Sudden Glottic Stenosis Caused by Cricoarytenoid Joint Involvement due to Rheumatoid Arthritis

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Abstract

A woman with rheumatoid arthritis (RA) experienced glottic stenosis approximately two months after switching from etanercept to tocilizumab. Cricoarytenoid joint (CAJ) arthritis due to RA was diagnosed. An awake tracheostomy saved the relievable airway, and the administration of methylprednisolone and infliximab ameliorated the flare-up and glottic stenosis. A follow-up examination revealed the recovery of the patient’s normal voice and good control of RA with infliximab and methotrexate. Although general physicians do not frequently encounter patients with symptomatic CAJ arthritis, this condition should be considered as it can be life-threatening. Therefore, when detected, it should be diagnosed and treated immediately.

Key words: cricoarytenoid joint arthritis, rheumatoid arthritis, airway obstruction, steeple sign


Introduction

Laryngeal involvement in rheumatoid arthritis (RA) patients, especially airway obstruction due to cricoarytenoid joint (CAJ) arthritis, is potentially life-threatening (1). While this is well known to otolaryngologists, the condition is not commonly encountered by general physicians (2). Promptly recognizing airway obstruction due to CAJ arthritis is essential for appropriate management. We experienced a thought-provoking case of CAJ arthritis that we describe in this report.

Case Report

A 54-year-old woman with polyarthralgia consulted her family physician in 2006. Although she had been treated with nonsteroidal anti-inflammatory drugs (NSAIDs) for eight months, her symptoms did not improve. She was then referred to our hospital in 2007. She presented with morning stiffness and polyarthritis involving both wrists and ankles, the right shoulder and both knee joints. The patient’s serum levels were as follows: rheumatoid factor (RF), 0 IU/mL; matrix metalloproteinase-3 (MMP-3), 298 ng/mg; and C-reactive protein (CRP), 12.7 mg/dL. Based on these findings, she was diagnosed with RA. Initially, she was treated with methotrexate (8 mg/week; the maximum dose at that time in Japan) and prednisolone (5 mg/day) for 14 months; however, these drugs did not fully control the RA disease activity. This led us to add etanercept (25 mg twice weekly) to the treatment regimen. Etanercept initially produced such a good response for one year that we began tapering the dose of prednisolone until the agent was completely discontinued. At that point, the swelling and joint tenderness returned, and the RA appeared to be reactivated. The roentgenographic changes were consistent with erosive changes in the proximal interphalangeal joints and carpal bones. The RF level was 3 IU/mL, and the MMP-3 value was 145 ng/mL. The CRP level had increased to 7.80 mg/dL, and the disease activity score-28 with C-reactive protein (DAS28-CRP) was 5.43. Therefore, tocilizumab (TCZ) (8 mg/kg for four weeks) was substituted in place of etanercept. Prednisolone (5 mg/day) was prescribed at the same time to provide bridging therapy. Although TCZ is generally infused every four weeks, the second infusion was administered after only three weeks. At that point, the pain and

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The patient’s percutaneous oxygen saturation was 95% on room air, and her body temperature was 37.8°C. A physical examination revealed swelling of the right wrist and both knee joints. No redness or swelling was evident in the laboratory data. This episode appeared to be associated with swelling of some joints with no obvious signs of infection in the arytenoids. Therefore, we surmised that the immobilization of the vocal cords was primarily caused by arthritis of the CAJ due to RA. Emergency awake tracheostomy was performed with an anesthetist at hand owing to serious concerns of deteriorating airway dysfunction. Methylprednisolone (500 mg) was administered for three consecutive days, and ceftriaxone sodium (1 g/day) was administered for one week because coexisting infection could not be ruled out.

On hospital day 7, the WBC count had recovered to 6,200/μL; however, fiberscopy showed no improvement in the CAJ motion, and the CRP level remained high at 8.63 mg/dL. Therefore, infliximab administration was initiated. At that point, the inflammation decreased and the articular symptoms and glottic stenosis gradually improved. On hospital day 14, the CRP level improved to 2.91 mg/dL and the DAS28-CRP level improved to 3.45. Laryngoscopy showed recovery of the glottis function, and the patient was able to speak with a speech bulb prosthesis. Three weeks later, the device was removed, and the tracheostomy orifice spontaneously closed. Follow-up performed for a further five weeks revealed DAS28-CRP remission and the recovery of the patient’s normal voice. The RF and MMP-3 values improved to 6 IU/mL and 82.4 ng/mL, respectively.

swelling of the joints remained unchanged; however, the CRP level had dropped to 0.19 mg/dL, and the DAS28-CRP level had reduced to 3.95. A third administration was scheduled after a four-week interval (Fig. 1).

