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Paradoxical Movement of Vocal Cords in Guillain - Barré Syndrome: Report a Case and Review of the Literature

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ABSTRACT

This is a report of a 50 year-old Thai male who was admitted to Siriraj Hospital because of progressive weakness of all extremities and increase shortness of breath. Endotracheal tube intubation was introduced for ventilatory support and secretion clearance. The diagnosis of Guillain-Barré syndrome was made based on typical presentation and CSF finding. The patient responsed well to intravenous immunoglobulin with successful extubation but developed upper airway obstruction from paradoxical movement of vocal cords which resolved in 5 days later. The review of the literature was done.

Keywords: Paradoxical movement; vocal cords; Guillain - Barré Syndrome; respiratory failure

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Guillain-Barré syndrome (GBS), also known as acute inflammatory demyelinating polyneuropa thy (AIDP), is the most common cause of acute progressive generalized paralysis and frequently severe, characterized by rapidly evolving symmetrical areflexic motor paralysis with or without sensory impairment. Although the cranial nerves are also frequently involved causing bulbar weakness, associated vocal cord paralysis has infrequently been reported. So far, from a review of previous literatures, only 5 cases were reported. Among them, 2 reported cases were bilateral vocal cord paralysis causing upper airway obstruction as a presentation of the disease. We hereby report a case of Guillain-Barré syndrome in which the patient presented with typical clinical feature of disease and vocal cord paralysis was diagnosed after the patient was hospitalized.

A 50-year-old Thai man was brought to the Emergency Room of Siriraj Hospital due to progressive difficulty in breathing for 1 day. He never had any history of medical problem. One week before admission, he felt tingling sensation and thickness of the skin on his feet, followed by on both hands. Soon he complained about leg weakness which rapidly progressed until he was unable to walk. In following morning he also developed weakness of his upper limbs, with increasing dysphagia, dysarthria, constipation and difficulty to clear up his bronchial secretion. A few days later he suffered from dyspnea and it progressed until he was brought to the hospital. He denied any fever or chill, muscle pain, urinary incontinence, recent upper respiratory tract infection, gastroenteritis, history of respiratory problem or allergy, neck trauma or any similar episode of weakness in the past. No remarkable food or environment exposures could be identified. He had been smoking about 2 packs per day for over 30 years but had not history of alcohol consumption. He denied any family history of weakness or any similar symptom.

Physical examination at the emergency room revealed a dyspneic man with symmetrically weak both upper and lower limbs; areflexia and bilateral facial muscle weakness were also detected. He reported numbness on both hands, forearms, legs and feet. His cardiovascular system examination showed heart rate of 90 beats per minute with regularity and his blood pressure was 160/90 mmHg. Chest auscultation revealed secretion sound but no other adventitious sound was found. First neurological evaluation showed grade 2 of peripheral muscle power without predominant proximal or distal pattern; and, facial diplegia was detected.

The patient was admitted to the medicine ward where he was intubated because he had unable to cough up the secretion and had impending respiratory failure. The physician who performed intubation noticed a decreased bilateral movement of the vocal cords. His lumbar puncture showed an elevated CSF protein of 265 mg/dl with only 2 WBC per ml of CSF. The serum creatine phosphate kinase (CPK) level was 202 U/L,(20-195 U/L).

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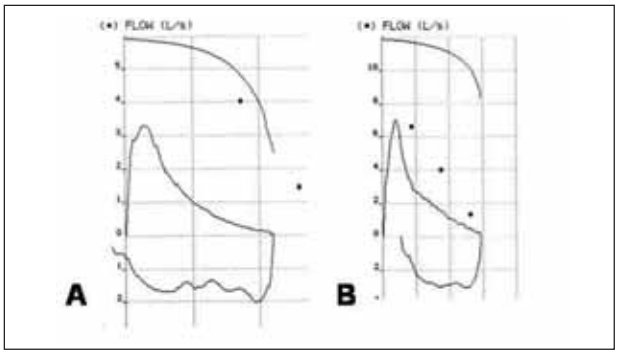


Fig 1. Flow volume loop (FV loop) of the patient A. On the day with sign of upper airway obstruction. The flow volume loop showed flattening of inspire limb indicated variable extrathoracic upper airway obstruction. ln B showed normalized FV loop when the patient was recover.

The serum potassium was 3.4 mmol/L and thyroid function test showed normal range. A clinical diagnosis of Guillain-Barré syndrome was assigned and intravenous immunoglobulin (IVIg) 20 grams/day for 5 days was selected for treatment.

