Case Report

Cricoarytenoid Arthritis with Rheumatoid Arthritis and Systemic Lupus Erythematosus

YUKI NANKE, SHIGERU KOTAKE, KOUICHI YONEMOTO, MASAKO HARA, MAKOTO HASEGAWA, and NAOYUKI KAMATANI

ABSTRACT. A 56-year-old woman with rheumatoid arthritis (RA) suddenly developed severe respiratory distress and laryngeal stridor, which required endotracheal intubation. She had had RA for 12 years, which had been controlled well with prednisolone (3 mg/day) at the orthopedic clinic. Laryngoscopy revealed cricoarytenoid arthritis. She was finally diagnosed as having overlap syndrome with RA and systemic lupus erythematosus. She was given high dose corticosteroids that improved her clinical symptoms and laryngoscopic findings. She represents the first patient with overlap syndrome who developed an acute airway obstruction due to cricoarytenoid arthritis. (J Rheumatol 2001;28:624–6)

Key Indexing Terms: CRICOARYTENOID ARTHRITIS SYSTEMIC LUPUS ERYTHEMATOSUS OVERLAP SYNDROME

Cricoarytenoid arthritis occurs in 25–35% of patients with rheumatoid arthritis (RA)1-3. Hoarseness, dyspnea, and fullness in the throat when swallowing or speaking are the predominant symptoms1,2,4,5. However, arthritis of cricoarytenoid joints with acute airway obstruction is uncommon and is rarely reported6-8. In addition, laryngeal involvement caused by systemic lupus erythematosus (SLE) is also seldom reported9,10. Teitel, et al reviewed the literature and found laryngeal edemas in 28% and vocal cord paralysis in 11% of patients with SLE11. This potentially life-threatening disease is not fully appreciated by many physicians caring for patients with collagen diseases. We describe a patient with RA who presented with cricoarytenoid arthritis causing acute upper airway obstruction, which required endotracheal intubation. She was later diagnosed as having overlap syndrome with RA and SLE. This case may give us insight into the clinical symptoms and medical procedures of cricoarytenoid arthritis in patients with collagen diseases.

CASE REPORT
A 56-year-old Japanese woman was diagnosed as having RA due to swelling of the small joints of the hands and morning stiffness in 1986. Gold sodium injections were initiated. In 1989, she came to the orthopedic clinic of our hospital, and D-penicillamine was administered instead of gold sodium injections. In 1990, her disease flared. D-penicillamine was stopped, and methotrexate and prednisolone (PSL) (10 mg/day) were initiated. Her joint symptoms had been controlled well by PSL (3 mg/day) since 1995. In late January 1998, she noted dyspnea and bloody sputum, hoarseness, and fullness of the throat when swallowing and speaking. On February 5, she was admitted to another facility for severe respiratory distress and laryngeal stridor. The laryngoscopic finding confirmed laryngeal edema and poor mobility of vocal cords (Figure 1), which required endotracheal intubation (Figure 2). She was diagnosed as having cricoarytenoid arthritis. High dose intravenous hydrocortisone was started and tapered to 40 mg over 5 days. She was transferred to our hospital February 10 for further investigation. Upon admission, she had slight inspiratory stridor. Her temperature was 36.2°C, heart rate 96/min, and blood pressure 130/76 mm Hg. There were slight basilar rales in both lungs. There was neither eruption nor infectious signs. Swan neck deformities in her hands and hammer toes were noted. There were rheumatoid nodules (1.5 cm in diameter) in both elbows, but no active synovitis was noted. Chest radiograph showed no abnormalities. Blood gases revealed a PO2 of 65.1 torr, and a PCO2 of 46.6 torr. The erythrocyte sedimentation rate was 76.4 mm/h. Dipstick test showed (1+) for protein and (1+) for occult blood, and an examination of urine sediment showed cellular casts of granular type (5/10). The white blood cell count was 9500/mg (normal 3300–9000) with 1.5% lymphocytes, red blood cell count 408 × 10^9/mg (308–500), hemoglobin 12.6 g/dl (11.5–15.0), and hematocrit 37.5% (34.8–45%). Total protein was 8.0 g/dl (6.7–8.3), albumin 3.0 g/dl (3.8–5.3). Serum creatinine level was 0.5 mg/dl (0.6–1.1) and C-reactive protein (CRP) 2.6 mg/dl (≤ 0.5). Immunological tests revealed a particle agglutination titer of 1:2560, antinuclear antibody (ANA) titer 1:5120 (homogeneous and speckled pattern), anti-SSA 16, anti-SSB 8, anti-Sm 8, and anti-RNP 16. Anti-topo I, anti-Jo-1, and anti-DNA antibodies were negative. Lupus anticoagulant, LE cells, and a direct Coombs test were all positive. Other serum data were C3 of 92 mg/dl (50–110), C4 of 25 mg/dl (13–45), CH50 of 35.7 U/dl (29–48), thrombin-antithrombin III complex (TAT) 25.5 ng/ml (< 3.2), plasmin α2 plasmin inhibitor complex (PIC) 5.6 vng/ml (< 1.0), and circulating immune complexes (C1q method) 9.9 μg/ml (normal < 2.9).

