

An update on the general management approach to common vasculitides

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ABSTRACT

Primary systemic vasculitides (PSV) are multisystem diseases associated with high morbidity and mortality, particularly if not treated in a timely manner. In recent decades, clinical trials have delivered considerable evidence to underpin optimal diagnostic and therapeutic approaches. This article provides a brief overview of PSV in adults, focusing on the latest updates and recommendations for the management of antineutrophil cytoplasmic antibody-associated vasculitis and giant cell arteritis.

Introduction

Primary systemic vasculitides (PSV) are a group of rare, chronic diseases characterised by inflammation of the blood vessels.¹ Vessels of any size in any organ can be affected. The disease spectrum ranges from mild, localised disease to potentially life- or organ-threatening manifestations. Due to their heterogeneity and rarity, the recognition and management of vasculitis poses a considerable challenge to many physicians. In this review, we begin with a general overview of vasculitis in adults, including classification and general management approaches. We then focus on two comparatively common diseases, antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) and giant cell arteritis (GCA), with the latest update on their management.

Types of vasculitis

Although this review focuses on PSV, the general approach to vasculitis requires consideration and exclusion of mimics and secondary vasculitis. Vasculitis may be mimicked by reactions to drugs, infections, malignancy or thromboembolic disease. Secondary vasculitis can develop in the context of inflammatory rheumatic diseases, such as rheumatoid arthritis and systemic lupus erythematosus. It is important to distinguish these from

PSV, as their management can differ completely; for example, immunosuppressive medication used for the treatment of presumed PSV may aggravate infectious diseases. Therefore, it is imperative to exclude other problems before making a diagnosis of PSV. Table 1 provides a summary of some mimics.^{2,3}

Currently, there are no validated diagnostic criteria for PSV. The Chapel Hill Consensus Conference (CHCC) in 2012 updated the definitions and nomenclature for vasculitis. Vasculitides are categorised primarily by vessel size, together with aetiology, pathogenesis, pathology, demographics and clinical features (Fig 1).¹ The ongoing Diagnostic and Classification Criteria in Vasculitis Study

Key points

Vasculitis should be suspected in any systemically ill patients with multiorgan dysfunctions, especially in patients with no conventional cardiovascular risk factors who presented with organ ischaemia symptoms.

The principles of managing antineutrophil cytoplasmic antibody-associated vasculitis are to induce and maintain disease remission using a combination of glucocorticoids and immunosuppressants.

Rituximab is an efficacious treatment option for remission induction and maintenance among patients with organ-threatening, life-threatening or relapsing disease. It is preferable when cyclophosphamide is contraindicated or fertility issue is a concern.

Colour Doppler ultrasound of temporal or axillary arteries has similar diagnostic values as temporal artery biopsy in giant cell arteritis.

Tocilizumab is a new therapeutic option for relapsing or refractory giant cell arteritis that dramatically reduces cumulative glucocorticoid exposure.

KEYWORDS: Vasculitis, ANCA, giant cell arteritis, tocilizumab, rituximab

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Table 1. Vasculitis mimics

Vasculitis mimics	Example
Infections	Infective endocarditis Bacteria eg meningococcaemia or gonococcaemia Spirochetes Fungi Mycobacteria Viruses
Drugs/therapy	Ergotamine Cocaine Phenylpropanolamine Warfarin Radiation therapy
Malignancy	Leukaemia cutis Plasma cell dyscrasia Metastatic carcinoma Paraneoplastic syndrome Lymphoma, especially involving nasal or paranasal cavity
Thromboembolic disorders	Atrial myxoma Cholesterol emboli from an atheroma Antiphospholipid syndrome Thrombotic thrombocytopenic purpura Sickle cell disease Procoagulant states Calciphylaxis
Other vasculopathy	Fibromuscular dysplasia Amyloid angiopathy Neurofibromatosis type I Coarctation of the aorta Chronic ergotism Scurvy Sweet's syndrome Köhlmeier–Degos disease

(DCVAS) Project is anticipated to develop and validate diagnostic criteria and improve classification criteria for PSV.⁴

Approach to a patient with possible PSV

Diagnosis of PSV is based on a combination of clinical, laboratory, histopathological and radiological features. There are several clinical manifestations that should raise suspicion of vasculitis. The most common are constitutional symptoms such as fever, malaise, unintentional weight loss and anorexia. These features are not unique to PSV; chronic infection, malignancy and autoimmune connective tissue diseases may present similarly. Symptoms indicating organ ischaemia (eg limb claudication, digital ulcer, stroke or myocardial infarction) are more specific, especially if they occur in young patients without conventional cardiovascular risk factors. Vasculitis should be suspected in constitutionally ill patients with multiorgan dysfunctions.

