Towards diagnostic criteria for the ANCA-associated vasculitides

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Key words: Vasculitis, classification, Wegener’s granulomatosis, microscopic polyangiitis, ANCA.

The primary systemic vasculitides are an important cause of mortality and morbidity in the community. Their incidence is >100 new cases per million per year (1). There are significant benefit with intensive chemotherapy but if the diagnosis is delayed, this may adversely affect the outcome. Up to 25% of patients on chronic haemodialysis programmes may have had undiagnosed systemic vasculitis as a major contributor to renal failure (2).

Classification criteria are useful to confirm that a group of patients with a clinical diagnosis have a similar or identical condition. However, in order to discriminate between patients with or without a specific disease, diagnostic criteria are required. There are currently no validated diagnostic criteria for primary systemic vasculitis although there are classification criteria that have been misapplied – the American College of Rheumatology (ACR) criteria (3) and the Chapel Hill Consensus Conference (CHCC) Definitions (4). When tested, the CHCC definitions supplemented with surrogate clinical and laboratory parameters failed to act as diagnostic criteria (5). Diagnostic criteria for Behçet’s disease, which is classified as a secondary vasculitis, have been developed for use in clinical trials (6).

Diagnostic criteria for primary systemic vasculitides would be most useful for practising clinicians who could rapidly and accurately assess the probability of vasculitis in the context of patients who present to them acutely, especially those with multi-system illnesses. There are currently no satisfactory serological tests to enable a rapid diagnosis of vasculitis. The ANCA test, when used by non-specialists has a diagnostic yield of only 0-12% for systemic vasculitis in a routine clinical setting (7). At present, ANCA testing is not part of the existing classification criteria for vasculitis. The considerable overlap of disease features across the ANCA related vasculitides is further justification to re-evaluate diagnostic and classification criteria (8).

References

Table I. Limitations of current definitions and classification of ANCA associated vasculitides.

<table>
<thead>
<tr>
<th>Disease</th>
<th>Current classification(s)</th>
<th>Problems</th>
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</thead>
<tbody>
<tr>
<td>Wegener’s granulomatosis</td>
<td>CHCC definition – Small vessel vasculitis ACR classification – Wegener’s granulomatosis</td>
<td>Overlap with MPA; range of organ involvement Some aspects are not vasculitic (eg nasal granulomata)</td>
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<tr>
<td>Microscopic polyangiitis</td>
<td>CHCC definition – Small vessel vasculitis ACR classification – no definition</td>
<td>Renal only variety may evolve into WG or MPA</td>
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<tr>
<td>Churg Strauss Syndrome</td>
<td>CHCC definition – Small vessel vasculitis ACR classification – Churg Strauss syndrome</td>
<td>Overlap with WG; renal disease less common than for WG and MPA; some aspects are non-vasculitic (eg allergies)</td>
</tr>
<tr>
<td>Unspecified small vessel vasculitis</td>
<td>CHCC definition – Unspecified small vessel vasculitis ACR classification – Unspecified small vessel vasculitis</td>
<td>May overlap features of MPA/WG/ CSS or be more limited; less well studied; some patients remain unclassifiable and should not be “forced” into an artificial system to describe the condition</td>
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