Cricoarytenoid Arthritis in Ankylosing Spondylitis*

A Cause of Acute Respiratory Failure and Cor Pulmonale

Daniel M. Libby, M.D.;† W. Shain Schley, M.D.;‡ and James P. Smith, M.D.§

A man with ankylosing spondylitis developed cor pulmonale and acute respiratory failure due to cricoarytenoid arthritis. He was successfully treated by endoscopic arytenoidectomy, and the surgical specimen showed ossified cartilage. Flow-volume curves documented extrathoracic airway obstruction produced by ankylosis of the cricoarytenoid joints.

Ankylosing spondylitis frequently involves peripheral joints, but cricoarytenoid joint involvement has only rarely been noted.1–3 A man with ankylosing spondylitis developed cor pulmonale and acute respiratory failure due to cricoarytenoid arthritis. The literature on this unique cause of upper airway obstruction is reviewed.

Case Report

A 69-year-old man with a 20-year history of ankylosing spondylitis involving the entire spine and sacroiliac joints entered the hospital with increasing breathlessness for two months and noisy breathing and hoarseness for one month. On admission, he was in respiratory distress with inspiratory stridor and expiratory rhonchi. There was pedal edema, jugular venous distension, and accentuation of the pulmonary component of the second heart sound. Severe anterior neck flexion deformity and limitation of temporomandibular joint motion were present. The hemoglobin level was 17.0 g/100 ml and the hematocrit value was 51.3 percent. Arterial blood gas levels measured with the patient breathing 40 percent oxygen were as follows: PaO₂, 82 mm Hg; PaCO₂, 84 mm Hg; and pH, 7.40. He was treated with antibiotics, bronchodilators, corticosteroids, and chest physiotherapy, but respiratory insufficiency worsened over the next 24 hours. Indirect laryngoscopy revealed erythema and edema of the vocal cords with bilateral fixation in the paramedian position. Hypoxemia and respiratory acidosis necessitated endotracheal intubation, performed with the endotracheal tube “sleeved” over a bronchofiberscope. One week later, after an unsuccessful attempt at extubation, he was transferred to The New York Hospital Respiratory Intensive Care Unit. After severe hypoxemia improved and arterial oxygenation became adequate with the patient breathing 24 percent oxygen, surgical consultation reconfirmed that the extreme anterior neck flexion made a definitive means of establishing a secure airway by cervical tracheotomy impossible.

Another trial without endotracheal tube was initiated. His respiratory status remained tenuous with audible stridor and persisting moderate hypoxemia and hypercapnia. After two weeks, when the edema from intubation should have subsided, direct fiberoptic laryngoscopy showed paramedian fixation of the vocal cords reducing the airway diameter to 1.5 to 2.0 mm. The patient underwent endoscopic right arytenoidectomy under general anesthesia with increase of the airway diameter to about 6.0 mm. Microscopic examination of the resected specimen revealed partial replacement with bone tissue and chronic inflammation (Fig 1).

Because the patient was critically ill, preoperative pulmonary function testing was not possible. Spirometry (Table 1) performed two weeks postoperatively showed a mild mixed ventilatory impairment, but flow-volume curves suggested significant residual extrathoracic (laryngeal) obstruction with mildly reduced expiratory flow rates but severe reduction of inspiratory flow. The ratio of expiratory to inspiratory flow at midvital capacity (E_{50}/I_{50}) was 3.0:

