ScienceDirect

Available online at www.indianjrheumatol.com and www.sciencedirect.com

Review Article

Classification of vasculitis: From historical controversies to present day pragmatic consensus

Richard A. Watts [a,b,*]

[a]Ipswich Hospital NHS Trust, Ipswich, [b]Norwich Medical School, University of East Anglia, Norwich, UK

ARTICLE INFO

Article history:
Received 30 May 2014
Accepted 3 June 2014
Available online 3 July 2014

Keywords:
ANCA
Granulomatosis with polyangiitis, microscopic polyangiitis

ABSTRACT

The classification of the systemic vasculitides has been controversial for several decades. However, over the past twenty years there have been several major developments, which means that there is pragmatic consensus regarding classification. These include the American College of Rheumatology criteria first published in 1990, and the Chapel Hill Consensus Conference definitions originally promulgated in 1994, but revised and extended in 2012. More recently the classical division of the ANCA vasculitides using clinical phenotype has come under scrutiny with evidence from epidemiological, genetic and outcome studies that perhaps these conditions should be classified on the basis of ANCA specificity into PR3-ANCA positive and MPO-ANCA positive groups. There remains, however, a major need for validated classification and diagnostic criteria, a need which hopefully the DCVAS project will address.

1. Introduction

The systemic vasculitides are a group of related disorders characterized by blood vessel inflammation leading to tissue or end organ injury. The classification of vasculitis has been controversial for many years. The purpose of this review is to describe the current classification of the vasculitides in their historical context and provide some indicators of future developments.

The initial descriptions of vasculitis were isolated case reports starting with William Heberden in the 1760s, Henoch, Schönlein, Kussamul and Meier in the nineteenth century.[1] In the twentieth century the first descriptions of granulomatosis with polyangiitis (Wegener's granulomatosis), eosinophilic granulomatosis with polyangiitis (Churg–Strauss syndrome), Takayasu arteritis and giant cell arteritis, Kawasaki disease were published. Wegener's granulomatosis, Churg–-Strauss syndrome and Henoch–Schönlein purpura were known by their eponyms until the present decade.

The initial classification of vasculitis was by Zeek in the 1950, who recognized five types of vasculitis based on vessel size.[2] Zeek reviewed the literature relating to vasculitis and periarteritis nodosa and used the generic term 'necrotizing angiitis' to indicate the specific damage to the blood vessel wall rather than the presence of inflammation alone; she classified these into five distinct entities: (i) hypersensitivity angiitis, (ii) allergic granulomatous angiitis, (iii) rheumatic arteritis, (iv) periarteritis nodosa, and (v) temporal arteritis. Most modern classifications are based on Zeek's work, which essentially combined histological changes and clinical

* Ipswich Hospital NHS Trust, Heath Road, Ipswich IP4 5PD, UK. Tel.: +44 1473702131.
E-mail address: richard.watts@ipswichhospital.nhs.uk.
http://dx.doi.org/10.1016/j.injr.2014.06.001

Table 1 – Chapel Hill Definitions as revised in 2011 (from Jennette et al, 2013).

CHCC 2011 name	CHCC 2011 definition
Large vessel vasculitis	Vasculitis affecting large arteries more often than other vasculitides. Large arteries are the aorta and its major branches. Any size artery may be affected.
Takayasu arteritis	Arteritis, often granulomatous, predominantly affecting the aorta and/or its major branches. Onset usually in patients younger than 50.
Giant cell arteritis	Arteritis, often granulomatous, usually affecting the aorta and/or its major branches, with a predilection for the branches of the carotid artery. Often involves the temporal artery. Onset usually in patients older than 50 and often associated with polymyalgia rheumatica.
Medium vessel vasculitis	Vasculitis predominantly affecting medium-sized arteries defined as the main visceral arteries and their branches. Any size artery may be affected. Inflammatory aneurysms and stenoses are common.
Polyarteritis nodosa	Necrotizing arteritis of medium-sized or small arteries without glomerulonephritis or vasculitis in arterioles, capillaries, or venules; and not associated with ANCA.
Kawasaki disease	Arteritis associated with the mucocutaneous lymph node syndrome and predominantly affecting medium-sized and small arteries. Coronary arteries are often involved. Aorta and large arteries may be involved. Usually occurs in infants and young children.
Small vessel vasculitis	Vasculitis predominantly affecting small vessels, defined as small intraparenchymal arteries, arterioles, capillaries and venules. Medium-sized arteries and veins may be affected.
ANCA associated vasculitis	Necrotizing vasculitis, with few or no immune deposits, predominantly affecting small vessels (i.e., capillaries, venules, arterioles and small arteries), associated with MPO-ANCA or PR3-ANCA. Not all patients have ANCA. Add a prefix indicating ANCA reactivity, e.g. PR3-ANCA, MPO-ANCA, ANCA negative.
Microscopic polyangiitis	Necrotizing vasculitis, with few or no immune deposits, predominantly affecting small vessels (i.e., capillaries, venules, or arterioles). Necrotizing arteritis involving small and medium-sized arteries may be present. Necrotizing glomerulonephritis is very common. Pulmonary capillaritis often occurs. Granulomatous inflammation is absent.
Granulomatosis with polyangiitis (Wegener's)	Necrotizing granulomatous inflammation usually involving the upper and lower respiratory tract, and necrotizing vasculitis affecting predominantly small to medium-sized vessels (e.g., capillaries, venules, arterioles, arteries and veins). Necrotizing glomerulonephritis is common.
Eosinophilic granulomatosis with polyangiitis (Churg–Strauss)	Eosinophil-rich and necrotizing granulomatous inflammation often involving the respiratory tract, and necrotizing vasculitis predominantly affecting small to medium-sized vessels, and associated with asthma and eosinophilia. ANCA is most frequent when glomerulonephritis is present.
Immune complex small vessel vasculitis	Vasculitis with moderate to marked vessel wall deposits of immunoglobulin and/or complement components predominantly affecting small vessels (i.e., capillaries, venules, arterioles and small arteries). Glomerulonephritis is frequent.
Anti-GBM disease	Vasculitis affecting glomerular capillaries, pulmonary capillaries, or both, with basement membrane deposition of anti-basement membrane autoantibodies. Lung involvement causes pulmonary haemorrhage, and renal involvement causes glomerulonephritis with necrosis and crescents.
Cryoglobulinemic vasculitis	Vasculitis with cryoglobulin immune deposits affecting small vessels (predominantly capillaries, venules, or arterioles) and associated with cryoglobulins in serum. Skin and glomeruli are often involved.
IgA Vasculitis (Henoch–Schönlein)	Vasculitis, with IgA1-dominant immune deposits, affecting small vessels (predominantly capillaries, venules, or arterioles). Often involves skin and gut, and frequently causes arthritis. Glomerulonephritis indistinguishable from IgA nephropathy may occur.
Hypocomplementemic urticarial vasculitis (anti-C1q vasculitis)	Vasculitis accompanied by urticaria and hypocomplementemia affecting small vessels (i.e., capillaries, venules, or arterioles), and associated with anti-C1q antibodies. Glomerulonephritis, arthritis, obstructive pulmonary disease, and ocular inflammation are common.

(*continued on next page*)