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REVIEW

Diagnosis and differential diagnosis of large-vessel vasculitides

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Abstract

There are no universally accepted diagnostic criteria for large-vessel vasculitides (LVV), including giant cell arteritis (GCA) and Takayasu arteritis (TAK). Currently, available classification criteria cannot be used for the diagnosis of GCA and TAK. Early diagnosis of these two diseases is quite challenging in clinical practice and may be accomplished only by combining the patient symptoms, physical examination findings, blood test results, imaging findings, and biopsy results, if available. Awareness of red flags which lead the clinician to investigate TAK in a young patient with persistent systemic inflammation is helpful for the early diagnosis. It should be noted that clinical presentation may be highly variable in a subgroup of GCA patients with predominant large-vessel involvement (LVI) and without prominent cranial symptoms. Imaging modalities are especially helpful for the diagnosis of this subgroup. Differential diagnosis between older patients with TAK and this subgroup of GCA patients presenting with LVI may be difficult. Various pathologies may mimic LVV either by causing systemic inflammation and constitutional symptoms, or by causing lumen narrowing with or without aneurysm formation in the aorta and its branches. Differential diagnosis of aortitis is crucial. Infectious aortitis including mycotic aneurysms due to septicemia or endocarditis, as well as causes such as syphilis and mycobacterial infections should always be excluded. On the other hand, the presence of non-infectious aortitis is not unique for TAK and GCA. It should be noted that aortitis, other large-vessel involvement or both, may occasionally be seen in various other autoimmune pathologies including ANCA-positive vasculitides, Behçet's disease, ankylosing spondylitis, sarcoidosis, and Sjögren's syndrome. Besides, aortitis may be idiopathic and isolated. Atherosclerosis should always be considered in the differential diagnosis of LVV. Other pathologies which may mimic LVV include, but not limited to, congenital causes of aortic coarctation and middle aortic syndrome, immunoglobulin G4-related disease, and hereditary disorders of connective tissue such as Marfan syndrome and Ehler–Danlos syndrome.

Keywords Vasculitis · Large vessel · Giant cell arteritis · Temporal arteritis · Takayasu · Diagnosis · Differential diagnosis · Criteria · Mimickers · Aortitis

Introduction

Vasculitides include a heterogeneous group of diseases, characterized mainly by inflammation of blood vessels which may result in damage and necrosis of the vessel wall. Patients with vasculitides typically present with constitutional symptoms such as fever, fatigue, weakness, and muscle and joint aches, accompanied by variable involvement of many organs and systems. The current trend is to classify vasculitides based upon the size of the involved vessels. However, the presence of anti-neutrophilic cytoplasmic antibodies (ANCA), being isolated in a single organ, or being associated with systemic disease, or having a probable etiology, are also considered within supportive parameters for classification [1].

Large vessels include aorta, its major branches, and extremity arteries and the analogous veins according to the 2012 International Chapel Hill Consensus Conference (CHCC) nomenclature of vasculitides. Large-vessel vasculitis (LVV) is defined as a vasculitis that affects large arteries more often than other vasculitides do [1]. In the present review, we concentrate on the diagnosis and differential

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diagnosis of large LVV, which include giant cell arteritis (GCA) and Takayasu arteritis (TAK).

Methods

We conducted a comprehensive review of the literature for English articles published between 1980 and 2018, using PubMed, Scopus, and Web of Science as databases. The keywords of "vasculitis", "large vessel", "giant cell arteritis", "temporal arteritis", and "Takayasu" "aortitis" were searched in combination with the keywords of "diagnosis", "differential diagnosis", "criteria", and "mimickers". We also manually searched the references of the selected articles for any relevant reference that we might have missed.

The general approach to the concepts of "diagnostic" and "classification" criteria

Currently, there are no universally accepted diagnostic criteria, neither for LVV nor other systemic vasculitides and other autoimmune diseases such as systemic lupus erythematosus (SLE). Diagnostic and classification criteria are not the same. Ideal diagnostic criteria must cover the different and heterogenic features of a disease, including those with unusual presentations. Naturally, such an approach will increase the sensitivity, at the expense of lowering specificity. However, classification criteria are standardized definitions aiming to cover familiar and homogenous clinical scenarios for a particular disease. Therefore, classification criteria do not aim to capture all patients, and may easily miss those patients with atypical features, resulting in lower sensitivity, but higher specificity. Therefore, classification criteria are not appropriate for the diagnosis of LVV in routine clinical care; instead, they serve to classify already diagnosed vasculitis patients and include them in clinical studies [2].

Ishikawa performed the first attempt to develop diagnostic criteria for TAK in 1988, and these criteria were later modified by Sharma et al. in 1995 (Table 1) [3–5]. The traditional classification criteria of the American College of Rheumatology (ACR) for TAK and GCA were defined in 1990 (Table 1) [6, 7]. Considerable progress occurred in diagnostic imaging modalities for both diseases, including magnetic resonance angiography (MRA), computerized tomography angiography (CTA), color Doppler ultrasonography (CDU), and positron emission tomography (PET) with 18F-fluorodeoxyglucose (18F-FDG-PET), since then. Arteriogram included in the 1990 ACR criteria for TAK is no longer the gold standard for the diagnosis of TAK [8]. Similarly, temporal artery biopsy (TAB) included in the 1990 ACR criteria for GCA is no longer the only tool for diagnosing GCA [8]. Thus, these criteria certainly need appropriate revisions: One current suggestion for revising the ACR criteria is to avoid TAB for the diagnosis of GCA [9]. There is also a growing tendency to use CDU for diagnosing GCA [8]. There is an ongoing multi-national observational study called as DCVAS (diagnostic and classification criteria for vasculitis study), designed to develop and validate diagnostic criteria for six different primary systemic vasculitides, including GCA and TAK [10].

