Giant cell arteritis

Renatta Varma and Anil Patel remind us of the variability in presentation of giant cell arteritis (GCA).1 Another patient's story presents yet another pitfall in GCA diagnosis.

A 76-year-old woman presented to the emergency department with right temporal "head pain." Her erythrocyte sedimentation rate was 65 mm/h. The diagnosis was "headache, rule out temporal arteritis." Prednisone, 40 mg per day, was prescribed, and the patient was referred for follow-up examination. Four days later, the patient's tenderness to percussion over the right temporal area persisted, and I increased the prednisone dosage. A biopsy was performed 2 days later.

The biopsy results, available 1 week later, showed no evidence of GCA, so the ophthalmologic surgeon stopped the patient's prednisone. The patient returned to me 4 days later with increasing "head pain." We discussed the possibility of false-negative biopsy findings, and I prescribed prednisone again (and osteoporosis prophylaxis). A consultation was arranged with a rheumatologist who concurred with the diagnosis of biopsy-negative CGA and continued to treat the patient accordingly.

Although the only confirmatory test for GCA is a positive biopsy, nondiagnostic biopsy specimens do not exclude the diagnosis. It is commonly accepted that, because of the patchy involvement of the arteries, biopsies may be nondiagnostic in many patients. Thus, because biopsies are invasive, some authors even suggest that biopsy may not be necessary.2,3

Furthermore, corticosteroid therapy, which should be started without delay, rapidly reduces the chance of a positive biopsy result.2 One week of corticosteroid treatment may reduce the chance of obtaining a positive biopsy to 10%.4 Therefore, biopsy should be performed within the first few days of therapy.⁴

Gary N. Fox Defiance Clinic Toledo, Ohio

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Renatta Varma and Anil Patel recently discussed herpes zoster (varicella zoster virus) infection as a differential diagnosis of GCA. Interestingly, 88% of arteries with histologically confirmed GCA have been found to harbour herpes simplex virus DNA.2 Consideration of both herpes zoster and herpes simplex as a cause in this case of necrotic scalp lesions may therefore be warranted.

Heike Varnholt

Department of Pathology Baystate Medical Center Springfield, Mass.

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[The authors respond:]

We concur with Gary Fox that if the clinical suspicion of GCA is high this condition should still be considered even if the biopsy results are nondiagnostic, especially because of its potential to cause devastating bilateral and irreversible blindness.1 However, we would caution against diagnosing giant cell arteritis without biopsy evidence (biopsy-negative GCA) because of the potential detrimental side effects of long-term steroid use.

Skip lesions are possible,^{2,3} but we find it useful to obtain a biopsy specimen at least 2-3 cm long and to ensure that the pathologist serially examines

the entire length of the specimen. If clinical suspicion remains despite a negative unilateral biopsy, then we recommend a contralateral biopsy as soon as possible, regardless of the time that has elapsed. Although steroid administration can affect a biopsy result, we have had numerous patients for whom a biopsy result was positive even after several weeks or months of steroid therapy. Furthermore, we consider temporal artery biopsy to be a relatively safe and simple office procedure. If 2 temporal artery biopsies are negative for GCA, we consult an internist to evaluate the patient's erythrocyte sedimentation rate or C-reactive protein level, or both, to search for a systemic infection or malignancy.

Anil D. Patel

Clinical Associate Professor Dean A. McGee Eye Institute Oklahoma City, Okla.

Renatta Varma

Fellow

Vitreoretinal Surgery Victoria, BC

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Corrections

The DOIs for a recent commentary and a recent review article2 should have read 10.1503/cmaj.051291 and 10.1503/cmaj .050141, respectively.

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