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CRICOARYTENOID ARTHRITIS IN ANKYLOSING SPONDYLITIS 641
Cricoarytenoid arthritis is most commonly seen in rheumatoid arthritis, is recognized clinically in 25 to 35 percent of cases, and is present in the majority of cases at autopsy. involvement of the cricoarytenoid joints also occurs in Reiter’s syndrome, systemic lupus erythematosus, progressive systemic sclerosis, Tietze’s syndrome, and relapsing polychondritis. Four cases of cricoarytenoid arthritis complicating ankylosing spondylitis have previously been described (Table 2). Symptoms of laryngeal involvement were present for several years in three patients, but for just four weeks in the other. In all four cases, ankylosing spondylitis was far advanced before cricoarytenoid arthritis caused airway compromise, and in only one case did respiratory failure result. The development of acute respiratory failure with cor pulmonale in our patient was shown to be due to progressive large airway narrowing as evidenced by the history of hoarseness and noisy breathing and the paramedian vocal cord fixation seen at laryngoscopy. Confirmation of its physiologic importance even after the airway diameter had been increased threefold by arytenoidec- tomy was obtained with the flow-volume loop which showed a persistent severe reduction in inspiratory airflow despite relative preservation of expiratory flow. The histologic findings in the cricoarytenoid joint involved with ankylosing spondylitis have only once been previously reported and consisted of osseous metaplasia with an accompanying pannus. In the present case, there was also metaplastic bone tissue (Fig 1) and chronic inflammatory infiltrate.

Cor pulmonale caused by upper airway obstruction has been noted in children with enlarged tonsils and adenoids and with subglottic stenosis following endotracheal intubation. It has been seen in adults with or without obesity or chronic bronchitis and emphysema who experience central or peripheral obstructive apneas while sleeping. The hypventilation induced by the obstruction or the abnormal central respiratory control mechanism lead to hypoxemia, hypercapnia, and acidosis—potent stimuli of pulmonary vasoconstriction,

### Table 1—Pulmonary Function Tests*

<table>
<thead>
<tr>
<th></th>
<th>Observed</th>
<th>Percent Predicted</th>
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</thead>
<tbody>
<tr>
<td>VC (ml)</td>
<td>2210</td>
<td>65</td>
</tr>
<tr>
<td>FEV₁ (ml)</td>
<td>1630</td>
<td>65</td>
</tr>
<tr>
<td>MVV (L/min)</td>
<td>49</td>
<td>54</td>
</tr>
<tr>
<td>E₁₀ (L/sec)</td>
<td>3.9</td>
<td>71</td>
</tr>
<tr>
<td>I₁₀ (L/sec)</td>
<td>1.3</td>
<td>24</td>
</tr>
</tbody>
</table>

*In airway obstruction due to cricoarytenoid arthritis complicating ankylosing spondylitis, indicating severe extrathoracic variable airway obstruction. VC indicates vital capacity; FEV₁, forced expiratory volume in one second; MVV, maximum voluntary ventilation; E₁₀ and I₁₀, instantaneous maximum expiratory and inspiratory flow rates at mid-vital capacity, obtained from the flow-volume loop.

(normal 0.8 to 1.2). Arterial blood gas levels on air were as follows: pH, 7.41; PaCO₂, 42 mm Hg; and PaO₂, 70 mm Hg. The hematoctrit value was 38 percent. Pedal edema and jugular venous distension were no longer present.

The patient experienced no serious respiratory difficulty over the next 18 months.

### Table 2—Clinical Characteristics of CA Associated with AS*

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Age, Sex</th>
<th>Time from Dx of AS to CA, yrs</th>
<th>Peripheral Joints</th>
<th>Extraarticular Manifestations of CA</th>
<th>Symptoms</th>
<th>Acute Airway Compromise</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Berendes and Miehlke, 1973¹</td>
<td>62, not reported</td>
<td>35</td>
<td>Shoulder, toe, elbow, knees, sternoclavicular, temporomandibular</td>
<td>None</td>
<td>Dyspnea for two years, hoarseness, aspiration</td>
<td>None</td>
<td>Intraarticular cortisone, tracheostomy</td>
</tr>
<tr>
<td>Wojtulweski et al, 1973²</td>
<td>64, M</td>
<td>12</td>
<td>Hips, knee</td>
<td>Iritis</td>
<td>Dyspnea, hoarseness, stridor for nine years</td>
<td>Acute respiratory failure</td>
<td>Tracheostomy, arytenoidec- tomy</td>
</tr>
<tr>
<td></td>
<td>51, M</td>
<td>11</td>
<td>Temporomandibular and “others”</td>
<td>Iritis</td>
<td>Hoarseness for ten years</td>
<td>Difficult endotracheal intubation during general anesthesia</td>
<td>Emergency crico-thyroid puncture, endotracheal intubation</td>
</tr>
<tr>
<td>Bienenstock and Lanyi, 1977¹</td>
<td>50, F</td>
<td>27</td>
<td>Hips</td>
<td>None</td>
<td>Dyspnea for four weeks, pain in throat radiating to the ear</td>
<td>None</td>
<td>Prednisone</td>
</tr>
</tbody>
</table>