The contribution of medical history, physical examination, and laboratory tests to the diagnosis of large-vessel vasculitides

Symptoms suggestive of TAK and GCA may change depending upon the time from the disease onset and the extent of organ/system involvement. Most of the initial symptoms are non-specific, mostly reflecting vague constitutional symptoms caused by systemic inflammation, which may increase as the disease progresses. Transmural, granulomatous, inflammatory process within the arterial wall leads to intimal hyperplasia and vessel occlusion, thereby causing ischemia and damage of the affected organs. Destruction of the large elastic arteries may lead to progressive dilatation, aneurysm formation, and potentially fatal vascular events such as dissection or rupture [11, 12].

Palpation of arterial pulses, blood pressure measurements of all extremities, and cardiac and neck auscultation for detecting bruits are crucial parts of the physical examination. There are no unique, highly sensitive or specific diagnostic tests. Elevated erythrocyte sedimentation rate (ESR), and serum C-reactive protein (CRP), leukocytosis with granulocytosis, thrombocytosis, and normochromic/normocytic anemia reflect the acute phase response [13, 14]. Leukopenia and thrombocytopenia are not expected in LVV except in case of immunosuppressive agent usage. As the systemic inflammation persists, hypoalbuminemia and polyclonal hyperglobulinemia may accompany [13]. However, the absence of an elevated acute phase response by itself is not sufficient to exclude the diagnosis of LVV [15]. Given that serum amyloid A (S-AA) protein increases rapidly even in the presence of minimal systemic inflammation, it may be useful to measure S-AA levels in the presence of normal ESR and CRP levels [16].

Given that vascular wall and systemic inflammation may be discordant especially in TAK, ESR and CRP levels may be normal despite ongoing disease activity [17]. Inflammatory molecules that are produced at sites of vascular inflammation may be expected to reflect the degree of current vascular wall inflammation better. Pentraxin-3 (PTX-3) is involved in the maintenance of vascular homeostasis, and measuring plasma levels of PTX-3 was suggested as a more reliable biomarker to reflect current vascular disease activity in TAK [18]. Although PTX-3 certainly had some

Table 1 Summary of the diagnostic and classification criteria for Takayasu arteritis (TAK) and giant cell arteritis (GCA)

	Ishikawa diagnostic criteria for TAK (1988) [4]	Sharma modification of Ishikawa's diagnostic criteria for TAK (1995) [5]	American College of Rheumatology Classification Criteria for TAK (1990) [6]	American College of Rheumatology Classification Criteria for GCA (1990) [7]
Number of criteria	An obligatory criterion, plus two major and nine minor criteria	Three major and ten minor criteria	Six criteria	Five criteria
Details of criteria	The obligatory criterion: occurrence of defined characteristic signs and symptoms[a] at the age ≤ 40 years, for at least 1 month Two major criteria: angiographic evidence of right and left mid subclavian artery lesions Nine minor criteria: included high ESR, unilateral or bilateral common carotid artery tenderness, hypertension, aortic regurgitation, and angiographic evidence of pulmonary artery, left mid common carotid, distal brachiocephalic trunk, descending thoracic and abdominal aorta lesions	Obligatory criterion (age ≤ 40 years) removed "Characteristic signs and symptoms of TAK" accepted as major criterion Other modifications Removal of age in the definition of hypertension Exclusion of the absence of aorto-iliac lesions Inclusion of coronary artery lesions in patients younger than 30 years in the absence of risk factors as a minor criterion	Age of onset before 40 years Extremity claudication Decreased brachial artery pulse Difference of more than 10 mmHg systolic pressure between two limbs Bruit over subclavian arteries or the aorta Angiographic evidence of narrowing or occlusion of the aorta, its primary branches, or large arteries in the proximal upper or lower extremities	Age at disease onset ≥ 50 years New headache Temporal artery abnormality: tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries ESR ≥ 50 mm/h by the Westergren method Abnormal artery biopsy: Arterial specimen showing vasculitis characterized by predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells
Special note	Absence of aorto-iliac involvement was a requisite	Acceptance of aorto-iliac involvement	–	–
Required criteria	Obligatory criterion plus two major criteria, OR A major criterion plus at least two minor criteria, OR At least four minor criteria	Two major criteria, OR A major criterion plus two minor criteria, OR Four minor criteria	At least three of the six criteria	At least three of the five criteria
Sensitivity	84%	92.5%	90.5%	93.5%
Specificity	100%	95.0%	97.8%	91.2%

ESR erythrocyte sedimentation rate

[a]Defined characteristic signs and symptoms of TAK: absence of pulses, differences in pulses in the arms, unobtainable blood pressure, significant blood pressure differences in the arms, easy limb fatigability or pain, unexplained fever or high ESR (≥ 20 mm/h; Westergren) or both, neck pain, transient amaurosis or blurred vision or syncope, dyspnea or palpitations or both, or hypertension or aortic regurgitation

advantages compared to CRP, and some studies reported promising results, unfortunately, PTX-3 could not solve the dilemma of effectively detecting ongoing vascular wall inflammation in TAK [19–21].

Role of imaging modalities for clinical diagnosis of large-vessel vasculitides

Imaging modalities are essential for the diagnosis of patients with LVV [8, 22–25]. Advantages and disadvantages of imaging methods including conventional angiography [23], MRA [26–30], CTA [26, 31, 32], CDU [25, 33–35], and 18F-FDG-PET-CT [36–40] were given in Table 2. In summary, the conventional angiography is no longer considered as the gold standard for the diagnosis of TAK. Currently, many physicians prefer to use MRA or CTA, or in selected cases 18F-FDG-PET-CT, for establishing the diagnosis of TAK.