A systematic and comprehensive history (Table 2) and examination (Fig 2) are essential. Laboratory tests are crucial for excluding mimics or secondary vasculitis and determining extent of organ involvement. The more specific serological, histopathological

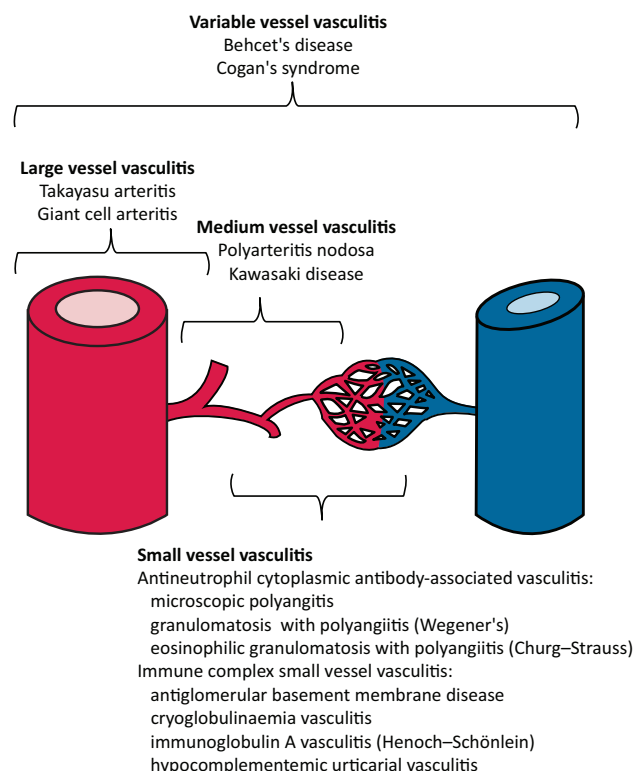


Fig 1. Types of vasculitis categorised according to the distribution of major vessel involvement.

and angiographic investigations are helpful in categorising PSV (Table 3).

AAV

AAV is characterised by necrotising vasculitis predominantly affecting small vessels, with few or no immune deposits. It is usually associated with ANCA specific for myeloperoxidase (MPO-ANCA) or proteinase 3 (PR3-ANCA) and commonly involves the respiratory tract and kidneys.¹

The estimated prevalence of AAV in England is 14.6/100,000 population. The median age of these patients is 65.3 years with approximately equal gender ratio. The prevalence of granulomatosis with polyangiitis (GPA) is 9.97/100,000, microscopic polyangiitis (MPA) is 1.40/100,000 and eosinophilic granulomatosis with polyangiitis (EGPA) is 3.18/100,000.⁵

Management of AAV

The British Society for Rheumatology (BSR) and British Health Professionals in Rheumatology (BHPR) published revised guidelines for the management of adults with AAV in 2014.⁶ The European League Against Rheumatism (EULAR) and European Renal Association – European Dialysis and Transplant Association (ERA-EDTA) and Canadian Vasculitis Research Network (CanVasc) updated similar recommendations in 2016.^{7,8} The Brazilian Society of Rheumatology (SBR) had published recommendations on the induction therapy of AAV in 2017.⁹