*AS indicates ankylosing spondylitis; CA, cricoarytenoid arthritis.

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right ventricular failure, and cor pulmonale. It has also been noted that physiologic dead space increases with exercise in subjects with upper airway obstruction, adding to the hypoxemia. An additional factor contributing to hypoxemia and hypercapnia is the inability to increase alveolar ventilation in response to the higher requirements for these substrates during infection or exercise.

The management of upper airway obstruction in ankylosing spondylitis may present a difficult problem. Temporomandibular and cervical ankylosis may prevent a direct view of the laryngeal structures, and attempts to extend the neck may lead to spinal fractures and neurologic compromise. Flexible fiberoptic (direct) laryngoscopy, as performed here, is the preferred method of visualizing the laryngeal structures when the rigid laryngoscope cannot be used because of cervical and temporomandibular ankylosis. Endotracheal intubation may have to be performed with the aid of the flexible bronchofiberscope. Cervical tracheotomy may be impossible, as it was in this case, when extreme anterior neck flexion prevents adequate exposure. Unilateral arytenoidectomy has been successful with insignificant morbidity in rheumatoid arthritis, and based on the experience here and in other cases, is recommended in ankylosing spondylitis. Arytenoidectomy, whether performed endoscopically or through a cervical incision, should always be preceded by tracheotomy to assure an adequate airway. Arytenoidectomy without prior tracheotomy is not recommended and was performed alone here only as a lifesaving measure because cervical tracheotomy was technically impossible.

As in rheumatoid arthritis, cricoarytenoid joint involvement in ankylosing spondylitis may be quite common pathologically and yet its clinical and physiologic importance underestimated due to the sedentary lifestyle of patients with disabling arthritis. Acute respiratory failure or cor pulmonale due to laryngeal obstruction may only represent a terminal phase in a gradually evolving process. Detection of physiologically-important cricoarytenoid arthritis is aided by the maximum expiratory and inspiratory flow-volume curves. When unexplained respiratory insufficiency or symptoms of laryngeal disease are present in a patient with ankylosing spondylitis, laryngoscopy and flow-volume curves should be performed.

REFERENCES


Reciprocating Tachycardia with Only Odd-numbered Beats in the Wolff-Parkinson-White Syndrome*

Julian M. Schamroth, M.B.B.Ch.; Dirk P. Myburgh, S.M., M.B.Ch.B.; and Leo Schamroth, M.D.

A case of Wolff-Parkinson-White syndrome with reciprocating tachycardia from retrograde Kent's bundle conduction is described. The paroxysms of reciprocating tachycardia manifested with the unusual, and hitherto unreported, feature of only odd-numbered beats. The phenomenon is explained on the basis of alternate anterograde conduction through two A-V nodal pathways.

The labyrinthine structure of the A-V node forms an ideal anatomic substrate for multiple intranodal conduction pathways. Furthermore, the elegant electrophysiologic studies of Moe and Mendez and Moe et al have shown that the A-V node contains at least two functionally separate A-V pathways with different refractory periods and conduction times. This was demonstrated in man by Denes and associates in 1973. It is possible for a critically timed impulse to find one pathway refractory and the other responsive. An impulse that is very premature will find no responsive tissue and will be blocked. One that is relatively late will find both A-V nodal pathways responsive and will be conducted through all fibers. Moe and associates showed, for example, that this principle could be used to explain the so-called supernormal phase of A-V conduction manifesting as alternation of A-V conduction times. The case presented here reflects a hitherto unreported phenomenon.

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