Recently, PET/CT was reported to be valuable in the assessment of local inflammatory and vascular remodeling events independent from systemic inflammation in TAK. PET/CT was also reported to detect even the lesions where the arterial wall was less than 4 mm [40]. The authors suggested that PET/CT might serve as a complementary modality in addition to the traditional inflammatory markers for the assessment of current disease activity. Interestingly, there are also attempts for combining functional PET data with morphological data derived from magnetic resonance imaging (MRI). Hybrid PET/MRI offers lower radiation exposure compared to PET/CT, which may be an advantage for the imaging of LVV [40, 41].

Table 2 Comparison of the imaging methods for the diagnosis of large-vessel vasculitides

	Advantages	Disadvantages
Conventional angiography [22–25]	Assessing the extent and localization of vascular involvement Detecting stenosis, occlusions, and aneurysms in large and medium-sized arteries	Visualizing only the lumen of the vessel, without giving any information about the vessel wall Missing minor, non-occlusive vascular lesions Lack of adequate resolution for small vessels Being an invasive method Causing radiation and contrast media exposure
Computerized tomography angiography (CTA) [22–26, 31, 32]	Evaluation of aorta and its primary branches Excellent anatomical characterization of structural changes Differentiating vascular and perivascular structures Detecting calcifications Shorter scanning time	Unable to visualize relatively small vessels Less resolution than CDU Causing radiation and contrast media exposure
Magnetic resonance angiography (MRA) [22–24, 26–30]	Evaluation of aorta and its primary branches Excellent anatomical characterization of structural changes Detecting vessel wall thickening, edema and contrast enhancement No risk of radiation exposure	Overestimation of vascular occlusions Inability to visualize small branch vessels and vascular calcifications More expensive Longer scanning time Less resolution than CDU
Color Doppler ultrasonography (CDU) [25, 33–35]	Evaluation of temporal, carotid, axillary, and femoral arteries Visualizing luminal changes, stenosis and aneurysms Detecting the characteristic, homogeneously thickened vessel wall, mural inflammation and edema Providing better resolution than MRA and CTA No risk of radiation exposure Cheaper	Diagnostic accuracy varies depending on the skill and experience of the operator May miss cases with early disease having patchy mural inflammation without transmural disease and associated tissue edema Fails to depict the thoracic aorta unless performed as a transesophageal examination
Positron emission tomography with 18F-fluorodeoxyglucose (18F-FDG-PET) [36–40]	Combining the functional information from PET and anatomical information from CT Most sensitive imaging method for early vessel inflammation Detecting early vascular inflammation and its location in the aorta and its branches	Vascular uptake of 18F-FDG not specific for vasculitis Discrimination between atherosclerotic and vasculitic lesions may be challenging No information for vessel wall structure and luminal flow High radiation exposure

Role of tissue biopsy for diagnosis of large-vessel vasculitides

Obtaining tissue biopsy and showing histopathological features of vessel wall inflammation are critical to confirm the diagnosis of LVV. However, in clinical practice, histopathological diagnosis using temporal artery biopsy (TAB) may only be possible in GCA, which is discussed in the following paragraphs.

Diagnosis of Takayasu arteritis

Definition of TAK

TAK is a frequently granulomatous vasculitis, predominantly affecting the aorta and its major branches. Age of onset is usually less than 50 years [1].

Clinical features of TAK

TAK presents with different symptoms and clinical findings, depending upon the duration and phase of the disease. The first phase is characterized by non-specific constitutional inflammatory symptoms, including fever of unknown origin. The second phase is characterized by vascular inflammation. Involvement of carotid arteries may cause carotidynia and neck pain. Similarly, mural inflammation in thoracic aorta may cause dorsal pain. In the late phase of the disease, severe narrowing or occlusions may occur mainly in the proximal parts of the arterial branches originating from arcus aorta. Decreased or absent upper extremity pulses, with or without discrepant measurements of arterial blood pressure between upper extremities, arterial bruits, and intermittent extremity claudication are among typical features of late-stage TAK. Severe hypertension may also occur in TAK; it may be caused by atypical coarctation of the aorta, loss of vascular compliance, aortic valve regurgitation due to aortitis, or renal artery stenosis [12, 42–44].

The occurrence of new and severe ischemic lesions is most common in TAK, although ischemic vascular manifestations including transient ischemic attack and stroke may occur in almost all types of vasculitides [45].

Diagnostic and classification criteria for TAK

Ishikawa diagnostic criteria for TAK were published in 1988 [4]. Ishikawa defined "characteristic signs and symptoms for TAK", and the occurrence of those signs and symptoms starting before 40 years of age for at least 1 month was accepted as an obligatory criterion. ACR classification criteria for TAK were published 2 years later than Ishikawa criteria [6]. The main criticism for Ishikawa 1988 and ACR 1990 criteria for TAK was the limitation of the age to less than 40 years, which led to modification of Ishikawa's criteria by Sharma et al. in 1996 [5]. They removed the obligatory criterion (age $\leq$ 40 years) and accepted the "characteristic signs and symptoms of TAK" as a major criterion. Ishikawa diagnostic criteria, Sharma modification of these criteria, and ACR 1990 classification criteria for TAK are summarized in Table 1.

ACR 1990 criteria generally cover the cases with TAK in the late stages, where it is not difficult to make the diagnosis. ACR 1990 criteria were also criticized for the selection of the control group used, which included patients mainly with small-vessel vasculitis, rather than those with atherosclerotic or congenital aortic diseases [3].

TAK may also be seen in childhood, and recently, classification criteria for childhood TAK were proposed by the European League Against Rheumatism (EULAR), the Pediatric Rheumatology European Society (PRES), and by the Pediatric Rheumatology International Trials Organization (PRINTO) for patients younger than 18 years. These criteria also aimed to include these patients in epidemiologic studies and clinical trials (Table 3) [46].

