Pulmonary fibrosis in patients with positive neutrophil cytoplasmic antibodies vasculitis

Sirs,
ANCA-positive vasculitis is a small-vessel systemic necrotising vasculitis (1). The most common presentation is pulmonary-renal syndrome, with necrotising glomerulonephritis and alveolar haemorrhage. In recent years, intestinal lung disease, often in the form of pulmonary fibrosis has been reported in patients with positive ANCA. Three patients with PF and ANCA-positive vasculitis are reported.

Case 1: A 72-year-old man, with a pulmonary fibrosis diagnosed by HRCT at age 60, was admitted with polyarthritis of the hands. He had peripheral neuropathy and Raynaud’s phenomenon. The immune was admitted for polyarthritis of the hands. The study highlighted a small-medium vessel vasculitis. Treatment with rituximab was initiated with good clinical evolution, remaining stable at present.

Pulmonary fibrosis is a rare clinical manifestation that has been associated with ANCA-positive vasculitis (2). In our case, pulmonary fibrosis can appear before, concomitantly or after the diagnosis of vasculitis (7). Concurrent diagnosis is the most common presentation (2, 6, 8). The pathogenesis remains unclear, although different mechanisms have been proposed. One of them is the presence of repeated episodes of subclinical alveolar haemorrhage secondary to diffuse capillaritis (6-7). Another mechanism is related to the increased oxidative stress and proinflammatory response against neutrophil cytoplasmic antibodies, specifically the anti-myeloperoxidase antibodies (2, 5, 8). The fibrotic reaction would be the reparative response to lung damage caused by these mechanisms. Some authors have proposed to consider that the interstitial lung affection as a limited form of microscopic polyangiitis (9).

In recent years around 131 clinical cases of pulmonary fibrosis and positive ANCA vasculitis have been published (3-4, 7). The average age of diagnosis of ANCA vasculitis is 68 years (10). The average age of our cases (Table I) is similar to those previously published. Multiple studies observed that pulmonary fibrosis regards a worse prognosis with the presence of positive anti-MPO antibody (7). Survival at 5 years in patients with ANCA vasculitis is about 60% and is reduced to 29% when it is associated with pulmonary fibrosis (3). One of our patients died at six years of diagnosis of ANCA vasculitis because of progression of lung disease. Unlike previous studies described, he had no positive anti-MPO antibodies as a poor prognostic factor. The other patient also died as a result of pneumonia, while the third patient remains clinically stable.

In conclusion, we wanted to point out the importance of pulmonary fibrosis as a clinical manifestation of patients with ANCA positive vasculitis, because it can help for the diagnosis and the treatment and may lead to improve the outcome of these patients.

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References

Table I. Main characteristics of patients.

<table>
<thead>
<tr>
<th>Gender</th>
<th>Age (years)</th>
<th>Clinical manifestations</th>
<th>Pulmonary fibrosis</th>
<th>ANCA pattern</th>
<th>Anti-MPO</th>
<th>Diagnostic orientation</th>
<th>Treatment</th>
<th>Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>M</td>
<td>72</td>
<td>Arthritis and renal involvement</td>
<td>Previous (3 years)</td>
<td>pANCA</td>
<td>Yes</td>
<td>MPA (renal biopsy)</td>
<td>CS+CYC, AZA</td>
<td>Death at 3 years</td>
</tr>
<tr>
<td>M</td>
<td>61</td>
<td>Arthritis, cutaneous involvement and peripheral neuropathy</td>
<td>Simultaneous</td>
<td>cANCA</td>
<td>No</td>
<td>ANCA-positive vasculitis (m/n biopsy)</td>
<td>CS+AZA, MMF</td>
<td>Death at 6 years</td>
</tr>
<tr>
<td>F</td>
<td>71</td>
<td>Arthritis, renal and cutaneous involvement</td>
<td>Previous (2 years)</td>
<td>pANCA</td>
<td>Yes</td>
<td>MPA (cutaneous biopsy)</td>
<td>CS+AZA, RTX</td>
<td>Clinical stability at 2 years</td>
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