Case report

Wegener’s granulomatosis: isolated involvement of the trachea and larynx

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SUMMARY A 26 year old man with subacute hoarseness and stridor was shown to have Wegener’s granulomatosis isolated to the trachea and larynx. Although isolated laryngeal Wegener’s is unusual, a review of the literature suggests that early treatment with cyclophosphamide is warranted.

Key words: vasculitis, cyclophosphamide, stridor.

Wegener’s granulomatosis typically presents as sinusitis, pulmonary infiltrates, and glomerulonephritis.1-3 Occasionally, Wegener’s granulomatosis appears, at least initially, to involve only one organ or one region, most commonly the lung.2-4 Wegener’s granulomatosis isolated to the tracheolarynx is rare and especially difficult to recognise since biopsies of this region rarely demonstrate vasculitis.2-11 In an effort to promote early diagnosis of this now treatable disorder we report a case of isolated tracheolaryngeal Wegener’s granulomatosis and review the pertinent literature.

Case report

A previously healthy 26 year old man was referred in July 1985 for evaluation of hoarseness, stridor, and dyspnoea. He had lost 3-5 kg but denied other symptoms. A detailed general physical examination was normal. Indirect laryngoscopy showed the subglottic larynx to be greatly narrowed circumferentially by boggy, erythematous folds of redundant tissue. The vocal cords were mobile.

The blood counts, chemistries, and urine analysis were normal. Skin tests for tuberculosis and coccidioidomycosis were negative. Chest and sinus x rays were normal. A computed axial tomographic

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Fig. 1 Laryngeal biopsy specimen showing small vessel vasculitis.
<table>
<thead>
<tr>
<th>Reference No</th>
<th>Sex</th>
<th>Age</th>
<th>Initial symptoms</th>
<th>Disease location</th>
<th>Laryngeal biopsy findings</th>
<th>Late manifestations</th>
<th>Therapy</th>
<th>Outcome (follow up)</th>
</tr>
</thead>
<tbody>
<tr>
<td>8</td>
<td>M</td>
<td>18</td>
<td>Hoarseness, dyspnoea</td>
<td>Upper trachea subglottis</td>
<td>Granuloma</td>
<td>Saddle nose, sinusitis, haematuria, proteinuria</td>
<td>Corticosteroids</td>
<td>Death</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>17</td>
<td>Hoarseness, sore throat, stridor</td>
<td>Subglottis, vocal cords</td>
<td>Not available</td>
<td>Skin vasculitis, glomerulonephritis</td>
<td>None stated</td>
<td>Tracheostomy, dialysis (14 mo)</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>55</td>
<td>Hoarseness</td>
<td>Subglottic mass</td>
<td>Granuloma</td>
<td>Glomerulonephritis</td>
<td>Cyclophosphamide, corticosteroids, azathioprine</td>
<td>Tracheostomy, transient renal failure, remission (9 mo)</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>39</td>
<td>Hoarseness, stridor</td>
<td>Subglottis</td>
<td>Granuloma inflammation</td>
<td>Glomerulonephritis, lung vasculitis</td>
<td>Radiation, corticosteroids</td>
<td>Tracheostomy, death from renal failure</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>14</td>
<td>Hoarseness</td>
<td>Subglottis</td>
<td>Granuloma inflammation</td>
<td>Haematuria, skin ulcers</td>
<td>Corticosteroids</td>
<td>Tracheostomy, skin ulcers, alive (10 mo)</td>
</tr>
<tr>
<td>11</td>
<td>M</td>
<td>51</td>
<td>Dyspnoea</td>
<td>Subglottis</td>
<td>Non-specific inflammation</td>
<td>Glomerulonephritis, skin purpura</td>
<td>Corticosteroids, cyclophosphamide</td>
<td>Tracheostomy, renal insufficiency, alive (1 mo)</td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>52</td>
<td>Hoarseness</td>
<td>Vocal cords, subglottis</td>
<td>Necrosis, granuloma</td>
<td>None</td>
<td>Corticosteroids</td>
<td>Death from tracheal obstruction (54 mo)</td>
</tr>
<tr>
<td>This report</td>
<td>M</td>
<td>26</td>
<td>Hoarseness, stridor</td>
<td>Subglottis to carina</td>
<td>Granuloma, necrosis, vasculitis</td>
<td>None</td>
<td>Corticosteroids, cyclophosphamide</td>
<td>Tracheostomy, improved (12 mo)</td>
</tr>
</tbody>
</table>
(CT) scan of the trachea showed concentric soft tissue, narrowing from the subglottic region to the left mainstem bronchus. A description of the CT findings has been reported previously. Indeed, three of the seven previously reported patients with laryngeal Wegener’s granulomatosis were adolescents. As with our patient, most other patients presented with subacute stridor or hoarseness, or both.

Additional similarities between our patient and those listed in Table 1 became evident when the larynx and upper airways were evaluated. In most patients, as in our case, direct laryngoscopy showed involvement of the subglottic region. Vocal cord involvement was less common. The disease can be localised, often to the posterior wall of the subglottic region, or diffuse, with circumferential narrowing. The mucosa may be erythematous, boggy, or ulcerated. Proliferation of granulation tissue produces mass lesions in some patients. Extension of the disease below the carina is quite rare. Tracheostomy was often required, as in our patient, to ensure adequate ventilation during the acute inflammatory phase of the disease.

The recognition of isolated laryngeal Wegener’s granulomatosis is made especially difficult by the fact that the histopathology is often not considered diagnostic. Typically, biopsy specimens from the upper respiratory tract of patients with Wegener’s granulomatosis show acute and chronic inflammation with necrosis. Granuloma are often present, but vasculitis is rarely found. Multiple laryngeal biopsies were performed in most of the seven previous cases, and vasculitis was found in none. In our patient, multiple biopsies at two different times were performed, one of which showed vasculitis, making this only the second time that vasculitis has been described on laryngeal tissue biopsy.

The tendency to dismiss the diagnosis of Wegener’s granulomatosis until and unless vasculitis is demonstrated is incorrect and results from a misconception of the clinicopathological features of Wegener’s granulomatosis. Although Wegener’s granulomatosis is usually classified as a form of vasculitis, Wegener himself emphasised the importance of the necrosis with granuloma formation. Of all the organs, the lung is the site where both the vasculitis and the granulomatosis necrosis are most frequently seen. Biopsy specimens from other sites, especially the upper respiratory tract, rarely show all the ‘classic’ pathological findings.

Thus the diagnosis of laryngeal Wegener’s granulomatosis depends on the presence of a consistent clinicopathological picture as well as the exclusion of the other causes of laryngeal obstruction, which have recently been reviewed in detail.

The outcome of primary laryngeal Wegener’s granulomatosis appears to depend on the timing and
the type of treatment.\textsuperscript{5–11} None of the seven previous cases received immunosuppressive therapy while the Wegener's granulomatosis was limited to the larynx. Subsequently, all seven developed either progressive laryngeal obstruction or disseminated disease (Table 1). Three patients, including our patient, were eventually treated with cyclophosphamide, and all improved. No patient remitted without cyclophosphamide therapy. This experience strongly suggests, but does not prove, the need for cyclophosphamide therapy in primary laryngeal Wegener’s granulomatosis. Given the very poor outcome experienced by most patients with isolated laryngeal Wegener’s granulomatosis who are untreated or treated after dissemination of their disease, early treatment with cyclophosphamide appears to be warranted.

References

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