Angiographic classifications and heterogeneity of TAK

Initial clinical findings of TAK may be different and heterogeneous based upon the location and extent of vessel involvement. For this reason, there have been attempts to classify patients with TAK according to involved vessels, based upon angiographic findings. Both Sheikhzadeh et al. [47] and Nasu [48] reported angiographic classifications for TAK, in 1982. However, Numano's angiographic classification for TAK is currently the most widely used classification [49]. Summary and comparison of these three angiographic classifications are given in Table 4.

Trying to make diagnosis of TAK earlier

The early diagnosis of TAK is the real goal, which is difficult to establish. For the early diagnosis, the clinician should consider the possibility of this disease in suspected cases. There are red flags which may notify the clinician to investigate TAK in a young patient with persistent systemic inflammation (Table 5) [50].

Heterogeneity of TAK should also be kept in mind for the early diagnosis. Genetic and ethnic factors may affect the primary location of vessel involvement. For example, vascular lesions tend to occur primarily in the ascending aorta, aortic arch, or its branches, and extend into the abdominal aorta in Japanese patients with TAK. On the other hand, abdominal aorta including renal arteries and thoracic aorta are primarily involved in Indian patients [49]. Type V

Table 3 EULAR/PRINTO/PRES criteria for childhood Takayasu arteritis [46]

Criterion	Definition
Angiographic abnormality (mandatory criterion)	Angiography (conventional, computed tomography, or magnetic resonance imaging) of the aorta or its main branches and pulmonary arteries showing aneurysm/dilatation, narrowing, occlusion or thickened arterial wall not due to fibromuscular dysplasia, or similar causes; changes usually focal or segmental
Pulse deficit or claudication	Lost/decreased/unequal peripheral artery pulse(s) Claudication: focal muscle pain induced by physical activity
Blood pressure (BP) discrepancy	Discrepancy of four limb systolic BP > 10 mmHg difference in any limb
Bruits	Audible murmurs or palpable thrills over large arteries
Hypertension	Systolic/diastolic BP greater than 95th percentile for height
Acute phase reactants	Erythrocyte sedimentation rate > 20 mm per first hour or C-reactive protein any value above normal (according to local laboratory)

Takayasu arteritis is classified when the mandatory criterion is present plus any other criteria

involvement of Numano's angiographic classification is most common in Indian patients with TAK (Table 4) [49]. For this reason, renovascular hypertension together with persistent systemic inflammation may lead the clinician to the possible early diagnosis of TAK in Indian patients [51].

In summary, the first step for the early diagnosis is a high index of suspicion in selected cases, and the second step is to confirm the diagnosis of TAK by appropriate imaging methods, as summarized in Table 2. Our approach is to use MRA for confirming the diagnosis of TAK. We tend to use CTA when MRA is not available for technical reasons. Although FDG-PET-CT scan has gained considerable acceptability, we use this imaging method for the diagnosis of TAK in selected difficult cases. Given that MRA has no radioactivity and is very useful both to assess wall thickening/edema and to detect the presence of aneurysms/thrombus, we and many authors recommend MRA in patients with TAK for close follow-up.

Diagnosis of giant cell arteritis (GCA)

Definition of GCA

GCA, also known as temporal arteritis, is an LVV, often granulomatous, and usually affecting the aorta and its major branches. GCA has a predilection for the branches of the carotid and vertebral arteries, especially the temporal artery [1].

Clinical features of GCA

It is usually seen in patients older than 50 years and often associated with polymyalgia rheumatica (PMR). Extensive systemic inflammation, accompanying vascular inflammation is present in the majority of the cases [1].

It is not difficult to diagnose GCA in the presence of a new-onset headache and temporal artery abnormalities, associated with a systemic inflammatory syndrome and proximal muscle pain in an elderly patient. Likewise, acute ocular symptoms such as impaired vision, diplopia, and amaurosis fugax should always remind the clinician the possibility of GCA. Such ophthalmological emergencies generally result from anterior ischemic optic neuropathy due to occlusion of the posterior ciliary or ophthalmic arteries. Similarly, claudication of the jaw or tongue or both, and upper respiratory symptoms such as a non-productive cough, sore throat, or hoarseness may direct the clinician to consider the possibility of GCA [52].

PMR is characterized by inflammatory pain and stiffness affecting the neck, shoulders, hips, and proximal extremities, with frequent occurrence of subdeltoid bursitis, biceps tenosynovitis, glenohumeral synovitis, or trochanteric bursitis. Symptoms are usually bilateral and more pronounced in the morning or after periods of inactivity, and there is severe systemic inflammation. PMR shares many epidemiologic and pathogenic features with GCA and may accompany GCA with a reported wide range of frequency from 17 to 66% [52].

Different subgroups of GCA

- Two different subgroups of GCA were reported concerning the severity of systemic inflammation. The first subgroup is mainly characterized by severe systemic inflammation, while, in the second subgroup, there is less inflammation, but a prominent vaso-occlusive process. Acute vision loss is generally seen in the second subgroup [17, 53].
- There is also a subgroup of GCA patients presenting with large-vessel involvement (LVI) without cranial arteritis [54]. The clinical presentation may be highly variable in this subgroup. Thoracic aorta and extremity arter-

Table 4 Proposed angiographic classifications for Takayasu arteritis

Sheikhzadeh 1982 [47]	Nasu 1982 [48]	Numano 1996 [49]
Type I: aortic arch type (cervicobrachial type)	Type I: primarily branches of aortic arch	Type I: branches of aortic arch
Type II: thoracoabdominal type	Type II: aortic arch and its branches	Type IIa: ascending aorta, aortic arch and its branches
Type III: peripheral type; iliac arteries	Type III: abdominal aorta and particularly renal arteries	Type IIb: thoracic descending aorta plus Type IIa
Type IV: combination of two or more of the previous types with or without pulmonary or coronary artery involvement	Type IV: whole aorta and its branches	Type III: thoracic descending aorta, abdominal aorta and/or renal arteries
		Type IV: abdominal aorta and/or renal arteries
		Type V: the combination of Type IIb and Type IV
		Note: involvement of coronary or pulmonary arteries should be indicated as C (+) or P (+), respectively

