Letter to the Editor

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Letter by de Boysson and Pagnoux Regarding Article, “Diagnostic Yield and Safety of Brain Biopsy for Suspected Primary Central Nervous System Angiitis”

To the Editor:

We read with interest the recent study by Torres et al1 on the diagnostic value and risks of brain biopsy in patients with suspected primary angiitis of the central nervous system (PACNS). Brain biopsy revealed PACNS in 11% of patients and showed perivascular infiltrate in 18% and alternative diagnoses in 30%. No baseline clinical, surgical, or imaging characteristics predicted a positive biopsy. Postbiopsy complications affected 16% of patients.

Findings from the other rare cohorts of PACNS patients not only are somewhat in agreement but also add some complementary information, which we think could be of interest to readers.2,3 In our previous report on the first 52 patients with a final diagnosis of PACNS enrolled in the French cohort, initiated in 2010 using the well-established network of the French Vasculitis Study Group, 19 of 31 (61%) who underwent brain biopsy had positive results.2 The cohort has now enrolled 109 patients. All have demonstration of vessel involvement on positive biopsy, digital subtraction angiography or magnetic resonance angiography. Among these 109 patients, 51 had a brain biopsy: 34 (67%) with vasculitis, 7 (13%) with a perivascular infiltrate, and 10 (20%) with negative results (and no alternative diagnoses).

Open-wedge biopsy was performed in 42 patients and revealed vasculitis in 26 (62%), whereas 9 patients underwent a stereotactic procedure that showed vasculitis in 8 (89%). Lesions identified on brain magnetic resonance imaging guided all stereotactic procedures and 25 of the 42 open-wedge biopsies. Altogether, 26 of 34 (76%) neuroimaging-guided biopsies were diagnostic. Seventeen patients had samples from a nonlesion site on magnetic resonance imaging (in the frontal area of the nondominant hemisphere), and 8 (47%) showed vasculitis. Six (12%) patients experienced a complication after the procedure (2 abscesses, 2 transient focal deficits, 1 transient fever, and 1 postoperative seizure).

When compared with patients with negative biopsy, patients with positive biopsy had, at disease onset, more seizures (65% versus 18%; P=0.001), abnormal cerebrospinal fluid analysis (ie, high protein level rate or leukocyte count; 87% versus 59%; P=0.03), and gadolinium enhancements (77% versus 20%; P=0.0003). Conversely, they showed less focal deficits (56% versus 100%; P=0.001) and less demonstration of vascular involvement on neurovascular imaging (on digital subtraction angiography [26% versus 94%] or magnetic resonance angiography [19% versus 82%]; P<0.0001 for both procedures).

Hence, our study indicates that the yield of a brain biopsy was higher in patients with predominant involvement of small vessels; three quarters of patients with a positive biopsy had negative findings on neurovascular imaging, suggesting isolated small-vessel PACNS. To increase the likelihood of obtaining a positive biopsy, some authors advocate targeted biopsy of lesions observed on imaging.4 Our study seems to confirm this as more than three quarters of our patients who had imaging-guided biopsies had a positive biopsy. However, nonlesion biopsy of the nondominant hemisphere’s frontal area may still be diagnostic. Sampling leptomeningeal tissue during the biopsy, especially in patients with gadolinium-enhanced lesions, may also raise diagnostic sensitivity.3,4

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Disclosures

None.

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