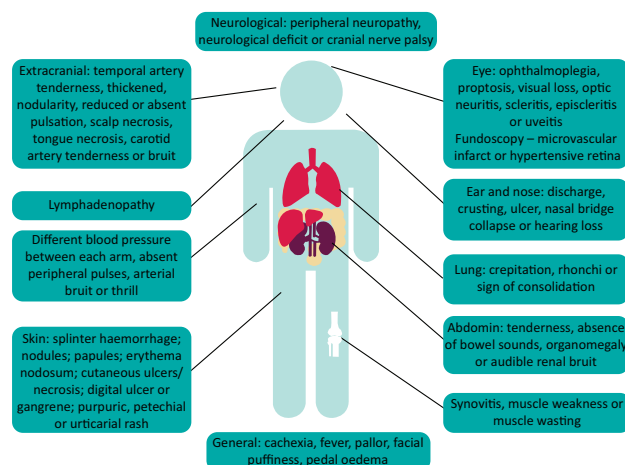
These latest guidelines and recommendations have simplified the stratification of AAV disease severity into non-severe and

Table 2. Summary of clinical history for patient with probable vasculitides

History	Descriptions
General	Onset of symptoms, progression Preceding infection, illness or trauma Previous medical illness Previous drug usage Family history of autoimmune rheumatic disease or chronic hepatitis infection Travel history Occupational history Recreational drug consumption or high-risk behaviour
Constitutional symptoms	Fever, sweats Weight loss, anorexia Malaise, fatigue
Skin, extremities	New, persistent skin rashes Raynaud's phenomenon Digital ulcer Cutaneous ulcer
Musculoskeletal	Myalgia, muscle weakness, stiffness Arthralgia
Ear, nose and throat	Persistent, recurrent, blood-stained nasal discharge or epistaxis Nasal crusting Sinusitis Nasal bridge collapse Ear discharge Hearing impairment
Eye	Painful, red eye Sight loss, changes to colour vision Diplopia
Nervous system	Wrist drop, foot drop Stroke Seizure Headache
Extracranial	Scalp tenderness / hyperaesthesia Jaw claudication, tongue claudication
Pulmonary	Asthma Breathlessness Haemoptysis
Cardiovascular	Limb claudication Chest pain
Gastrointestinal	Abdominal pain Diarrhoea Haematemesis, melaena, haematochezia
Renal	Frothy urine, haematuria Hypertension
Genitourinary	Scrotal pain

severe groups, namely, 'non-organ threatening disease' and 'organ- or life-threatening disease'.

The general principles of managing AAV are to induce and maintain disease remission using the least cytotoxic medications. Latest BSR/BHPR and EULAR/ERA-EDTA recommendations also

**Fig 2. Summary of possible clinical findings in patients with primary systemic vasculitides.**

highlight the importance of patient education and participation in disease management.

Remission induction

Organ- or life-threatening AAV

Latest guidelines advocate combination of either cyclophosphamide (CYC) or rituximab (RTX) with glucocorticoids in inducing remission in patients with organ- or life-threatening AAV.

In the CYCLOPS study, pulsed CYC was associated with fewer leukopenia episodes and lower cumulative dose than oral regimens and thus deemed to have lower toxicity.¹⁰ Although pulsed CYC was associated with a higher relapse risk than oral CYC in an extended follow-up study, it was not associated with increased mortality or long-term morbidity.¹¹ Oral CYC is therefore no longer a preferred choice for induction therapy in BSR/BHPR and EULAR/ERA-EDTA recommendations, but is still considered as an option in CanVasc and SBR recommendations.^{6–9} BSR/BHPR recommends intravenous CYC 15 mg/kg at 2-week intervals for first three pulses and thereafter at 3-week intervals, reduced for age and renal function. Each course should be a minimum of 3 months but not exceed 6 months.⁶

RTX has emerged as a new remission induction option for severe AAV after it was demonstrated to be non-inferior to CYC in the RAVE and RITUXVAS trials.^{12,13} The RAVE trial, which recruited younger AAV patients with less severe renal disease, demonstrated similar rates of adverse events compared to oral CYC.¹² RITUXVAS trial, using intravenous CYC as a comparator, showed equivalent sustained-remission and infection rates.¹³ The RITUXVAS 2-year follow-up study showed comparable disease remission, adverse events and mortality.¹⁴ This B-cell-depleting anti-CD20 monoclonal antibody is now recognised as a first-line alternative by all guidelines, with minor difference in restrictions on its use. The UK National Institute for Health and Care Excellence (NICE) considers RTX to be cost effective in treating AAV.¹⁵ The BSR/BHPR gives preference to RTX when CYC avoidance is needed, eg pre-existing high cumulative dose of CYC, infection, intolerance, uroepithelial malignancy or infertility concern.⁶ Dosing regimens are either 375 mg/m²/week for 4 weeks or two infusions of 1 g, 2 weeks apart.^{6,16} The EULAR/ERA-EDTA emphasised the lack of strong data on the use of RTX for EGPA.⁷