Table 5 Red flags to investigate Takayasu arteritis in a young patient with persistent systemic inflammation [50]

Carotidynia
Hypertension
Angina pectoris
Vertigo and syncope
Extremity claudication
Absent/weak peripheral pulses
Discrepant blood pressure in the upper limbs (> 10 mmHg)
Arterial bruits
Aortic regurgitation

ies may be involved in those patients. Although lower extremities may also be affected, upper extremities are affected more often, which may cause symptoms of vascular insufficiency such as claudication. This subgroup of GCA patients with LVI was reported to be younger, more likely to be women and to have less cranial symptoms and PMR. More importantly, GCA patients with LVI had an increased risk of aortic dilation [55]. Therefore, the presence of aortic insufficiency murmur in cardiac auscultation may implicate the possibility of this subgroup of GCA in a suspected patient.

- Recently, it has been suggested that there are two more different subgroups of GCA with regard to type of subclavian arterial involvement: The first subgroup, being more common, is characterized by wall thickening, stenosis, and occlusion of subclavian arteries, while dilation of subclavian arteries characterizes the second less common subgroup. Interestingly, aortic dilation has been reported to be more common and aortic wall thickening less common in the second subgroup. The authors recommend that patients with GCA having subclavian artery dilation should be evaluated and monitored carefully for possible aortic dilation and aneurysm [56].

Classification criteria for GCA

ACR 1990 criteria for the classification of GCA (Table 1) [7] concentrate on occurrence in older patients, new-onset typical headache, prominent constitutional symptoms, abnormalities of temporal artery in physical examination, and typical histological findings obtained from TAB. Ocular symptoms and claudication of the jaw and tongue are not included within these criteria. More importantly, the subgroup of GCA with LVI is not covered in these criteria.

Histologic diagnosis of GCA

TAB is still vital for diagnosing GCA, especially in patients with symptoms of cranial arteritis. The artery specimen should be at least 1 cm in length to avoid missing inflammatory segments of the vessel. Supporting this view, the rate

of positive findings was only 25.7% in specimens less than 1 cm in length, while 74.3% in specimens ≥ 1 cm in length [57]. Whether TAB should be made unilateral or bilateral is controversial. Some authors recommend routine bilateral TAB for all patients, while others perform unilateral biopsy initially, and make a contralateral biopsy only if the initial biopsy is negative. We also prefer to perform unilateral TAB initially. Breuer et al. reported that performing bilateral TAB increased the diagnostic sensitivity of the procedure by up to 12.7%, compared to unilateral TAB [58]. However, the rate of discordant results in bilateral TAB was calculated as 5.9% in another study [59]. If the patient needs urgent corticosteroid (CS) treatment such as in the presence of an ophthalmic emergency, the treatment should not be delayed. Fortunately, TAB remains a valuable diagnostic procedure even after several weeks of CS treatment. Nevertheless, approximately 10% of patients with the clinical diagnosis of GCA will have negative TAB [52].

Diagnosis of GCA with cranial symptoms by imaging methods

Imaging of the superficial temporal arteries with CDU or MRA is an alternative method for the diagnosis of GCA with cranial symptoms, and may be tried in selected cases [8]. CDU may evaluate temporal, carotid, axillary, and femoral arteries, by visualizing luminal changes, stenosis, and aneurysms. CDU may detect the characteristic, homogeneously thickened vessel wall and mural inflammation in the presence of vasculitis. The hypoechoic area surrounding the lumen of the artery is thought to represent edema and vascular inflammation of the arterial wall and called as 'halo sign' which may contribute to the diagnosis of GCA [25] (Fig. 1).

Overall sensitivity and specificity of CDU for detecting abnormalities such as halo sign, stenosis, or occlusion in GCA were 88% and 78%, respectively, as compared to TAB [8, 25]. A recent study aimed to evaluate the intima-media thickness of temporal, facial, and axillary arteries involved in GCA for determining cut-off values which may be helpful for diagnosis and follow-up of GCA. The authors reported that CDU could correctly distinguish vasculitic from normal arteries in patients with suspected GCA [34].

As discussed above, discontinuous and skip inflammatory changes in the temporal artery may cause false-negative biopsy results in nearly 13–44% of patients, mainly if the length of the biopsy specimen is too short. Therefore CDU-guided TAB might be expected to improve the sensitivity for GCA diagnosis. However, in a recent study, CDU-guided TAB did not result in improved sensitivity [35]. The authors reported that the probability of a positive TAB is high in the presence of the halo sign on CDU, regardless of whether the TAB is guided by CDU or not [35].

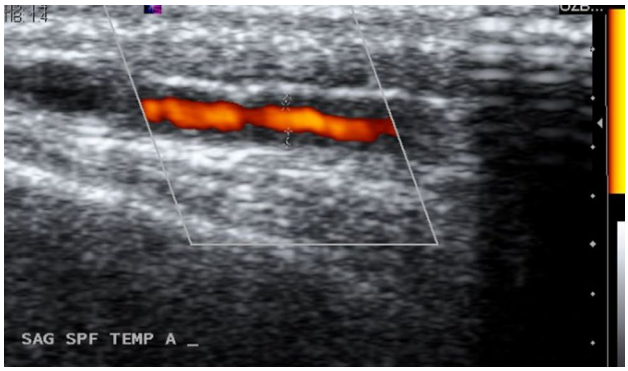

Fig. 1 Power Doppler ultrasound image of the right superficial temporal artery of a 73-year-old woman delineates blood flow and concentric thickening of the vessel wall ("halo sign"). The thickness of the hypoechoic wall was measured as 0.5 mm all along its preauricular segment

TAB versus CDU of superficial temporal arteries for the diagnosis of GCA with cranial symptoms

TABUL study is an excellent multicenter prospective study which compared CDU and TAB in the new cases of suspected GCA. A training program was given to sonographers to standardize the CDU assessment of temporal and axillary arteries. Analysis of 381 patients who underwent both USG and TAB within 10 days of starting treatment for suspected GCA showed that sensitivities of TAB and USG were 39% and 54%, respectively; while the specificities were 100% and 81%, respectively [33].

