Large vessel vasculitis: which imaging method?

Marco A Cimminona, Dario Camellinob

a Research Laboratory and Academic Division of Rheumatology, University of Genova, Genova, Italy
b Autoimmunology Laboratory, Department of Internal Medicine, University of Genova, Genova, Italy

Imaging has dramatically changed our understanding of large vessel vasculitides. They can be identified with ultrasound, 18F-fluorodeoxyglucose positron emission tomography (PET), combined or not with computed tomography (CT), and magnetic resonance imaging (MRI) [1]. With these techniques, large vessel vasculitides have also been recognised in patients presenting with only nonspecific signs or symptoms, such as malaise and fever, or increased inflammatory indices. Large vessel vasculitides are not a homogeneous group of conditions since they include several diseases in addition to giant-cell arteritis and Takayasu’s arteritis [2]: IgG4-related disease, rheumatoid arthritis, systemic lupus erythematosus, Behcet’s syndrome and Cogan’s syndrome can also affect the aorta and its branches. In their retrospective study published in Swiss Medical Weekly [3], Adler et al. showed a high positive predictive value (92%) of magnetic resonance angiography (MRA) of the aorta for diagnosing large vessel vasculitides. Their most striking finding was that glucocorticoid treatment for more than 5 days reduced the chance of classifying MRA as positive by 89.3%. In giant-cell arteritis, MRI is very effective for assessing inflammation of the temporal arteries, with a sensitivity of 93.6% (assuming temporal artery biopsy to be the reference standard) [4].

In this study, 51/75 patients who underwent MRA were judged as “imaging negative”. Six of these patients, however, were considered to have large vessel vasculitis on the basis of their clinical picture, and five showed positive histology of the temporal artery, despite “imaging negative”. Six of these patients, however, were considered to have large vessel vasculitis on the basis of their clinical picture, and five showed positive histology of the temporal artery, despite “imaging negative”. In a small and heterogeneous cohort of patients, a poor correlation between this score and clinical and laboratory parameters was found. The most important limitation of ultrasound, MRA and PET studies in large vessel vasculitis is the lack of histological confirmation of imaging findings: a definite diagnosis by an expert clinician is usually taken as reference standard [10], an approach representing “circular reasoning”. The cases, usually identified on the basis of clinical suspicion, are investigated with an imaging tool and the confirmation of the diagnosis (and therefore of the truthfulness of imaging findings) is then based on clinical evaluation, which is influenced by the results of imaging. Except for anecdotal reports [11], histological confirmation of large vessel vasculitis is not feasible in the vast majority of patients and most of our knowledge still relies on old necropsy studies [12].

A possible limitation of the study by Adler et al. is that it merged all patients with large vessel vasculitis. Takayasu’s arteritis and giant-cell arteritis are differentiated on the basis of age, but the debate as to whether their differences exceed their similarities is still open [13]. Moreover, isolated aortitis is an increasingly recognised entity, which should probably be separated from giant-cell and Takayasu’s arteritis, because it is characterised by younger age compared with giant-cell arteritis and a higher risk of aortic damage requiring surgery. Isolated aortitis has been classified separately in the latest Chapel Hill Consensus Conference as “single organ” vasculitis [2], but many reports consider it together with other large vessel vasculitides. The scenario is further complicated by the observation of large vessel vasculitis at imaging in at least one third of patients with apparently clinical isolated polymyalgia rheumatica [14], raising the doubt whether it represents an incomplete form of giant-cell arteritis or a disease per se. Giant-cell arteritis, Takayasu’s arteritis, vasculitis associated with polymyalgia rheumatica and isolated aortitis share pathological characteristics, but evidence for considering these forms of large vessel vasculitis a single entity is still lacking. As a consequence, each of these diseases should probably be studied separately.

In the end, which is the best imaging method for detecting large vessel vasculitis? The answer has yet to come. A PubMed search of the terms “large vessel vasculitis AND positron emission tomography” performed on 23 October 2016 yielded 95 results published in the last five years, in comparison with 83 for “large vessel vasculitis AND magnetic resonance imaging”. This indicates that both tech-
niques continue to be under the spotlight, at least in the research field.

Disclosure statement
No financial support and no other potential conflict of interest relevant to this article was reported.

Correspondence
Marco Arnedo Cimmino, MD
Università di Genova
Viale Benedetto XV, 6
i-16132 Genova
cimmino[at]unige.it

References