Table 3. Laboratory investigations useful in the evaluation of suspected vasculitides

Preliminary investigations	More specific investigations
Full blood count	Perinuclear ANCA (pANCA) and cytoplasmic ANCA (cANCA)
Urinalysis, urine protein:creatinine ratio	Antiproteinase 3 (antiPR3) and antimyeloperoxidase (antiMPO) antibody
Renal profile	Antiglomerular basement membrane antibody
Liver function test	Serum cryoglobulins
Creatinine kinase	Immunoglobulin E level
Fasting lipid profile, fasting blood sugar	Appropriate tissue biopsy
C-reactive protein, erythrocyte sedimentation rate	Bronchoscopy, bronchoalveolar lavage
Procalcitonin level	Angiographic examinations
Blood culture	Magnetic resonance angiography / magnetic resonance imaging
Hepatitis B and hepatitis C screening	Positron emission tomography
HIV test	Nerve conduction studies or electromyography
Other serology such as parvovirus B19, cytomegalovirus or specific fungal serology test	
Antinuclear antibody, antidouble stranded deoxyribonucleic acid (anti-dsDNA), extractable nuclear antigen	
Antiphospholipid antibodies	
Rheumatoid factor	
Complements level	
Urine toxicology screening	
Chest X-ray or computed tomography of the thorax	
Sinus X-ray or computed tomography	
Electrocardiography, echocardiography	
Serum protein electrophoresis, serum free light chains, urine Bence Jones protein	

High-dose glucocorticoids at 1 mg/kg/day or equivalent remains the cornerstone of remission induction therapy. The BSR/BHPR recommends intravenous methylprednisolone 250–500 mg to be given prior to or with the first two pulses of CYC.⁶ CanVasc and SBR recommend intravenous methylprednisolone 500–1,000 mg for 1–3 days as initial part of induction therapy in severe AAV.^{8,9} All guidelines acknowledge little data supporting the benefits of intravenous methylprednisolone in severe AAV. The BSR/BHPR and EULAR/DRA-EDTA recommend tapering prednisolone to 15 mg or 7.5–10 mg at 12 weeks, respectively. Detailed prednisolone-tapering protocols have been outlined in CanVasc and SBR recommendations.^{8,9}

Newer therapies continue to emerge. The ADVOCATE trial investigated avacopan (a novel orally-administered selective antagonist of complement fragment C5a receptor (C5aR)) against prednisolone for remission induction, each in combination with CYC or RTX, followed by azathioprine for maintenance. The avacopan combination was non-inferior to prednisolone combination at week 26 and superior in sustained remission at week 52. A remarkable reduction in glucocorticoid-related toxicity was observed in the avacopan arm.¹⁷

Severe organ-/life-threatening AAV

The BSR/BHPR, EULAR/ERA-EDTA and SBR recommend consideration of plasma exchange (PLEX) for patients with new or relapsing AAV and severe renal failure (creatinine >500 µmol/L) or severe diffuse alveolar haemorrhage, in addition to pulsed CYC and glucocorticoids.^{6,7,9} CanVasc recommendations restrict PLEX as adjuvant to patients with severe AAV refractory to standard remission induction therapy.⁸ More data are needed

on PLEX efficacy. In the MEPEX trial, PLEX improved rate of renal recovery at 3 months when compared with intravenous methylprednisolone, while patient survival and severe adverse event rates were similar at 12 months.¹⁸ However PEXIVAS (a factorial trial design comparing PLEX vs no PLEX among patients with severe AAV receiving CYC or RTX) did not show adjuvant PLEX to reduce death or end-stage renal failure after up to 7 years of follow-up. Results did not support a treatment effect for the 191 participants with pulmonary haemorrhage. The design also allowed comparison of reduced vs standard dose glucocorticoid regimens; outcomes were non-inferior when using 50% reduced glucocorticoids dose.¹⁹

