Positive Temporal Artery Ultrasound and Biopsy After 5 Months of High-Dose Glucocorticoids

To the Editor:

We read with interest the article by Majerovich et al describing the duration of glucocorticoid (GC) therapy and temporal artery biopsy (TAB) positivity in giant cell arteritis (GCA).1 Their results are noteworthy, indicating GC therapy does not appear to significantly affect TAB positivity to at least 6 weeks; findings that the authors noted are also supported by our institution’s experience.2 It is our group’s opinion that GCA should be confirmed by biopsy and/or imaging (temporal artery or large-vessel), and patients with clinical diagnosis should be restricted to those with characteristic features for whom appropriate investigations were negative or nondiagnostic and alternative etiologies were excluded.

Current GCA management consensus guidelines by the American College of Rheumatology (ACR) conditionally recommend obtaining a TAB within 2 weeks of starting oral GCs, compared to waiting longer than 2 weeks.3 We agree with Majerovich and colleagues1 that TAB, particularly in patients with cranial symptoms, can still be diagnostic beyond 2 weeks, especially if other objective methods of disease confirmation have not yet been attempted, or were done and were inconclusive. Temporal artery ultrasound (TAUS) has replaced TAB in many centers as the first-line tool to evaluate patients for possible GCA. The sensitivity of TAUS for the diagnosis of GCA, however, declines more rapidly in comparison to TAB, with some studies showing a reduction from 92% sensitivity with 0 to 1 days of GC to 80% on days 2 to 4, and down to 50% after > 4 days.4 It is noteworthy that the ACR consensus statements do not yet provide a suggested timeline or cut-off for optimal evaluation if TAUS is performed.5 European guidelines, which conversely favor TAUS over TAB, recommend obtaining TAUS within 1 week of GC initiation to assist with increased sensitivity.6 Herein we present an illustrative case in which both TAUS and TAB were positive after 5 months of GC in a patient with initial clinical diagnosis of GCA but incomplete response to long-term high-dose GC.

A 70-year-old male presented to a local neurology clinic with a 6-month history of 20-pound unintentional weight loss and 1-month history of progressive bilateral headache, temporal artery tenderness, and prominent temporal artery vessels (Figure 1A). Erythrocyte sedimentation rate (ESR) was 30 mm/h and C-reactive protein (CRP) was 13.1 mg/L prior to GC. Oral prednisone 60 mg/day was initiated for presumption of GCA and resulted in initial symptom reduction. Neither TAB, TAUS, nor large-vessel angiography were obtained initially. He was unable to taper prednisone < 25 mg/day because of recurrence of headache, scalp tenderness, and bilateral leg pain. Five months after initiation of GC, he presented for a second opinion, still on 25 mg/day of prednisone.

On presentation, physical examination was notable for ongoing bilateral enlargement of the temporal arteries (Figure 1B) despite normal CRP (< 3.0 mg/L) and ESR (5 mm/h). Serum IgG4 was within normal limits. Antineutrophil cytoplasmic antibodies and monoclonal gammopathy screen were negative. TAUS was performed and showed circumferential wall thickening with halo sign involving both the frontal and parietal branches of both temporal arteries (Figures 1C,D). TAB was pursued and confirmed histopathologic features consistent with GCA (Figure 1E-G). Computed tomography (CT) with angiography of the chest identified an ascending aortic aneurysm of 48 mm without evidence of wall thickening.18F-fluorodeoxyglucose positron emission tomography/CT was negative for arterial hypermetabolism and no features of neoplasia were observed. Due to refractory symptoms and requirement of chronic GC therapy, he was started on tocilizumab (TCZ) 162 mg subcutaneous weekly. Cranial symptoms improved and he was able to subsequently taper off GC in 6 months without return of cranial features. Due to ongoing clinical remission, his local care team optimized the TCZ dose after 12 months of weekly administration to a frequency of every 14 days.

This case highlights some key concepts. First, it reinforces that in select cases of GCA, persistent vasculitis can be detected by both TAUS and TAB in a longer time frame than typical of suggested consensus thresholds. Second, even in the context of normalization of the systemic inflammatory response, localized arterial inflammation can exist despite prolonged use of high-dose GC. Third, confirmation of disease, specifically in patients with atypical presentations or lack of expected therapeutic response is critical because non-GCA alternative diagnoses such as polyarthritis nodosa, antineutrophilic cytoplasmic antibody–associated vasculitis, IgG4-related disease, and lymphoma, among others, can result in inflammation of the temporal arteries, resulting in lack of typical response to GC. In such circumstances, histopathologic findings on TAB may hold key additional information to help make the correct diagnosis and subsequently guide modification of treatment approaches.6,9 Although TAUS is a reasonable first-line tool for evaluation of suspected GCA, we agree with Majerovich et al1 that TAB remains an important diagnostic study and the lack of ability to perform TAB within 2 weeks of GC use should not be a barrier to pursuing TAB.

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