

## INSTRUCTIONS FOR LETTERS TO THE EDITOR

Editorial comment in the form of a Letter to the Editor is invited. The length of a letter should not exceed 800 words, with a maximum of 10 references and no more than 2 figures or tables; and no subdivision for an abstract, methods, or results. Letters should have no more than 4 authors. Financial associations or other possible conflicts of interest should be disclosed.

Letters should be submitted via our online submission system, available at the Manuscript Central website: http://mc.manuscriptcentral.com/jrheum For additional information, contact the Managing Editor, The Journal of Rheumatology, E-mail: jrheum@jrheum.com

## Nonsystemic Vasculitic Neuropathy: A Clinicopathological Study of 22 Cases

To the Editor:

We read with great interest the recent article by Kararizou,  $et al^1$  about the clinicopathological study of nonsystemic vasculitis neuropathy, and the accompanying editorial by Michael Collins<sup>2</sup>.

In providing medical care for more than 1500 patients with different kinds of vasculitides, we similarly see patients with nonsystemic vasculitic neuropathy. We were therefore amazed, particularly with regard to the editorial, that a comment about the importance of antineutrophil cytoplasmic antibody (ANCA) testing in these clinical conditions was missing, above all since the manuscript was published in a rheumatology journal. Did Kararizou, *et al* really omit to determine the ANCA status in this study population?

In contrast to the outlined standarized procedure, we would emphasize that ANCA testing should be included in the investigation in patients with undefined neuropathy. Sensory symptoms, pain, and paresthesia might be *the* initial or sole symptom of ANCA-associated vasculitides (AAV)<sup>3</sup>. We were able to show in one of the largest cohort studies of patients with Wegener's granulomatosis, the classic example of AAV, that peripheral nervous system involvement was present in 42% of subjects (Table 1)<sup>4</sup>. Importantly, more than one-third of these cases developed symptoms even before the diagnosis of AAV.

In other forms of AAV, such as Churg-Strauss syndrome, involvement of the peripheral nervous system is even more frequent and seems to be a typical early manifestation, in contrast to vasculitis of the central nervous system<sup>5</sup>. If a vasculitic neuropathy is diagnosed, interdisciplinary surveillance (e.g., ENT, ophthalmologist, and nephrologist) might uncover additional (subclinical) vasculitic manifestations.

In the presented study<sup>1</sup>, additional ANCA testing, as well as an interdisciplinary approach to the patients with a vasculitic neuropathy, might have classified some patients as AAV. The exact diagnosis and the actual disease extent can be decisive for the prognosis, especially as the patients with AAV and vasculitic neuropathy seem to have a more severe disease course<sup>6</sup>.

*Table 1.* Pathophysiological characteristics of peripheral neuropathy in 56 patients with generalized Wegener's granulomatosis. Reprinted with permission, from *Arch Neurol* 2001; 45:1215-21<sup>6</sup>.

	No. (%) of Patients
All patients with peripheral neuropathy	56 (100)
Symmetrical sensorimotor polyneuropathy	31 (44)
Mononeuritis multiplex	25 (44)
Affected nerves	
Peroneal	23
Tibial	6
Sural	2
Ulnar	6
Median	3
Radial	2
Axonal neuropathy	41 (73.2)
Demyelinating neuropathy	2 (3.6)
Both types of lesion present	9 (16.1)
Acute or subacute onset	25 (44.6)
Chronic prolonged onset	28 (50)

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## Dr. Kararizou replies

To the Editor:

We thank Drs. Aries and Gross for their comments on our article<sup>1</sup>.

They emphasize the importance of antineutrophil cytoplasmic antibody (ANCA) testing as a serological marker associated with vasculitis and consequently as a differentiating factor of nonsystemic versus systemic vasculitis

We acknowledge the importance of their comment, as nonsystemic and Wegener's vasculitis both affect the small epineurial blood vessels<sup>2-5</sup>. As noted in our article, the cellular composition of the inflammatory infiltrates appears to be similar in both nonsystemic vasculitic neuropathy and in the systemic form (especially in Wegener's) and so, in the case where granuloma formation is absent, this can be confusing<sup>2-5</sup>.

As our study is a retrospective one, spanning a period of 20 years, ANCA testing was not available at the time some of our patients were examined. Thus we can only report the results concerning 17 out of 22 patients, which were negative for perinuclear and cytoplasmic ANCA.

We are now completing a study in a large group on systemic vasculitic

polyneuropathy; the study comprises patients with positive ANCA, many of them having prominent or exclusively sensory symptoms.

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# Bilateral Ocular Myositis as a Late Complication of Dermatomyositis

To the Editor:

Dr. Kokotis and colleagues have given an excellent description of an interesting clinical case<sup>1</sup>. It was recognized as bilateral ocular myositis based on the clinical features and orbital magnetic resonance imaging (MRI) show-

ing enlargement of the recti extraocular muscles (their Figure 1). Nevertheless, if we make a closer inspection of Figure 1, it will not be difficult to find that only coronal sections of the orbital MRI were presented, but the important radiological section, an axial section, of the involved recti muscles was missing. Axial imaging plays a key role in diagnosis, absence of which may blur the distinction between this case and thyroid eye disease<sup>2,3</sup>.

The clinical manifestation of ocular myositis may sometimes be vague and difficult to distinguish from thyroid eye disease or Graves' ophthalmopathy<sup>2</sup>. Both these conditions may produce extraocular muscle enlargement on orbital imaging; but characteristically, extraocular muscle enlargement of thyroid eye disease tends to be tendon-sparing, whereas in ocular myositis, the inflammatory change is more diffuse and involves the whole muscle bellies and tendons<sup>2,3</sup>. This marked imaging disparity is best appreciated from the axial sections of orbital scans<sup>2</sup>. Apart from computerized tomography or MRI orbital imaging, ophthalmic echography (B-mode ultrasonogram) may act as a supplement to the clinical difficulty in distinction between the two.

Surprisingly, given that thyroid eye disease is the most common cause of bilateral proptosis in adults, the authors have apparently given no consideration of it as a differential during the clinical investigation. In the interest of readers, the authors are encouraged to provide more information for this patient, particularly investigatory results such as axial MRI of the orbit and thyroid function tests.

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