# Acromegaly Presenting With Bilateral Vocal Cord Immobility: Case Report and Review of the Literature

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**Summary: Objective.** To present a case of bilateral vocal cord immobility (BVCI) in a patient with acromegaly and review the current literature describing this presentation.
**Design.** Case report and literature review.
**Setting.** Academic tertiary care center.
**Methods.** English language literature search of online journal databases.
**Results.** A 56-year-old man presented with 3 months of progressive stridor and shortness of breath. Transnasal flexible endoscopy revealed BVCI. A tracheostomy was performed to secure his airway. Further history was suggestive of acromegaly and imaging demonstrated a pituitary macroadenoma. The diagnosis of acromegaly was made. The patient was treated with octreotide followed by an endoscopic trans sphenoidal resection of the pituitary adenoma. Sixteen months after his initial presentation, a right laser arytenoidectomy was performed and the patient was subsequently decannulated. In the literature to date, 11 cases of BVCI in acromegaly have been reported. These patients often present with stridor and require a tracheostomy. With treatment of their acromegaly, these patients may regain vocal cord mobility and may be decannulated.
**Conclusion.** Acromegaly with BVCI is a rare presentation. Acute management of the airway of patients with acromegaly presenting with BVCI typically requires a tracheostomy. A period of 15 months should be allowed for restoration of vocal cord mobility before airway opening procedures are considered.
**Key Words:** Acromegaly–Vocal cord fixation–Vocal cord immobility–Airway–Stridor.

## INTRODUCTION

Acromegaly is a rare endocrine disorder caused by excessive growth hormone secretion, which is often associated with a pituitary adenoma.[1,2] Manifestations include insidious, characteristic facial and extremity changes, including enlarged supraorbital ridges, prognathism, macroglossia, and enlarged hands and feet. Moreover, respiratory manifestations are also well documented in the literature. Death caused by respiratory disease is three times more likely in patients with acromegaly than in the general population.[3] Pathologic changes in the craniofacial bones, rib cage, soft tissues of the head and neck, and lungs are thought to contribute to respiratory disease, including obstructive sleep apnea.[4] However, acute upper airway obstruction is uncommon and bilateral vocal cord immobility (BVCI) is extremely rare. This report describes a patient with an initial presentation of acromegaly consisting of stridor secondary to BVCI. A systematic literature review of similar patients was also performed to summarize available diagnostic and treatment data.

## CASE REPORT

A 56-year-old man presented to the emergency department with a 3-month history of progressive stridor and shortness of breath, which had acutely worsened. He was unable to lie flat without becoming dyspneic and had been frequently waking at night. Of note, he had been intubated for procedures for nephrolithiasis three times in the last year. Beyond this, his medical history was unremarkable other than diet-controlled suspected type 2 diabetes mellitus. He was a lifetime nonsmoker.

On presentation to the emergency department, he was assessed by the otolaryngology—head and neck surgery resident on call who performed flexible transnasal endoscopy on the patient revealing BVCI (Figure 1). The patient was admitted to hospital for observation and a trial of intravenous steroids. His stridor continued to progress overnight and it was decided that a tracheostomy would be necessary to secure his airway. The patient underwent urgent awake intubation, tracheostomy, suspension laryngoscopy, and bronchoscopy. Bilateral arytenoid fixation and minimally mobile vocal cords were observed. The vocal cords, subglottis, and trachea were unremarkable. However, the supraglottic mucosa was edematous and was biopsied.

A broad differential diagnosis for the BVCI was considered, including malignancy, intubation trauma, central nervous system lesion, diabetic neuropathy, recurrent laryngeal nerve injury, myasthenia gravis, sarcoidosis, amyloidosis, Wegener's granulomatosis, rheumatoid arthritis, and systemic lupus erythematosus. Iatrogenic trauma to the recurrent laryngeal nerve was ruled out from the patient's history. The biopsied tissue was negative for granulomatous processes. Serum antinuclear antibodies, antineutrophil cytoplasmic antibodies, and rheumatoid factor were not detected. The patient's erythrocyte sedimentation rate and C-reactive protein measurements were within normal limits. His diabetes was adequately controlled with a hemoglobin A1c of 6.9%. A computed tomography scan of the head, neck, and chest was done to rule out a possible vagal lesion or other structural cause for the vocal cord paralysis. A probable $1.1 \times 1.8 \times 1.2$ cm pituitary macroadenoma was identified on the study. The pituitary adenoma was later confirmed with a magnetic resonance imaging.

