

Large vessel vasculitis

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The large vessel vasculitides, Takayasu's arteritis (TAK) and giant cell arteritis of the elderly (GCA), have traditionally been classified as distinct disorders, primarily based on the ethnicity and age of affected individuals. However, their indistinguishable histopathologic findings have led some to question the discrimination between these diseases. Hall proposed that TAK, GCA, and polymyalgia rheumatica (PMR) composed an "unholy trinity"—representing a continuum of disease (1). However, subsequent studies concluded that these diseases were readily distinguished on the basis of vascular manifestations, symptoms, age and ethnicity (2-4). Further study has led us to challenge these assumptions and suggest that they are more similar than previously appreciated.

Although often reported to be a disease found primarily in young, Far Eastern females, TAK has been identified worldwide. Furthermore, US and Italian cohorts are primarily composed of Caucasian women, and studies from India document a significant prevalence of TAK in males (5-8). Although age is considered a diagnostic criterion in the US, other ethnic cohorts have been less stringent in this regard, with several reporting > 15% of their cohort over the age of 40 at the time of presentation (6, 9). In addition, multiple cohort studies document GCA symptoms in TAK, including headache, visual disturbances, and polymyalgias, albeit at a lower prevalence.

The earliest reports of GCA emphasized temporal artery involvement (temporal arteritis) and others later expanded observations to GCA affecting the extracranial branches (cranial arteritis) of the carotid arteries (10). This limited view of the range of vascular damage changed following publication of a meticulous autopsy study of large vessel vasculitis by Ostberg in 1972. By removing the entire aorta and major branch vessels to the elbows and knees and sectioning the specimens at 2 cm intervals, she found evidence of large artery involvement in all subjects with GCA, similar to her findings in TAK (11). The advent of PET scanning, computed tomography and magnetic

resonance vascular imaging has reinforced the notion that involvement of aortic branch vessels in GCA is common, although the frequency of clinically significant vascular stenoses and aneurysms is less than in TAK (12). In a longitudinal cohort study, we found that 72% of GCA (31/43) patients who had vascular imaging studies had large artery vasculitis, most frequently of the aorta and subclavian and axillary arteries (13).

Novel technologies have provided new insights into the large vessel vasculitides that have caused us to reconsider the conventional classification of these diseases. It is possible that bias in our approach to data collection and physical examination has constrained our ability to assess disease manifestations equally in patients with large vessel vasculitis. Further prospective study is necessary to validate the consideration of these disorders as a spectrum of a single disease.

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