis or eosinophilic granulomatosis with polyangiitis
Microscopic polyangiitis
Churg-Strauss syndrome
Buerger disease
Vasculitis involving arteries and veins of various sizes
Behçet disease
Relapsing polychondritis

RA, rheumatoid arthritis; SLE, systemic lupus erythematosus.