Localised AAV

Mycophenolate mofetil (MMF) or methotrexate (MTX) in combination with glucocorticoids are recommended for remission induction in AAV patients without evidence of organ damage. Long-term data from the NORAM study showed that MTX was safe and as effective as oral CYC in achieving remission in non-organ-threatening AAV, but the median time to relapse and cumulative relapse-free survival were shorter in MTX-treated patients.²⁰ In the MYCYC trial, MMF was non-inferior to pulsed CYC for remission induction in patients with AAV but resulted in higher relapse rate, especially in those with PR3-ANCA positivity.²¹ Regarding long-term CYC toxicity, both BSR/BHPR and EULAR/ERA-EDTA have phased out CYC, instead recommending a combination of glucocorticoids with either MTX (up to 25–30 mg once per week) or MMF (up to 3 g/day) for localised AAV.^{6,7} CanVasc and SBR recommendations advocate only combination of glucocorticoids and MTX.^{8,9} Generally, MTX dose reduction is

required in renal impairment and MMF is preferred over MTX in moderate or severe renal impairment.^{6,8,9}

Refractory disease

Treatment for refractory disease remains challenging and should be managed at expert centres. BSR/BHPR and CanVasc favour use of RTX in those who fail to achieve remission with CYC and glucocorticoids.^{6,8} EULAR/ERA-EDTA advocates switching from CYC to RTX or vice versa.⁷ Adjunctive intravenous immunoglobulin has been suggested by BSR/BHPR, EULAR/ERA-EDTA and CanVasc for patients who have refractory disease or persistent low activity.^{6–8} BSR/BHPR and CanVasc recommend mepolizumab (anti-interleukin-5 monoclonal antibody) for refractory EGPA.^{6,8}

Remission maintenance

Once remission is achieved, low-dose glucocorticoids combined with azathioprine, MTX, MMF, leflunomide or RTX are recommended for maintenance by BSR/BHPR, EULAR/ERA-EDTA and CanVasc, with various preferences.

RTX was superior to azathioprine in maintaining remission (after CYC induction) for AAV in the MAINRITSAN trial, where more patients had sustained remission at month 28 with comparable safety.²² The advantage of RTX over azathioprine in preventing relapse continued at 60 months' follow-up.²³ The RITAZAREM trial demonstrated RTX to be superior to azathioprine in achieving sustained-remission among patients with relapsing disease at baseline and given RTX as induction therapy.²⁴ The cost of RTX can be prohibitive. BSR released consensus guidelines on the use of RTX for maintenance in new and relapsing AAV following RTX or CYC induction. They advocate the fixed interval dosing, either 500 mg or 1,000 mg administered every 6 months for a period of 2 years.²⁵

Azathioprine, the standard-of-care therapy for remission maintenance, was superior to MMF in the IMPROVE study.²⁰ Although leflunomide was superior to MTX, it is preferred as a second-line treatment due to its adverse effects profile.^{6–8,26}

BSR/BHPR and EULAR/ERA-EDTA recommend at least 24 months of remission maintenance therapy, whereas CanVasc recommends 18 months.^{6–8} Some experts advocate 36 months in patients who are PR3-ANCA positive.⁷ In patients who relapse after a prior course of RTX maintenance, with persistent ANCA positivity, or where the effect of relapse would be organ- or life-threatening despite 2 years of RTX maintenance therapy, an extended duration of RTX up to 5 years could be considered.²⁵ If sustained remission is achieved for at least 1 year on maintenance therapy, BSR/BHPR suggests tapering glucocorticoids, followed by withdrawing immunosuppressants 6 months later.⁶

Relapse of AAV

Latest BSR/BHPR guidelines advocate the use of RTX or further course of CYC in major relapse.⁶ The EULAR/ERA-EDTA and CanVasc more strongly recommend RTX over CYC, given concerned over cumulative CYC dose.^{7,8} RAVE long-term follow-up data showed that those with severe relapsing disease treated with RTX had superior remission maintenance and fewer severe relapses at 6 and 12 months.²⁷ BSR/BHPR, EULAR/ERA-EDTA and CanVasc recommend optimisation or modification of remission maintenance agent and increase glucocorticoid dose for minor relapse, without specifying choice of immunosuppressant and duration of therapy.^{6–8}

Long-term follow-up and monitoring

Long-term follow-up and monitoring for disease relapse and its complications is imperative since AAV is a chronic relapsing disease. The role of biomarkers (eg ANCA, PR3-ANCA, MPO-ANCA or CD19) in predicting disease relapse requires further research. Regular monitoring and assessment for infections, haemorrhagic cystitis, cancer (ie bladder or cervical), infertility, cardiovascular and thromboembolic risks, and osteoporosis is warranted.