On the last day of IVIg treatment, his neurological examination was improved to motor power grade[4]. In bed, he could move all his limbs, but still suffered from dysphagia and dysarthria. He was successfully weaned off the ventilator and extubation followed. After a moment of extubation he showed dyspneic breathing with inspiratory stridor. The spirometry showed flattening of inspiratory limbs compatible with variable extrathoracic airway obstruction (Fig 1A). The stridor responded to noninvasive ventilator (BiPAP ventilator with the setting of IPAP 10 cmH_2O and EPAP 5 cmH_2O). A bronchoscopy was performed for diagnosed the cause of the stridor. It showed bilateral vocal cord paralysis with paradoxical movement with the phase of respiration (Fig 2). The vocal cords showed no swelling and the rest of the airway was clear. The patient remained in the Respiratory Care Unit for BiPAP ventilator until 2 days later when he had no stridor and was comfortable with only canula oxygen 3 liters per minute was administrated.

At this time the patient gained his muscle strength to the point that he could moved to sit on his bed by himself; the facial diplegia was resolved; he could breathe without any assisting machine; the bowel and urinary bladder functions also improved gradually. Five days later a repeated spirometry showed normal flow volume loop (Fig 1B) which was confirmed by normal appearance of the vocal cords without any paradoxical movement detected by bronchoscopy.

DISCUSSION

Guillain-Barré syndrome (GBS) is a common cause of acute or subacute inflammatory demyelinating polyneuropathy; it occurs worldwide and in any season. It affects children as well as the adult of all age and both sexes.[1] The reported incidence rates varied from 0.4 to 1.7 cases per 100,000 persons per year.[1] A mild respiratory and gastrointestinal infections precede the neuropathic symptom by 1 to 3 weeks in about 60 to 70 percent of the cases.[1,2] Serologic studies reported that *Campylobacter jejuni* is the most frequent identifiable antecedent, and others include viral illness (Cytomegalovirus, Epstein-Barr virus,

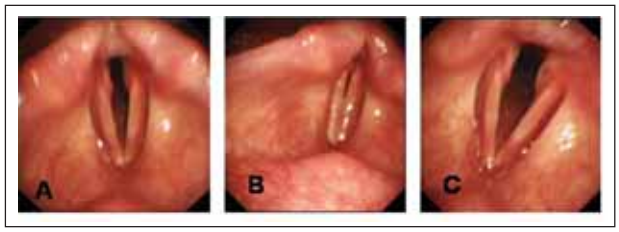


Fig 2. The bronchoscopic finding of bilateral paralyzed vocal cords in mid position (A). The paradoxical movement of the vocal cords was demonstrated by adduction of the vocal cords during deep inspiration (B) causing upper airway obstruction and abduction of the vocal cords on expiration

HIV), bacterial (*Mycoplasma pneumoniae*, Lyme's disease), the administration of antirabies vaccines and swine influenza vaccine was associated with a slight increase in the incidence of GBS.[1,2] The evidence of GBS pathogenesis is supported by autoimmune basis which both cellular and humoral immunological reactions.[1]

The earliest symptom of GBS is paresthesia and slightly numbness on the peripheral of both limbs, while the major clinical manifestation is rapid progressive areflexic motor paralysis symmetrically over a period of several days to a week or two, the proximal as well as the distal muscles of the limbs are involved. Usually, the lower extremities are affected before the upper (Landry's ascending paralysis), the trunk and neck; the cranial muscles may also be affected, and almost 30 percent of the patients require ventilatory assistance due to respiratory muscle weakness.[1,2] Disturbance of autonomic function (sinus tachycardia, bradycardia, facial flushing, fluctuating hyper-hypotension, loss of sweating, profuse diaphoresis) are common and frequent persist for more than a week.[1]

Diagnosis of GBS is made by recognition of the pattern of the rapidly evolving paralysis with areflexia, other systemic symptoms and other characteristic antecedent events. Also, CSF findings are markedly elevated protein concentration (100 to 1,000 mg/dl) with normal pressure, sugar, without accompanying pleocytosis (excepted in HIV patient).[2,3] Electrophysiologic studies are characteristics of a demyelinating process with slow conductive velocities. The conditions which should be differentiated for GBS include Polymyositis (West nine virus, enterovirus), carcinomatosis meningitis, Myasthenia gravis, botulism, tick paralysis, tetradotoxin, paralytic porphyria, diptheria and electrolyte imbalances.[1,2]

The specific treatment of GBS is either a high dose intravenous immunoglobulin (IVIg) or plasmapheresis, as they are equally effective; a combination of the two therapies is not significantly better than either alone. Glucocorticoid has not been found to be effective in GBS.[1,3] The early intervention for supportive treatment possible decreased morbidity and mortality, such as respiratory failure, unstable arrhythmia.[2,4] In the worsening phase of GBS, most patients require monitoring in a critical care setting, with particular attention to vital capacity, cardiovascular status and chest physiotherapy.[1] The rehabilitation program should be recognized to regain the patient's functional abilities. The majority of patients (about 85 percent) recovered nearly completely within several months to a year. However, about 10 percent were with pronounced residual capacity, and the mortality rate is 3 to 5 percent even in the best-equipped setting.[1]