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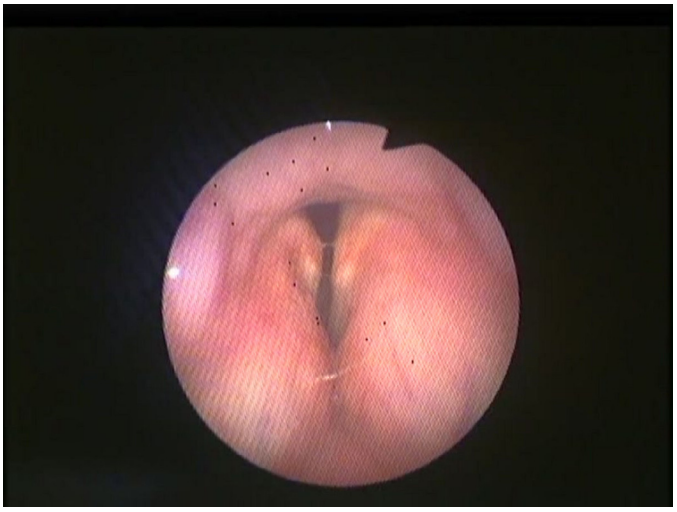

**FIGURE 1.** Flexible transnasal endoscopy demonstrating BVCI.

On closer observation, it was noted that the patient had very large hands. Upon further questioning, the patient reported that his wedding ring no longer fit, his shoes were tighter, and he felt that his tongue was enlarging. As such, acromegaly was suspected. An insulin-like growth factor-1 level of 1201 μg/L (reference range 80–270 μg/L) and human growth hormone of 70 μg/L (reference range <5.1 μg/L) confirmed the diagnosis. He had normal levels of thyroid-stimulating hormone, adrenocorticotropic hormone, prolactin, luteinizing hormone, and follicle-stimulating hormone. Both endocrinology and neurosurgery departments were consulted. The multidisciplinary consensus was to treat the patient with octreotide and reserve surgery for a salvage situation. Follow-up computed tomography scan and magnetic resonance imaging 4 months later showed minimal change in the adenoma, and thus, surgical management was pursued.

Nine months after the pituitary adenoma was discovered, it was resected using an image-guided endoscopic transsphenoidal approach, with no complications, in a combined fashion with an otolaryngologist and a neurosurgeon. Repeated insulin-like growth factor-1 and human growth hormone values were found to have declined to normal limits. Unfortunately, repeat flexible nasopharyngoscopy over the first 16 postoperative months failed to demonstrate any change in the patient's vocal cord status. The patient then underwent a unilateral endoscopic laser arytenoidectomy. One month later, he was successfully decannulated. At 6 months post decannulation, he remains tracheostomy-free.

## METHODS

An English language literature search was performed on PubMed, MEDLINE(R) In-process & Other Non-indexed Citations, MEDLINE (1950 to present), Embase, and Scopus for cases of BVCI and acromegaly. The search terms used included *acromegaly* AND *vocal cord*, *acromegaly* AND *paralysis*, *acromegaly* AND *fixation*, *acromegaly* AND *airway*, *acromegaly* AND *larynx*, and *acromegaly* AND *laryngeal*. Bibliographies of pertinent studies were also reviewed for relevant articles. Articles were further reviewed if they described both a case of acromegaly and a case of abnormal vocal cord mobility on airway endoscopy. To be included as a case, complete BVCI, fixation, or paralysis had to be described. All cases where partial mobility was present or where vocal cord immobility was directly preceded by neck surgery were excluded.

Prior to commencement, health research ethics board approval was obtained.

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

## RESULTS

Twelve articles describing complete BVCI in acromegaly were identified in the literature search. Ten of these articles met our inclusion criteria. A summary of these cases is presented in Table 1.[5–12] Two additional cases of acromegaly and BVCI are reported.[6,10] However, both of these cases presented immediately following thyroidectomy. Therefore, injury to the recurrent laryngeal nerve intraoperatively cannot be definitively excluded as the cause of vocal cord immobility. Additional cases were excluded as they did not describe complete BVCI.

Including the presented case, a total of 11 cases have been reported in the literature to date.[5–12] The mean age at presentation was 50.3 ± 13.2 years. Ten patients were male and one was female. Of these 11 cases, nine required tracheostomy. With treatment of acromegaly, most of the patients had improvement of their respiratory symptoms and some regained bilateral vocal cord mobility with the average documented time to restored vocal cord mobility being 6.7 ± 5.9 months (range 2–15 months).[5,9,11] However, at least three patients had persistent partial vocal cord immobility.[6,8] There are four cases in which successful decannulations were documented, with the mean time to decannulation being 9.8 ± 7.8 months (range 1–18 months).[5,8,9] Four patients required adjunctive airway procedures, including arytenoidectomy, in order to achieve decannulation.[8–10] Unfortunately, there is limited follow-up information provided in many of the cases, with no documentation of vocal cord mobility or decannulation following treatment.

## DISCUSSION

BVCI is rarely found in acromegaly. Although there are several proposed mechanisms, the cause of vocal cord immobility in acromegaly is unknown. Grotting and Pemberton proposed four potential mechanisms.[10] These include arthritic changes of the cricoarytenoid joint, cartilaginous enlargement impairing the mobility of the cricoarytenoid joint, stretching of the recurrent laryngeal nerves secondary to laryngeal enlargement, and recurrent laryngeal nerve injury caused by thyroid enlargement. Others have attributed the vocal cord fixation to a mass effect caused by thickened laryngeal structures.[5,7,13] Axonal demyelination and myopathies have also been reported in acromegaly and may contribute to vocal cord immobility.[6]

As the laryngeal enlargement subsides, vocal cord mobility may be regained. This reversibility of vocal cord fixation with treatment of acromegaly is suggestive of a reversible etiology. In addition, there are case reports of acromegalic patients presenting with stridor and dyspnea caused by laryngeal mucosal hypertrophy with or without partial vocal cord immobility