GCA

GCA is a large vessel vasculitis, primarily affecting the aorta and its extracranial branches, with a predilection for the temporal artery.¹ It commonly occurs in patients older than 50 years with highest incidence among people that are 70–79 years old. The incidence of GCA ranges between 6/100,000 in northern Italy to 22/100,000 in the UK.^{28–31}

Recognition and diagnosis of GCA

EULAR and BSR updated their recommendations on diagnosis and treatment of GCA in 2018 and 2020, respectively.^{32,33}

GCA is distinguished into three phenotypes: cranial GCA, large vessel GCA (LV-GCA) and isolated polymyalgia rheumatica (PMR). Patients aged over 50 years, presenting with abrupt-onset headache, scalp tenderness, visual loss, jaw claudication, limb claudication, polymyalgia or constitutional symptoms should raise suspicion of GCA. Several mimics of GCA and PMR (Table 4) should be excluded via thorough history, physical examination, and investigations (Tables 2 and 3; Fig 2).^{2,34,35}

High erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) are hallmarks of GCA. A small proportion of patients with GCA (<4%) have normal ESR and CRP.^{36,37} Normal acute-phase reactants make a diagnosis of GCA very unlikely but do not rule it out.

Confirmatory diagnostic test for GCA

The latest recommendations advocate utilising either temporal artery biopsy (TAB) or ultrasound of the temporal and/or axillary arteries to confirm diagnosis of GCA.^{32,33,38}

Table 4. Mimics of giant cell arteritis and polymyalgia rheumatica

Mimics of giant cell arteritis	Mimics of polymyalgia rheumatica
Sinusitis	Elderly-onset rheumatoid arthritis
Dental or temporo-mandibular local disease	Osteoarthritis
Non-arteritic anterior ischaemic optic neuropathy	Fibromyalgia
Subacute thyroiditis	Shoulder bursitis / tendonitis
Chronic infections	Cervical spondylosis
Trigeminal neuralgia	Ankylosing spondylitis
Malignancy	Statin-induced myositis
atherosclerotic cardiovascular disease	Systemic lupus erythematosus
	Hypothyroidism
	Chronic infections
	Polymyositis
	Malignancy (eg multiple myeloma)
	Amyloidosis

TAB has been regarded as the gold standard to diagnose GCA for decades. In one meta-analysis of studies investigating TAB positivity among patients satisfying the 1990 American College of Rheumatology classification criteria of GCA, the estimated sensitivity of TAB for GCA diagnosis was 77%, with high between-study heterogeneity.^{39,40}

Vascular imaging as an emerging diagnostic tool for GCA

High-resolution colour Doppler ultrasound (CDS) of the temporal or axillary arteries, showing concentric hypoechoic mural thickening 'halo sign', and 'compression sign' where the hypoechoic wall swelling is non-compressible upon application of pressure with ultrasound probe, are the most important sonographic abnormalities for GCA.⁴¹ These findings are interpreted as inflammatory vessel wall oedema.⁴² Using CDS, LV-GCA was observed in 53% of one GCA cohort, where circumferential, homogeneous, hypoechogenic mural thickening was most frequently observed in the axillary arteries.⁴³ In the TABUL study, CDS of temporal and axillary arteries had a higher sensitivity than TAB (54% vs 39%) but lower specificity (81% vs 100%) in diagnosing GCA.⁴⁴ In short, CDS of temporal or axillary arteries, best done within 1 week of glucocorticoid initiation, is valuable to confirm GCA in high probability cases.³²