Vocal cord paralysis in GBS has infrequently been reported. It may be unreported or unrecognized in intubated cases.[3,4] There were a total of 5 reported cases in the literatures. All reported patients presented the symptoms and signs of upper airway obstruction. In two cases, vocal cord paralysis occurred in the evolution of Guillain - Barré syndrome. Mechanical ventilator was needed in all cases. The paradoxical movement of the vocal cord was reported in only one case out of five as a 33 -year- old women presented with acute onset of respiratory distress and stridor.[5] The vocal cords were normal in appearance and relatively immobile in the paramedian position. Paradoxical motion of the vocal cords on inspiration was detected by fiberoptic laryngoscopy. Subsequently, the progressive weakness of the limbs occurred which led to elective intubation. The patient was ventilated for 6 weeks after treatment and was successfully decanulated without any recurrent stridor.

A review of etiology of bilateral vocal cord paralysis in 240 adults showed only 3 patients who had the paralysis caused by GBS. The most common (58 percent) was thyroidectomy, followed by neurological disease (22 percent) such as poliomyelitis, Parkinson s disease, cerebrovascular accident, GBS, multiple sclerosis, CNS infection, Charcot-Marie-Tooth disease and others, while malignancies of the neck were 6 percent of them and the rest 14 percent were miscellaneous.[6]

This reported patient presented with typical manifestation of GBS from the beginning. He was later intubated and received complete a 5-day course of IVIg treatment. He responded well to the point that he could be extubated successfully, while we detected upper airway obstruction caused by bilateral paradoxical movement of the vocal cords. The other causes of upper airway obstruction were excluded by bronchoscopic examination. The vocal cord paralysis can cause upper airway obstruction in some but not in every patient depending on the distance between the two paralyzed cords. Also, in rare situation that paralyzed cord moved paradoxically. Even in the five reported cases, only one showed paradoxical movement of the paralyzed cord. This movement could not explain by the venturi effect of the airflow because the adduction of the cord appeared only during inspiratory phase but the contribution of this phenomenon could not be excluded based on the findings that paradoxical movement occurred during the recovery of the respiratory muscles. The paradoxical movement of vocal cord was found mostly in paradoxical vocal cord dysfunction syndrome which is closely linked to psychogenic respiratory distress. This means there is some influence of higher brain function and the appearance of this movement during the recovery of the paralyzed cord which may unmask the higher brain control of the cords in some patients. This abnormal movement can also be found in other diseases such as chronic cough, muscle tension dysphonia[7] and multiple system atrophy. Recovery of the function of the vocal cords parallels with the disappearance of its paradoxical movement which occurred in one week in this case. The successful treatment of paradoxical vocal cord movement with noninvasive ventilation was reported by Nonake M, et al.[8] in patients with multiple-system atrophy and we also had the same success in this reported case.

In conclusion, we report a case of Guillain-Barré syndrome who developed paradoxical movement of paralyzed vocal cords causing upper airway obstruction. It responded to noninvasive ventilation. The paradoxical movement resolved in 12 days after the first dose of IVIg treatment.

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บทคัดย่อ

การขยับของเส้นเสียงแบบ Paradoxical ในผู้ป่วย Guillain- Barré syndrome: รายงานผู้ป่วย 1 ราย

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ภาควิชาอายุรศาสตร์, คณะแพทยศาสตร์ศิริราชพยาบาล, มหาวิทยาลัยมหิดล, กทม. 10700, ประเทศไทย.

รายงายผู้ป่วยชาย อายุ 50 ปี ได้รับไว้ในโรงพยาบาลศิริราช ด้วยอาการสำคัญอ่อนแรงมากขึ้นเรื่อย ๆ มา 1 สัปดาห์และหายใจไม่สะดวกมา 1 วัน หลังเข้ารับการรักษาได้รับการใส่ท่อช่วยหายใจ เนื่องจากการหายใจวาย และได้รับการวินิจฉัยว่าเป็นโรค Guillain-Barré syndrome ผู้ป่วยตอบสนองดีต่อการรักษาด้วย immunoglobulin ทางเส้นเลือดดำ จนสามารถหายใจได้เอง หลังจากถอดท่อช่วยหายใจเกิดอาการหายใจลำบาก ตรวจพบว่ามีลักษณะของหลอด ลมใหญ่อุดกั้นจาก spirometry และพบว่าเส้นเสียงมีการขยับแบบ paradoxical ตอบสนองดีต่อการใช้เครื่องช่วยหายใจแบบภายนอก อาการหายใจลำบาก ดีขึ้นใน 5 วัน หลังจากนั้นสามารถหยุดการช่วยหายใจได้ ผู้วิจัยได้รวบรวมสารนิพนธ์เกี่ยวกับการเกิดภาวะนี้