Epidemiology of Neurovasculitis

David S. Younger, MD, MPH, MS[a,b,*]

KEYWORDS

• Global • Burden • Vasculitis

KEY POINTS

- Vasculitis is defined as inflammation of blood vessel walls for at least some time during the course of the disease and affects arteries and veins of varying calibers.
- The Chapel Hill Consensus Conferences have provided consensus on nosology and definitions for the commonest forms of adult-onset vasculitides.
- The Pediatric Rheumatology European Society and the European League against Rheumatism have proposed specific classification criteria for the commonest childhood vasculitides.
- Although not included in the 2013 Global Burden of Disease Study, adult and childhood vasculitides are a significant source of morbidity and mortality globally.
- Management relies on the use of immunosuppressant and immune modulatory therapy.

CLASSIFICATION AND NOSOLOGY

Vasculitis is defined as inflammation of blood vessel walls for at least some time during the course of the disease and affects arteries and veins of varying calibers. Two Chapel Hill Consensus Conferences (CHCC), one in 1994[1] and the other in 2012,[2] provided consensus on nosology and definitions for the commonest forms of vasculitis. The revised CHCC nomenclature serves as a guide for the categorization of diverse forms of vasculitis based on the vessels involved and provides a scheme for the neurologic aspects thereof (**Box 1**). Large vessel vasculitis (LVV), including giant cell arteritis (GCA) and Takayasu arteritis (TAK), affects the aorta, its major branches, and analogous veins. Medium vessel vasculitis (MVV), inclusive of polyarteritis nodosa (PAN) and Kawasaki disease (KD), involves main visceral arteries and veins and initial branches. The category of small vessel vasculitis (SVV) recognizes involvement of intraparenchymal arteries, arterioles, capillaries, veins, and venules, with a disease mechanism related to antineutrophil cytoplasmic antibody (ANCA) and immune

The author has nothing to disclose.

[a] Division of Neuroepidemiology, Department of Neurology, New York University School of Medicine, New York, NY, USA; [b] College of Global Public Health, New York University, New York, NY, USA

* Corresponding author. 333 East 34th Street, 1J, New York, NY 10016.

E-mail address: david.younger@nyumc.org

Neurol Clin 34 (2016) 887–917

http://dx.doi.org/10.1016/j.ncl.2016.06.006 **neurologic.theclinics.com**

0733-8619/16/$ – see front matter

Box 1
Classification of primary systemic vasculitides

Large vessel vasculitis

Giant cell arteritis

Takayasu arteritis

Medium vessel vasculitis

Polyarteritis nodosa

Kawasaki disease

Small vessel vasculitis

ANCA-associated vasculitis
- Microscopic polyangiitis
- Granulomatosis with polyangiitis (Wegener)
- Eosinophilic granulomatosis with polyangiitis (Churg-Strauss)

Immune-complex vasculitis
- Cryoglobulinemia
- IgA vasculitis (Henoch-Schönlein)
- Hypocomplementemic urticarial vasculitis (anti-C1q)

Variable vessel vasculitis

Behçet disease

Cogan syndrome

Single-organ vasculitis

Primary CNS vasculitis

Idiopathic aortitis (IgG4)

complexes. The category of ANCA-associated vasculitis (AAV) includes granulomatosis with polyangiitis (GPA), Wegener granulomatosis (WG) type, eosinophilic granulomatosis with polyangiitis (EGPA) Churg-Strauss syndrome, and microscopic polyangiitis (MPA) (microscopic polyarteritis), whereas vasculitic disorders associated with immune complexes include immunoglobulin A (IgA) vasculitis (IgAV) (Henoch-Schönlein purpura [HSP]), cryglobulinemic vasculitis (CV), and hypocomplementemia urticarial vasculitis (HUV) associated with C1q antibodies. Vasculitis without a predominant vessel size and caliber, respectively, from small to large, involving arteries, veins, and capillaries, comprises the category of variable vessel vasculitis (VVV), characteristic of Behçet disease (BD) and Cogan syndrome (CS). The category of vasculitis associated with systemic disease includes vasculitis associated with rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE) and other connective tissue disorders, wherein the vasculitic process is secondary to or associated with the underlying systemic disorder. There is a category of vasculitis associated with a probable specific cause, such as substance abuse and infection designated by the specific vasculitic disorder with a prefix to denote the causative agent. The category of single-organ vasculitis (SOV) involves arteries or veins of any size in a single organ without features to indicate that it is a limited expression of a systemic vasculitis characterized by primary central nervous system (CNS) vasculitis, nonsystemic peripheral nerve vasculitis (PNV), and isolated aortitis.

Recognizing that certain forms of vasculitis are more common in childhood and that some vasculitides display different disease courses compared with adult forms,[3] the