High-resolution magnetic resonance imaging (MRI) of cranial arteries can be considered as an alternative to CDS for GCA diagnosis.^{32,33,38} MRI can detect vessel wall oedema and contrast enhancement, signifying vessel inflammation. Using TAB as a reference standard, MRI of the cranial arteries had a sensitivity of 94% and specificity of 78% in diagnosing GCA. The corresponding negative predictive value of MRI cranial arteries was 98%, thus potentially useful for ruling out GCA.⁴⁵

Use of imaging to evaluate involvement of the aorta and its proximal branches in GCA is limited due to lack of direct evidence. Most of the published evidence is extrapolated from other diseases, such as Takayasu arteritis.³⁸

Computed tomography angiography of the aorta and its major branches can reveal vessel wall thickening with contrast enhancement, and structural lesions (stenosis, dilatation or aneurysms).⁴⁶ Contrast-enhanced magnetic resonance angiography was found to reliably identify aortitis in large-vessel vasculitides but changes were very sensitive to glucocorticoids.⁴⁷ Positron emission tomography – computed tomography (PET-CT) can demonstrate inflammatory cell infiltration of the aorta and its branches.⁴⁸ It has the advantage of being additionally able to detect malignancy or occult infections.³⁸

Conventional angiography is not recommended for the diagnosis of GCA because safer modalities available. It is mainly used for vascular intervention such as angioplasty or stenting.³⁸

Management of GCA

Glucocorticoids remain the mainstay of treatment for GCA. Oral glucocorticoids at 1 mg/kg/day should be initiated immediately while referring patients for urgent specialist evaluation.^{32,33} High-dose glucocorticoids are continued until resolution of symptoms and normalisation of inflammatory markers. Despite the lack of data regarding the optimal duration of glucocorticoid therapy, BSR recommends to taper over 12–18 months, whereas EULAR advocates 24 months or more.^{32,33}

A substantial number of patients with GCA have refractory or relapsing disease when glucocorticoids are tapered. They are conventionally treated with glucocorticoid dose escalation, which subjects them to excessive cumulative adverse effects of glucocorticoids. MTX might be combined with glucocorticoid taper for those at high risk of glucocorticoid toxicity or who relapse, however the evidence remains equivocal.⁴⁹

Tocilizumab, an interleukin-6 inhibitor, was approved for GCA by US and European regulatory authorities in 2017. In the GiACTA trial, 1 year of weekly tocilizumab plus 26-week prednisolone taper (56%) had higher rate of sustained remission at 1 year, compared with placebo and 26-week prednisolone taper (14%) or 52-week taper (18%; $p < 0.001$).⁵⁰ Results from 2-year long-term extension of GiACTA trial showed 47% of patients in weekly tocilizumab plus 26-week prednisolone taper maintained clinical remission, with similar safety profile compared with placebo and prednisolone taper. This arm of patients had the lowest cumulative glucocorticoid dose over the 3-year study.⁵¹ Subcutaneous tocilizumab 162 mg weekly has been approved by NHS England for patients with relapsing or refractory GCA for 1 year.⁵²

Contrary to previous recommendations, anticoagulant, antiplatelet or cholesterol-lowering agents are now not routinely recommended for treatment of GCA unless indicated.^{32,33}

Summary

Significant advancement has been made in the management of common vasculitides, especially for AAV and GCA. Diagnosis of PSV depends largely on excluding mimics and secondary vasculitis, recognising a combination of clinical, laboratory, histopathological and radiological features. Validated diagnostic criteria are much anticipated.

Intravenous CYC is favoured over oral CYC for AAV due to lower cumulative toxicity. RTX has provided a treatment revolution for AAV, both for remission induction and maintenance, among patients with organ-threatening, life-threatening or relapsing disease. There is still a lack of robust data to support the use of PLEX in imminent life-threatening AAV. More research is needed to guide treatment for refractory or relapsing AAV, and to identify biomarkers indicating a predisposition to poorly controlled disease.

CDS of temporal or axillary arteries has similar diagnostic value as TAB in diagnosis GCA. Tocilizumab provides an additional therapeutic option for relapsing or refractory GCA, lowering risk of glucocorticoid toxicity in susceptible patients. Further cost-effectiveness analyses are needed to support the extension commissioning of tocilizumab in GCA. ■

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