Churg-Strauss Syndrome Complicated by Chronic Periaortitis: A Case Report and Review of the Literature

Kenji Fujii¹ and Yuji Hidaka²

Abstract

We present a case of Churg-Strauss syndrome complicated by chronic periaortitis. A 68-year-old man presented with wheezing, dyspnea, purpurae, and numbness of the extremities. Antineutrophil cytoplasmic antibodies were absent; however, eosinophilia, a pulmonary infiltrative shadow on chest X-ray, eosinophilic vasculitis on histologic examination of skin and kidney, and mononeuritis multiplex were detected. Churg-Strauss syndrome was diagnosed. Contrast-enhanced abdominal computed tomography revealed a periaortic soft tissue mass extending from the subphrenic abdominal aorta to the proximal area of the bilateral iliac arteries. This indicated chronic periaortitis, probably caused by vasculitic activities. Both disorders improved with steroid therapy.

Key words: Churg-Strauss syndrome, chronic periaortitis, IgG4-related disease


Introduction

Churg-Strauss syndrome is a rare form of eosinophilic vasculitis affecting the small- to medium-sized blood vessels with diffuse involvement of cardiovascular organs. The large blood vessels are reportedly not affected (1). Chronic periaortitis is a fibroinflammatory disease which spreads from the abdominal aorta to the iliac arteries and is reportedly associated with systemic autoimmune diseases (2). Here we report a case of Churg-Strauss syndrome complicated by chronic periaortitis and present a review of the literature describing vasculitis in small- and medium-sized blood vessels complicated by periaortitis. We also discuss antineutrophil cytoplasmic antibody (ANCA)-positive periaortitis.

Case Report

A 68-year-old man with a history of chronic rhinitis presented with wheezing, effort-related dyspnea, appetite loss, fatigue, purpurae, and numbness of the extremities. On physical examination, he was afebrile with normal blood pressure; no laterality of the upper arms was observed. Lung auscultation revealed scattered, expiratory wheezes and coarse crackles over the right lower lobe. The abdomen was unremarkable. Palpable purpurae were observed extending from the left thorax to the anterior aspect of the abdomen as well as on the left forearm and the anterior surface of both legs. Neurological examination revealed bilaterally symmetrical weakness of the triceps surae muscle. Urinalysis revealed 1+ proteinuria and 3+ occult blood. Hematologic test results were as follows: hemoglobin level, 9.3 g/dL; white blood cell count, 10,200/μL with 56% neutrophils, 21% lymphocytes, 3% monocytes, 20% eosinophils, and 0% basophils; platelets, 230×10³/μL; erythrocyte sedimentation rate, 124 mm/h; C-reactive protein, 2.46 mg/dL; urea nitrogen, 21 mg/dL; and creatinine, 1.2 mg/dL. Immunoglobulin (Ig) G level was elevated to 3,558 mg/dL and rheumatoid factor was elevated to 54.8 IU/mL. Tests for the presence of antinuclear, anti-double-stranded DNA, anticardiolipin, antitiglomerular basement membrane, and ANCA all proved negative, as were the results of an enzyme-linked immunosorbent assay for antibodies to proteinase 3 and myeloperoxidase. Neurophysiological examination revealed multiple

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Received for publication June 11, 2011; Accepted for publication September 29, 2011

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mononeuropathies in the right median, bilateral tibial, and peroneal nerves. Chest X-ray revealed bilateral reticular opacities and right pleural effusion. Thoracentesis yielded yellow fluid containing 1625 white blood cells/μL with 43% neutrophils, 36% lymphocytes, and 21% eosinophils. Electrocardiogram and cardiac ultrasonography results were normal. Abdominal contrast-enhanced computed tomography (CT) revealed a periaortic soft tissue mass extending from the subphrenic abdominal aorta to the proximal area of the bilateral common iliac arteries (Fig. 1Aa and 2Aa). A skin biopsy revealed leukocytoclastic vasculitis with predominantly eosinophilic invasion without necrotizing vasculitis. A renal biopsy revealed severe perivascular inflammatory cell infiltration with eosinophils around the renal arterioles and renal interstitium; however, crescent formation or necrotizing angitis was not seen (Fig. 3).

