Supplementary material

Table S1. AAV and their mimickers, categorised according to associated autoimmune features (ANCA-AI) or lack of (ANCA-O)
Abbreviations: AAV, ANCA-associated vasculitis; APLS, antiphospholipid syndrome; CNS, central nervous system; CTD, connective tissue disease; GPA, granulomatosis with polyangiitis; IBD, inflammatory bowel disease; MPA, microscopic polyangiitis; SLE, systemic lupus erythematous.

AAV n=49	ANCA-AI n=99		ANCA-O n=140
	Non-AAV vasculitides n=11	Other auto-immune disorders n=88	
GPA, n=16 MPA, n=33	Giant-cell arteritis, n=3 Cryoglobulinemic vasculitis, n=3 Hypocomplementemic urticarial vasculitis, n=2 Behçet's disease, n=2 IgA-vasculitis, n=1	IBD, n=16 SLE, n=14 Unclassified inflammatory Rheumatism, n=10 Rheumatoid arthritis, n=9 Systemic sclerosis, n=7 Idiopathic pulmonary fibrosis, n=5 Primary Sjögren syndrome, n=5 Unclassified CTD, n=4 Pernicious anemia, n=3 Myasthenia gravis, n=2 APLS, n=2 Other, n=11	Infectious disorders, n=38 Myocardial infarction and/or stroke, n=25 Hematological disorders, n=13 Non-inflammatory rheumatic disorders, n=12 Deep vein thrombosis and/or pulmonary embolism, n=9 Asthma, n=7 Cancer, n=6 Alcoholic liver cirrhosis, n=6 Other disorders, n=24

Table S2. Features of subjects presenting with small-vessel vasculitides and positive ANCA, in relation to 2022 ACR/EULAR classification criteria and definitive diagnosis.
Abbreviations: AAV, ANCA-associated vasculitis; APLS, antiphospholipid syndrome; CNS, central nervous system; CTD, connective tissue disease; GPA, granulomatosis with polyangiitis; IBD, inflammatory bowel disease; MPA, microscopic polyangiitis; SLE, systemic lupus erythematous; yo, years old.

Demographics	Comorbid features	Clinician's diagnosis	Clinical features	Lung involvement	Kidney involvement	Other organ involvement	ANCA type, titre	Histopathology	2022 ACR/EULAR classification criteria	Definitive diagnosis
Male, 46 yo	None	Behçet's disease	Relapsing pericarditis	None	None	Polyarthralgia Relapsing bipolar aphtosis	PR3-ANCA, 36 U/mL	Digestive biopsy without IBD	GPA ACR/EULAR scoring: 5	Behçet's disease
Male, 41 yo	Smoking	Behcet's disease	Posterior uveitis with hemorrhagic retinal vasculitis	None	None	Inflammatory brainstem syndrome Relapsing oral aphtosis Folliculitis	PR3-ANCA, 33 U/mL	Not performed	GPA ACR/EULAR scoring: 5	Behcet's disease
Male, 88 yo	B-cell lymphoma	Cryoglobulinemic vasculitis	Purpura	None	Yes Glomerular proteinuria	Multiple lacunar strokes Splenic ischemia	PR3-ANCA 102 U/mL	Not performed	GPA ACR/EULAR scoring: 5	Cryoglobulinemic vasculitis
Female, 71 yo	Angioedema Hypertension	Hypocomplementemic urticarial vasculitis	Mononeuropathy simplex	None	None	Muscle pain Urticaria	PR3-ANCA, 36 U/mL	Small vessel vasculitis, C3 deposit, without necrosis nor granuloma	GPA ACR/EULAR scoring: 5	Hypocomplementemic urticarial vasculitis
Female, 30 yo	Angioedema Urticaria	Hypocomplementemic urticarial vasculitis	Purpura	None	None	Urticaria Relapsing angioedema Arthralgia	PR3-ANCA, 40 U/mL	Small vessel vasculitis, C3 deposit, without necrosis nor granuloma	GPA ACR/EULAR scoring: 5	Hypocomplementemic urticarial vasculitis
Female, 50 yo	None	IgA vasculitis	Purpura	None	None	None	PR3-ANCA, 30 U/mL	Small vessel vasculitis without necrosis, granuloma, or deposit	GPA ACR/EULAR scoring: 5	Non-specific small-vessel vasculitis
Male, 50 yo	None	Cryoglobulinemic vasculitis	Muscle pain and purpura	None	None	None	PR3-ANCA, 31 U/mL	Small vessel vasculitis without necrosis nor granuloma	GPA ACR/EULAR scoring: 5	Cryoglobulinemic vasculitis
Male, 75 yo	Smoking Auricular fibrillation	Cryoglobulinemic vasculitis	Purpura	None	None	Arthralgia	PR3-ANCA, 22 U/mL	Small vessel vasculitis with immune complex deposit (IgM/C3)	No AAV ACR/EULAR scoring: 1	Cryoglobulinemic vasculitis