Hand radiographs revealed stage IV RA. Magnetic resonance imaging (MRI) of the neck showed soft tissue swelling with a deviation to the left (Figure 3A). She was finally diagnosed as having cricoarytenoid arthritis...
with RA (stage IV, class 3). She also satisfied the American College of Rheumatology criteria for SLE12. Corticosteroid was tapered to 10 mg/day over 7 weeks. On the 20th day of admission, followup laryngoscopy and MRI were performed (Figure 3B). Movement of the right vocal cord was diminished, although the left vocal cord had full range of movement, and laryngeal edema was resolved. She was discharged in good condition on June 13. In September 2000, we saw her at our outpatient clinic taking prednisolone 9 mg/day in good condition.

**DISCUSSION**

This patient had RA for more than 10 years and suddenly developed airway obstruction due to cricoarytenoid arthritis. After treatment with high dose intravenous hydrocortisone, it was found that she met the criteria for the classification of SLE12 (protein urea, lymphopenia, positive for ANA and lupus anticoagulant). The most common etiology of cricoarytenoid arthritis is generalized arthritis. Cricoarytenoid arthritis can be induced by penetrating trauma with secondary infection, or from internal trauma to the larynx from rough endoscopy or intubation3. Cricoarytenoid arthritis occurs in up to 35% of patients with RA1,3, and is
present in the majority of cases at autopsy. On the other hand, it is possible that cricoarytenoid arthritis develops in SLE, although laryngeal involvement caused by SLE is rare. It can range from mild ulceration, vocal cord paralysis, and edema to necrotizing vasculitis with airway obstruction. In most cases, symptoms such as hoarseness, dyspnea, and vocal cord paralysis related to SLE are resolved with PSL. Thus it is difficult to determine whether cricoarytenoid arthritis of this patient was due to RA or to SLE.

The pathophysiology of laryngeal inflammation in SLE is not well understood. This complication, which occurs mainly in patients with active disease, would suggest that an immune or vasculitis process is responsible. Indeed, oral mucosal immunoglobulin and complement deposition have been described. Tissue deposition of immune complexes leads to complement activation and eventually to neutrophil and mononuclear cell infiltration, resulting in localized inflammation. The laryngeal basement membranes may be better suited to binding immune complexes of a particular size or charge. Alternatively, they may have particular laryngeal antigens that lead to localized formation of immune complexes. We speculate that an immune or vasculitis process may have been responsible in this patient.

Cricoarytenoid arthritis sometimes leads to death, although life-threatening arytenoid fixation is rare. However, the cricoarytenoid joint is not readily accessible for inspection or palpation during routine examination, and symptoms of laryngeal inflammation are often unnoticed by the patient. Thus, not only otolaryngologists but also rheumatologists should be aware of the early symptoms of cricoarytenoid arthritis. To our knowledge, this is the first case with overlap syndrome with airway obstruction due to cricoarytenoid arthritis.

In summary, involvement of the larynx in overlap syndrome has not been reported. This is the first report of overlap syndrome that caused airway obstruction due to cricoarytenoid arthritis, for which corticosteroid was effective. Our case may provide insight into the clinical features and medical procedures for cricoarytenoid arthritis in patients with collagen disease.

REFERENCES