The patient fulfilled the 1990 American College of Rheumatology classification for Churg-Strauss syndrome (3) with the following criteria: asthma, >10% increase in eosinophils, a pulmonary infiltrative shadow on chest X-rays, eosinophilic vasculitis on histologic examination of skin and kidney, and mononeuritis multiplex. None of the temporal and cervical arteries, the thoracic aorta, or their branches was involved; chronic periaortitis was diagnosed.

Treatment was initiated with 60 mg/day prednisone, which led to an improvement in asthma, eosinophilia, cutaneous symptoms, and renal dysfunction; subsequently, the pulmonary infiltrative shadow almost completely disappeared. However, a slight numbness of the lower extremities persisted. Nevertheless, there was no onset of abdominal symptoms or ureteric obstruction. The perivascular lesion involving the abdominal aorta was significantly improved within 2 months of treatment (Fig. 1Bb and 2Bb); thereafter, his clinical course was favorable.

**Discussion**

Clinically and histologically, the patient described in this case report had a typical presentation of Churg-Strauss syndrome. His eosinophilia, pulmonary invasion, and renal dysfunction were promptly relieved by steroid therapy without any exacerbation in clinical course. Although aortic lesions

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Figure 1. Contrast-enhanced computed tomography (CT) at the level of the first lumbar vertebra. Arrows designate the A ring of an enhanced, periaortic, soft tissue mass is observed (arrow) on initial contrast-enhanced CT (Aa). Improvement and contraction of the ring two months after initiation of therapy (arrow) (Bb).

Figure 2. Contrast-enhanced computed tomography (CT) above the bifurcation of the common iliac artery. Arrows designate the A ring of an enhanced, periaortic, soft tissue mass is observed (arrow) on initial contrast-enhanced CT (Aa). Improvement and contraction of the ring two months after initiation of therapy (arrow) (Bb).
are not usually associated with Churg-Strauss syndrome (1), the improvement of the aortic lesion with steroid therapy as observed in this case suggests a possible association of the lesion with this syndrome. Furthermore, the patient had no evidence of large-vessel vasculitis, and the lesion distribution was mainly from the abdominal aorta to the iliac arteries. This was indicative of chronic periaortitis. Chronic periaortitis usually encompasses idiopathic retroperitoneal fibrosis, inflammatory abdominal aortic aneurysms, and perianeurysmal retroperitoneal fibrosis (4). The definitive diagnostic test for chronic periaortitis is defined as the presence of a periaortic soft tissue mass forming a rind of abdominal tissue around the aorta, with a very extensive spread demonstrated by contrast-enhanced CT scanning or magnetic resonance imaging (5).

Table 1. Reports of Vasculitis of the Small- and Medium-Sized Blood Vessels Vasculitis Complicated by Periaortitis and Antineutrophil Cytoplasmic Antibodies (ANCA)-Positive Periaortitis

<table>
<thead>
<tr>
<th>Disease</th>
<th>Cases</th>
<th>C-ANCA:</th>
<th>P-ANCA:</th>
<th>ANCA-positive:</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wegener granulomatosis</td>
<td>5</td>
<td>3</td>
<td>1</td>
<td>1</td>
<td>6, 7, 8</td>
</tr>
<tr>
<td>Cryoglobulinemia associated with hepatitis C</td>
<td>1</td>
<td>ND</td>
<td></td>
<td></td>
<td>11</td>
</tr>
<tr>
<td>Henoch–Schönlein purpura</td>
<td>1</td>
<td>ND</td>
<td></td>
<td></td>
<td>12</td>
</tr>
<tr>
<td>Polyarteritis nodosa</td>
<td>2</td>
<td>ND</td>
<td></td>
<td></td>
<td>13, 14</td>
</tr>
<tr>
<td>Churg–Strauss syndrome</td>
<td>1</td>
<td>negative</td>
<td></td>
<td></td>
<td>17</td>
</tr>
<tr>
<td>ANCA-positive periaortitis</td>
<td>7</td>
<td>1</td>
<td>6</td>
<td></td>
<td>2, 15, 16</td>
</tr>
</tbody>
</table>