As stenosis in the upper airway was suspected, inhalation administration of epinephrine and betamethasone was performed. However, the patient’s respiratory status did not improve. Therefore, we immediately consulted an otolaryngologist; fiberscopy revealed immobilized vocal cords in the adducted position with a narrow glottis opening. Furthermore, only slight edema, without bright redness, was evident on the larynx mucous membrane, and no evidence of trauma to the CAJ was apparent. We did not detect any other neurological deficits. The CRP level was 26.1 mg/dL, the RF level was 18 IU/mL, the white blood cell (WBC) count was 9,500/μL and the DAS28-CRP level was 6.06. The red blood cell and platelet counts were normal (407×10^4/μL and 32.6×10^4/μL, respectively). No other abnormal findings were evident in the laboratory data.

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Discussion

Although the symptoms may be vague or absent, the prevalence of laryngeal involvement in patients with RA ranges from 13% to 75% in various clinical studies and from 45% to 88% in postmortem reports (3). CAJ arthritis is a typical manifestation of laryngeal involvement in RA patients. Laryngoscopy reveals red swelling of the arytenoids in the acute phase, edema of the vocal folds in the chronic phase and thickened mucosa over the arytenoids in both phases (4). James et al. reported that other causes of stridor in RA patients include ischemic neuropathy caused by vasculitis, cervicomedullary compression due to rheumatoid involvement of the cervical spine, confluent rheumatoid nodules and laryngeal amyloidosis (1). In addition to RA, the differential diagnosis of glottal dysfunction includes infection, recurrent laryngeal nerve palsy and trauma. In our case, the glottal symptoms occurred in connection with inflammation of the right wrist and both knee joints. The patient had a history of RA but no history of trauma. Laryngoscopy showed only slight edema, without bright redness around the immobilized vocal cords. Furthermore, there were no neurological deficits, and treatment for the inflammation of RA improved the movability of the vocal cords in the absence of nervous system therapy. For these reasons, we considered that the glottal dysfunction was primarily caused by complications of RA due to CAJ arthritis, which can flare up in the chronic state. Coexisting infection could not be excluded outright due to the patient’s low-grade fever and slight elevation in the WBC count. Regrettably, we were not able to obtain photographs of the patient’s vocal cords on admission, as the portable laryngoscope was not fitted with a camera.

With regard to diagnosis, low-voltage radiographs, computed tomography (CT) and magnetic resonance imaging (MRI) are useful for detecting upper airway obstruction, and the diagnosis of CAJ arthritis can be confirmed by fiberoptic laryngoscopic examinations (3, 5). However, laryngoscopic specialists are not always available in emergencies. CT and MRI examinations require time, and transferring an unstable patient to the appropriate scanning room entails risk. Therefore, the “steeple sign” is helpful for detecting CAJ arthritis. This sign is observed on anteroposterior radiographs of the soft tissue of the neck. The normal lateral convexity of the subglottic trachea is lost, and narrowing of the subglottic lumen produces an inverted V configuration (Fig. 2). This sign indicates the presence of edema in the trachea (6). Our patient exhibited a visible steeple sign on the first chest radiograph, which improved a few weeks later. The existence of this sign on radiographs in RA patients implies the presence of CAJ arthritis. Computed radiography systems are now common and facilitate obtaining enlarged images of the upper airway using chest X-rays as well as adjusting the density of the photographic images on the monitor of the associated computer terminal. This enables clinicians to observe the soft tissue of the neck more clearly. Therefore, chest X-rays including the cervical region should be ordered in the first instance.

Concerning treatment in the acute phase, airway management is the most important consideration because hypoxia can cause irreversible brain damage and be life-threatening. Although it has been reported that the administration of steroids and epinephrine inhalation is effective for treating CAJ arthritis (2), tracheostomy is the preferred treatment in patients with stridor and bilateral midline arytenoid fixation (5). Furthermore, intubation is a difficult and rough maneuver that can damage the arytenoids in patients with bilateral CAJ arthritis. It is essential to establish airway patency before the onset of severe dyspnea. Of course, fundamental and definitive therapy for the flare-up of RA should be successively provided.

We herein reported a case of CAJ involvement due to RA. Although general physicians do not frequently encoun-
ter symptomatic CAJ arthritis patients, the condition should be kept in mind, as it can be life-threatening. Therefore, if detected, it should be diagnosed and treated immediately. We believe that obtaining a chest radiograph including the cervical region is helpful for detecting CAJ arthritis, and tracheostomy remains the definitive treatment for this condition in the acute phase.

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

References