Diagnosis of GCA with large-vessel involvement by imaging methods

In such cases presenting with LVI, without cranial arteritis, appropriate imaging methods including MRA or CTA may confirm the diagnosis of GCA. However, 18F-FDG-PET/CT scanning is generally the first option in selected cases, and may also be useful to exclude malignancy in the differential diagnosis of GCA.

Similarities and differences between Takayasu arteritis and giant cell arteritis

Whether TAK and GCA represent a spectrum of the same disease is an ongoing debate [60–63]. Despite similarities [64–66], there are also striking differences between GCA and TAK including major histocompatibility complex associations [67], localization of prominent vessel wall inflammation [68], type of aortic involvement [69], and distribution of the involved arteries [70]. Cytokine response patterns to CS treatment [61, 71–73], and clinical responses to anti-TNF treatment [74–76] and abatacept treatment [77, 78] are

also different. Similarities and differences between GCA and TAK are summarized in Table 6. Although the debate goes on, we believe that GCA and TAK are different diseases.

Differential diagnosis between TAK and the subgroup of GCA with LVI

Since typical cranial symptoms are rare in the subgroup of GCA with LVI, differential diagnosis with older TAK patients may occasionally be difficult. Peripheral vascular abnormalities on examination, lower extremity claudication, fever, arthralgia/myalgia, and stroke/transient ischemic attacks are more common in TAK, while upper extremity claudication occurs more often in GCA with LVI. The presence of aortic aneurysms is more likely in GCA with LVI, while aortic stenotic lesions occur more commonly in TAK. Aortic aneurysms occur mostly in the ascending thoracic aorta in GCA [62, 63, 69].

Differential diagnosis between large-vessel vasculitides and mimickers

Various pathologies may mimic LVV either by causing systemic inflammation and constitutional symptoms or by causing lumen narrowing or aneurysm formation in the aorta and its branches. As a general rule, infectious and malignant diseases should be considered in the differential diagnosis of elevated acute phase response, before making the diagnosis of LVV, especially of PMR. These pathologies may not only mimic LVV, but may also cause secondary vasculitis with a wide range of vasculitic symptoms [79, 80]. In selected cases, 18F-FDG-PET/CT may be useful to exclude malignancy in the differential diagnosis of GCA, as well as to confirm the diagnosis of GCA with LVI.

Since aortitis is a critical component seen both in TAK and in the subgroup of GCA with LVI, any pathology causing aortitis may confuse differential diagnosis. The early diagnosis of aortitis is of primary importance to prevent serious complications, such as aneurysmal rupture or aortic dissection. The term "aortitis" is, indeed, a histopathological term implicating inflammation of media and adventitia, accompanied with the classic appearance of "tree-barking" of the aortic intima [81]. However, in clinical practice, the diagnosis of aortitis is generally made based on imaging techniques such as FDG-PET/CT, MRA, or CTA [81–83].

Aortitis may be infectious or non-infectious. Among infectious agents, the most common pathogenic species involved are Salmonella, *Staphylococcus aureus, Streptococcus pneumonia, Mycobacterium tuberculosis*, human immunodeficiency virus, and, even if now rare, *Treponema pallidum* [83]. In most cases of infectious aortitis, the responsible microorganism may colonize the aorta by entering via the vasa vasorum. The presence of atherosclerotic plaque, aneurysm sac, or any other injury in any segment of the aortic wall may predispose to the colonization of microorganisms [84–89]. Going into details of infectious aortitis is beyond the scope of this review. However, the message to the clinician is straightforward: in the presence of aortitis, infectious causes should always be excluded using the standard microbiological diagnostic tests.

Non-infectious causes of aortitis should be considered after the exclusion of infectious aortitis. Although GCA and TAK are the most frequent causes of autoimmune aortitis, other systemic autoimmune diseases, such as SLE [90], Sjögren's syndrome [91], rheumatoid arthritis [92], ANCA-associated vasculitides [93, 94], HLA-B27 associated spondyloarthropathies [95], psoriatic arthritis [96], sarcoidosis [97], Cogan's syndrome [98], relapsing polychondritis [99], inflammatory bowel diseases [91, 100, 101], Behçet's disease [102], and IgG4-related disease [103], and possibly some other diseases may also cause occurrence of aortitis. Aortitis may also occur in PMR patients without GCA. In the presence of atypical symptoms such as unexplained thigh pain, low back pain or lower limb pain, and an inadequate response to low-to-medium doses of CS, associated aortitis should be suspected in PMR [91].

It should be noted that aortitis may occasionally be idiopathic and may occur in isolation, with a prevalence ranging between 4.3 and 8.4% [104, 105]. Histologically isolated aortitis is virtually indistinguishable from GCA and TAK [106]. The risk factors for the development of isolated aortitis were reported as advanced age, history of connective tissue disease, diabetes mellitus, and heart valve pathology [105].

Differences between isolated aortitis and GCA-associated aortitis have recently been described by Talarico et al. [107]. Isolated aortitis is seen predominantly in male and younger patients when compared to GCA. Aortic arch, and thoracic and abdominal aortas are involved, while aortic branches seem to be spared. Unfortunately, whether patients with isolated aortitis represent variants of GCA or TAK, or how they will evolve in the future is indefinite [107].