Abbreviations: ANCA, antineutrophil cytoplasmic antibodies; C-ANCA, cytoplasmic antineutrophil cytoplasmic antibody; P-ANCA, perinuclear antineutrophil cytoplasmic antibody; ND, not done.

The pathogenesis of chronic aortitis mainly involves an exaggerated inflammatory response to advanced atherosclerosis, with systemic autoimmunity as a contributing factor (2). In previous study of with idiopathic aortitis patients (6), systemic autoimmune diseases, including systemic lupus erythematosus, rheumatic fever, large-vessel vasculitis (Takayasu arteritis/giant cell arteritis), and small- and medium-vessel vasculitis (Wegener granulomatosis/polymarteritis nodosa), were found in 16 of 52 patients (31%). There have been reports (Table 1) of vasculitis of the small and medium blood vessels complicated by periaortitis (6-14) as well as reports of ANCA-positive periaortitis (2, 15, 16); furthermore, ANCA is reportedly considered a contributing factor in cases of ANCA-positive, small-vessel vasculitis with periaortitis (8, 9, 15). However, to our

Figure 3. Hematoxylin and Eosin stained section of the skin biopsy specimen showing leukocytoclastic vasculitis with eosinophilic invasion (A), and that of the renal biopsy specimen showing severe perivascular inflammatory cell infiltration with eosinophils around the renal arterioles and renal interstitium (B) in Hematoxylin and Eosin staining.
knowledge, only one case of Churg-Strauss syndrome complicated by chronic periaortitis has been previously reported, and that case was negative for ANCA (17). Vaglio et al (2) reported that ANCA-positive chronic periaortitis was seen in only 3 of 16 patients (19%) in their study; of the remaining 13 ANCA-negative cases, abdominal vascular lesions with necrotizing vasculitis of small blood vessels in the vasa vaso-

rum were detected in 9 (69%). Histological vasa vasoritis of the aorta with granulomatous and necrotizing angiitis has been documented in a previous report (17). Therefore, the periaortitis described in the present case may also be related to the immunopathology of small blood vessels around the aorta. The periaortitis was successfully treated without the development of an aortic aneurysm or ureteric obstruction, although eosinophilic infiltration of the vasa vasorum was unclear in our case.

In addition, an elevation of IgG was detected in the present patient. Unfortunately, we could not measure his IgG4 level because of lack of serum sample. IgG4-related systemic disease is associated not only with thoracic aortic lesions (18) but also with lesions of the abdominal aorta (19, 20). Though IgG4-related disease is clinically quite different from Churg-Strauss syndrome, IgG4 elevation in Churg-Strauss syndrome has been reported recently (21). Furthermore allergic symptoms, such as bronchial asthma and allergic rhinitis, were detected in approximately half of IgG4-related disease, including Mikulicz’s disease (22) and autoimmune pancreatitis (23). Therefore it is suggested that the allergic reaction based on Th2 cytokines, such as interleukin (IL)-4, IL-5 and IL-10, may play an important role in both Churg-Strauss syndrome and IgG4-related disease.

In conclusion, chronic periaortitis might be involved in Churg-Strauss syndrome and the clinical manifestation of IgG4-related disease may occasionally be similar to it. Therefore we need to have a more specific approach for the differentiation of these diseases clinically and experimen-

	The authors state that they have no Conflict of Interest (COI).

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