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INVITED REVIEW

Diagnosis of Giant Cell Arteritis and Classification Criteria

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Giant cell arteritis (GCA) is a systemic granulomatous vasculitis of the aorta and its large branches. It frequently affects patients older than 50, and a female predominance has been observed in [1]. Although increased mortality has not been reported in population-based cohort studies on GCA, the development of permanent organ damage is a major concern. Vision loss due to ischemic optic neuropathy is one of the main findings in GCA and can be prevented with early diagnosis and treatment [2].

The 1990 American College of Rheumatology (ACR) Classification Criteria for GCA are widely used for classification and consist of 5 items. Patients meeting 3 out of 5 five items are classified as GCA (Table 1) [3]. The presence of fragmentation of the internal elastic lamina, in addition to arterial wall inflammation and mononuclear cell infiltration in temporal artery biopsy, is considered the gold standard for diagnosing GCA [4]. These criteria have significant drawbacks, including their limitation to cranial findings and development before the modern imaging methods.

A decrease in the sensitivity of the 1990 criteria is observed in recent cohort studies. This finding could be explained by the enrolment of patients with different clinical features in the cohorts depending on the improvement in the clinician's perspective over the years [5]. Implementing cross-sectional imaging methods, including Positron Emission Tomography – Computed Tomography (PET-CT), improved the diagnosis of GCA and facilitated the recognition of disease patterns without cranial manifestations.

Patients with isolated extra-cranial involvement reported a diagnostic delay of 2 to 5 months compared to classical DHA patients [6]. Late recognition of large-vessel involvement could cause permanent organ damage. Thoracic aorta dilatation/aneurysm has been reported in 15% and large artery stenosis in 30% of patients with proven aortic inflammation [7].

New classification criteria for giant cell arteritis have been planned based on the scope of DCVAS (The Diagnostic and Classification Criteria for Vasculitis) study data. They are expected to be published in 2022. Sudden vision loss, tongue or jaw claudication, C-reactive protein level, temporal artery ultrasound (US), PET-CT, and angiographic findings were the new items set by [8].

In the last decade, there have been significant advances regarding the role of imaging in diagnosing GCA. "EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice," published in 2018, emphasized the importance of early imaging but stated that imaging should not delay the initiation of treatment. The US of temporal and axillary arteries was recommended as the initial imaging modality of suspected cranial GCA [9]. In patients with a high probability of GCA (jaw claudication and increased acute phase reactants), positive findings in the temporal artery in the US were sufficient for diagnosis without an additional test (biopsy or further imaging) [10].

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Magnetic resonance imaging (MRI) may be used as an alternative to the US, but the main limitations of using MRI are high costs and restricted availability. The negative predictive value of MRI in combination with normal findings in temporal artery biopsy was found to be 98% [11].

PET-CT is a technique that can show inflammation in large vessels that we cannot investigate, and its sensitivity is higher than in MRI and CT field [12]. PET-CT may also be useful in the diagnosis of polymyalgia rheumatica and in distinguishing GCA from systemic diseases such as malignancies and chronic infections.

In conclusion, GCA should be at the top differential diagnosis list in patients with certain findings and a high probability of diagnosis (sudden vision loss, typical headache, jaw claudication, and increased acute phase reactants) due to the risk of permanent organ damage and treatment should be initiated as soon as possible. Even if treatment is started, early imaging methods and biopsy can be applied to confirm the diagnosis. The ACR 1990 Classification Criteria are insufficient to identify patients who are resistant to first-line agents and would participate in clinical trials evaluating the efficacy of further treatment options such as biologics. The new criteria to be published this year are expected to fill this gap. It should be kept in mind that these criteria were created for classification purposes and treatment should be decided individually based on clinical features.

Table 1. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis

1- Patient age > 50 years
2- New onset of localized headache
3- Abnormality of temporal artery (temporal artery tenderness, reduced pulsation)
4- Elevated erythrocyte sedimentation rate > 50 mm/1st hour
5- Abnormal temporal artery biopsy

REFERENCES

[1] Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, et al. 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum. 2013;65,1:1-11.

[2] Diamantopoulos AP, Haugeberg G, Lindland A, Myklebust G. The fast-track ultrasound clinic for early diagnosis of giant cell arteritis significantly reduces permanent visual impairment: towards a more effective strategy to improve clinical outcome in giant cell arteritis? Rheumatology (Oxford). 2016;55,1:66-70.

[3] Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, et al. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. Arthritis Rheum. 1990;33,8:1122-8.

[4] Mahr A, Saba M, Kambouchner M, Polivka M, Baudrimont M, et al. Temporal artery biopsy for diagnosing giant cell arteritis: the longer, the better? Ann Rheum Dis. 2006;65,6:826-8.

[5] Seeliger B, Sznajd J, Robson JC, Judge A, Craven A, et al. Are the 1990 American College of Rheumatology vasculitis classification criteria still valid? Rheumatology (Oxford). 2017;56,7:1154-1161.

[6] Tomelleri A, Campochiaro C, Sartorelli S, Farina N, Baldissera E, et al. Presenting features and outcomes of cranial-limited and large-vessel giant cell arteritis: a retrospective cohort study. Scand J Rheumatol. 2022;51,1:59-66.

[7] de Boysson H, Liozon E, Espitia O, Daumas A, Vautier M, et al. Different patterns and specific outcomes of large-vessel involvements in giant cell arteritis. J Autoimmun. 2019;103:102283.

[8] Craven A, Robson J, Ponte C, Grayson PC, Suppiah R, et al. ACR/EULAR-endorsed study to develop Diagnostic and Classification Criteria for Vasculitis (DCVAS). Clin Exp Nephrol. 2013;17,5:619-621.

[9] Dejaco C, Ramiro S, Duftner C, Besson FL, Bley TA, et al. EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice. Ann Rheum Dis. 2018;77,5:636-643.

[10] Luqmani R, Lee E, Singh S, Gillett M, Schmidt WA, et al. The Role of Ultrasound Compared to Biopsy of Temporal Arteries in the Diagnosis and Treatment of Giant Cell Arteritis (TABUL): a diagnostic accuracy and cost-effectiveness study. Health Technol Assess. 2016;20,90:1-238.

[11] Rhéaume M, Rebello R, Pagnoux C, Carette S, Clements-Baker M, et al. High-Resolution Magnetic Resonance Imaging of Scalp Arteries for the Diagnosis of Giant Cell Arteritis: Results of a Prospective Cohort Study. Arthritis Rheumatol. 2017;69,1:161-168.

[12] Soussan M, Nicolas P, Schramm C, Katsahian S, Pop G, et al. Management of large-vessel vasculitis with FDG-PET: a systematic literature review and meta-analysis. Medicine (Baltimore). 2015;94,14:e622.