After a differential diagnosis of aortitis is over, other pathologies which may mimic LVV should also be considered. Among pathologies affecting aorta and its branches, atherosclerosis is probably the most common mimicker, especially in older patients. Association between inflammation and atherosclerosis is well known, and vasculitic/atherosclerotic lesions may be present together even in young patients [108, 109]. Given that atherosclerosis is also an inflammatory process, atherosclerotic plaques may show increased uptake of gadolinium contrast in MRA and increased FDG uptake in PET-CT imaging, resulting in further confusion in differential diagnosis [110]. However, there are some clues which may help to differentiate

Table 6 Similarities and differences between Takayasu arteritis (TAK) and giant cell arteritis (GCA)

Similarities	Differences		
	Parameter	TAK	GCA
Female dominance [60–63] Common role of cell mediated immunity in pathogenesis [60–63] Granulomatous inflammation in vessel wall [60–63] IL-12B as the most prominent genetic factor for both diseases [66] High serum levels and vascular expressions of interleukin (IL)-6 and IL-17 [61, 63] Favorable response to corticosteroid and anti-IL-6 treatments [63–65]	Demographic data [60–63]	Young females from Far-Eastern and Asian countries	Older patients, especially Caucasian origin
	Association with major histocompatibility complex (MHC) [67]	MHC class I alleles	MHC class II alleles
	Histopathology [68]	Predominant adventitial scarring	Vessel wall inflammation prominent in the inner layer
	Symptoms and physical findings [60–63]	Peripheral vascular abnormalities including bruits, absent pulses and blood pressure discrepancies	Cranial symptoms including headache, jaw or tongue claudication, scalp tenderness, and vision loss
	Aortic involvement [69]	Stenotic lesions	Thoracic aneurysmal dilatation
	Carotide involvement [70]	Branches of the internal carotid artery more common	Branches of the external carotid artery more common
	Cluster analysis of involved arteries [70]	Left subclavian artery together with bilateral carotids, and also involvement of mesenteric artery	Symmetric subclavian with concomitant axillary arteries
	Cytokine response patterns to treatment [61, 71–73]	Unlike Th17 cytokines, serum levels of Th1 cytokines are easily suppressed in TAK	Unlike Th1 cytokines, serum levels of Th17 cytokines are easily suppressed in GCA
	Response to biologic treatments [61, 74–78]	While anti-TNF treatment is generally effective, abatacept is not	While abatacept is effective, anti-TNF treatment is not

Table 7 Pathologies which should be considered in the differential diagnosis of giant cell arteritis and/or Takayasu arteritis

Mimicker	Definition	Comments for differential diagnosis
Atherosclerosis [108–116]	Inflammatory vascular disease Traditional risk factors plus inflammation contribute May cause aortic aneurysms and lumen narrowing in aortic branches and medium-sized arteries	Localized in bifurcation sites and in ostiums, rather than proximal parts of the arteries Location of aortic aneurysms: abdominal (mostly intrarenal), rather than thoracic Upper limb arteries rarely involved Punctate, linear calcifications and discrete plaque lesions Localized hot spots, rather than linear smooth signals in PET-CT Localized thickening of intima-media with ultrasonography, rather than concentric, smooth thickening, and long-segment stenosis
Mycotic aneurysm [84, 85, 87, 89]	May occur as a complication of septicemia, usually in the context of a bacterial infection, classically spreading from the heart, as in infective endocarditis Thoracic and abdominal aorta, peripheral, intracranial, and abdominal arteries involved	Presence of fever, abdominal pain, pulsatile abdominal mass and cardiac murmurs, plus positive blood cultures Transesophageal echocardiography may confirm the diagnosis of infective endocarditis by showing vegetations affecting the cardiac structures, aorta root, and ascending aorta
Syphilis (Sy) [84–86]	Cardiovascular Sy may cause inflammation in the ascending aorta, resulting in aortic wall thickening, aneurysm formation, aortic valvular incompetence and coronary artery disease	Positive serology for Sy
Tuberculosis (TB) [84–86]	*Mycobacterium tuberculosis* may cause granulomatous arteritis leading to vessel wall thickening, stenosis, and aneurysm in the aorta and its branches, thereby mimicking LVV	Unlike typical arterial stenosis of TAK, TB aortitis causes vessel wall erosion of thoracic and abdominal aorta resulting with formation of true or false aneurysms
HIV (human immunodeficiency virus) infection [88]	Wide range of vasculitides may be seen Multiple aneurysm formation or occlusive disease may occur	Immunocompromised host Positive serology for HIV
Immunoglobulin G4-related disease (IgG4-RD) [103]	A fibroinflammatory condition involving many organs including pancreas, major salivary glands, orbital tissue, lungs, kidneys, biliary tree, lymph nodes, and retroperitoneum May cause true aortitis, creating confusion with LVV	Increased serum IgG4 Histology shows lymphoplasmacytic infiltrate with increased IgG4 plasma cells, storiform fibrosis, obliterative phlebitis and mild-to-moderate tissue eosinophilia
Congenital aortic coarctation [117, 118]	Commonly located in the junction of distal aortic arch and descending aorta, immediately beyond the origin of the left subclavian artery	Unlike TAK, more common in males Location of coarctation and absence of systemic inflammation may be helpful for differential diagnosis
Middle aortic syndrome [119, 120]	Segmental narrowing of the abdominal or distal descending thoracic aorta	May be caused by various pathologies Main differential diagnosis incudes TAK, MFS, EDS, LDS, and NF1
Marfan syndrome (MFS) [86, 121]	Autosomal dominant disorder of the connective tissue matrix Results from mutations in the fibrillin-1 gene May affect the wall of the thoracic aorta leading to aneurysm formation, dissection, and, sometimes, aortic regurgitation, thereby mimicking LVV	Typical Marfanoid body status and clinical features including lens dislocation No systemic inflammation No arterial wall thickening or stenosis with imaging Histopathology showing cystic medial necrosis without inflammation
Ehlers–Danlos Syndrome Type IV (EDS) [86, 122]	Autosomal dominant disorder of the connective tissue matrix Results from mutations in the type III procollagen gene Descending and abdominal aorta may be affected, resulting in dissection, rupture, or aneurysm	No systemic inflammation No arterial wall thickening or stenosis with imaging Similar histopathology with MFS, i.e., cystic medial necrosis without inflammation

Table 7 (continued)

Mimicker	Definition	Comments for differential diagnosis
Loeys–Dietz syndrome (LDS) [86, 123]	Another genetic disorder of the connective tissue matrix Results from mutations in the TGF-β receptor gene May cause tortuosity, aneurysms, and dissections in thoracic and abdominal aortaç	No systemic inflammation Genetic testing needed Additional presence of hypertelorism, bifid uvula, cleft palate, and bicuspid aortic valve helpful in diagnosis
Neurofibromatosis type 1 (NF1) (von Recklinghausen disease) [124, 125]	May cause vascular aneurysms/arteriovenous malformations, renal artery stenosis, coarctation of aorta, or segmental narrowing of abdominal or distal descending thoracic aorta	Absence of systemic inflammation Neurocutaneous tumors, plexiform tumors, optic gliomas, hamartomatous Lisch nodules in the iris, café au lait macules and learning disabilities may be helpful in differential diagnosis
Fibromuscular dysplasia [86, 126]	Non-inflammatory vasculopathy of small and medium-sized arteries including renal, internal carotid, and vertebral arteries May lead to aneurysm, stenosis, occlusion, and dissection Renovascular hypertension is frequent Headache and limb claudication may mimic GCA or TAK	No systemic inflammation Middle and distal portions of renal, internal carotid, and vertebral arteries most commonly affected; aortic involvement rare Classic "string-of-beads" appearance, or focal concentric narrowing and diffuse tubular stenosis with angiography No arterial wall thickening, edema, or contrast uptake on magnetic resonance angiography
Primary amyloidosis [127, 128]	Amyloid deposition in temporal artery may mimic GCA	Presence of organomegaly and isolated proteinuria not expected in GCA Congo red staining with biopsy confirms amyloid deposition
Erdheim–Chester disease (ECD) [129]	Non-Langerhans histiocytosis affecting multiple organs, with heterogeneous presentation Vascular involvement may mimic LVV	Histological features include xanthogranulomatous infiltration of foamy histiocytes surrounded by fibrosis Cortical osteosclerosis and associated pain of long bones typical
Segmental arterial mediolysis (SAM) [130]	Frequently involves abdominal visceral arteries Causes vacuolar degeneration of smooth muscle cells in the arterial media, predisposing to dissecting aneurysms, stenosis and occlusions in medium-sized and large vessels	Presence of postprandial abdominal pain caused by intestinal ischemia with or without intraabdominal bleeding Vascular lesions generally limited to a single anatomic location Absence of other inflammatory, infectious, or heritable diseases
Post-radiation lesions [131]	Injury to vascular endothelial cells causes intimal thickening and irregularity, plus focal fibrosis and necrosis	History of previous malignancy and radiotherapy

atherosclerosis from vasculitis, as summarized in Table 7 [111–116].

Other pathologies which may mimic LVV include congenital causes of aortic coarctation and middle aortic syndrome [117–120], hereditary disorders (Marfan syndrome [121], Ehler–Danlos syndrome [122], Loeys–Dietz syndrome [123], neurofibromatosis [124, 125], fibromuscular dysplasia [126]), primary amyloidosis [127, 128], Erdheim–Chester disease [129], segmental arterial mediolysis (SAM) [130], and post-radiation vascular lesions [131]. Practical points for differentiating those mimickers from LVV were also given in Table 7.

Conclusion

The diagnosis of TAK and GCA may be accomplished by combining the patient symptoms, physical examination findings, blood test results, imaging findings, and biopsy results if available. Disease heterogeneity determined by the type and extent of vessel involvement should be considered for the early diagnosis of TAK. Persistent systemic inflammation plus presence any of the red flags should warn the clinician to investigate TAK. In a young patient with hypertension, the clinician should check all the peripheral pulses and measure the blood pressure in all limbs. A possible diagnosis of TAK should be confirmed using appropriate imaging methods in suspected cases. The conventional angiography is no longer the gold standard imaging technique for diagnosing TAK.

Similarly, TAB is no longer the only tool for diagnosing GCA presenting with cranial symptoms, and there is a growing tendency to use CDU for the evaluation of superficial temporal arteries. For the early diagnosis of GCA, ESR and CRP should be performed as initial screening tests in any elderly patient presenting with a unilateral headache. In case of high acute phase response, the next step should be scanning of the superficial temporal artery with CDU.

It should be noted that clinical presentation may be highly variable in a subgroup of GCA patients presenting with LVI, without prominent cranial symptoms. PET-CT, MRA, or CTA may be helpful for the detection of aortitis in this subgroup. There are also other different subgroups of GCA, which the clinician should be aware of, to reach the ultimate diagnosis.

In the presence of aortitis, differential diagnosis should be performed before reaching the diagnosis of LVV. The initial step should be to exclude infectious causes of aortitis. Even if aortitis is of non-infectious type, this finding is not unique for LVV. It should be noted that aortitis or other large-vessel involvement may occasionally be seen in various other autoimmune pathologies, as well. Moreover, aortitis may also be idiopathic and isolated. After a differential diagnosis of aortitis is completed, other pathologies which may mimic LVV, including atherosclerosis, congenital aortic problems, and hereditary collagen tissue disorders, should be excluded before diagnosing LVV.

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Compliance with ethical standards

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