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Isolated vocal cord paralysis mimicking respiratory weakness in MuSK myasthenia

[1]Naveen Kumar Paramasivan *MD*, [1]Kavadisseril Vivekanandan Vysakha *MD DM*, [2]Kiran Vishnu Narayan *MD DM*, [3]Jayaprabha Sathyabhama *MS*, [1]Sruthi S Nair *MD DM*

[1]Department of Neurology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram; [2]Department of Pulmonary Medicine, and [3]Department of Otorhinolaryngology, Government Medical College, Thiruvananthapuram, India

Abstract

Vocal cord paralysis is a rare manifestation of myasthenia gravis and challenging to diagnose with presentations other than stridor. We discuss a patient with MuSK myasthenia gravis who had respiratory difficulty due to vocal cord palsy and the diagnostic difficulties encountered in establishing the etiology for the dyspnea. The diagnosis was captured by spirometry flow volume loop showing flattening of the inspiratory portion suggestive of extrathoracic upper airway obstruction. Early diagnosis can prevent the need for tracheostomy and other emergent measures.

Keywords: MuSK, myasthenia gravis, vocal cord palsy, stridor, respiratory distress

INTRODUCTION

Myasthenia gravis is a common neurological cause of respiratory distress. Shortness of breath in myasthenia is often presumed to be due to respiratory pump weakness but has infrequently been reported to be related to vocal cord paralysis. We present a case of muscle specific kinase (MuSK) myasthenia gravis who had dyspnea related to vocal cord palsy as the presenting feature of a relapse. We discuss the diagnostic difficulties encountered in determining the cause of respiratory difficulty in this patient and the use of ancillary tests in reaching a conclusion.

CASE REPORT

A 36-year-old lady presented in October 2012 with insidious onset, progressive ptosis, diplopia and fatigable limb weakness of 5 years duration and recent worsening with nasal regurgitation and hoarseness. A decrement of 24% was recorded from left spinal accessory nerve on repetitive nerve study (RNS), and increased jitter was noted from extensor digitorum communis and frontalis muscles in single fiber electromyography (SFEMG), consistent with myasthenia gravis. Neostigmine test and acetyl choline receptor antibodies were negative. Baseline CT thorax and ear-nose-throat evaluation were normal. She was initiated on Pyridostigmine 180 mg/day and prednisolone 10 mg/day. Within one month, she presented with worsening symptoms of bulbar weakness and respiratory distress requiring mechanical ventilation. Anti-MuSK antibody titre obtained at this time was positive (2.6 nmol/L, normal <0.05 nmol/l).

Following recovery from the crisis, she attained remission and was maintained on oral prednisolone (10mg/day) and azathioprine (100 mg/day) for nearly 5 years. In 2017, she started having dyspnea on exertion with sudden awakening from sleep due to breathlessness. There was no orthopnea, nocturnal wheezing, dyspeptic symptoms, recent weight gain or loud snoring. Clinical evaluation did not show fatigability, respiratory paradox or stridor. The single breath count was 44. A baseline finger pulse oximetry saturation showed 98% with no exercise desaturation but she could complete only 3 minutes with 200 metres on a 6-minute exercise walk test. The test was prematurely stopped due to limiting dyspnea and harsh breath sounds. Speech and swallow evaluation were normal. RNS was negative from spinal accessory, facial and ulnar nerves. A transthoracic echocardiography showed normal cardiac function and pulmonary arteries. A chest radiograph showed adequate lung volumes with normal cardiac silhouette and normal diaphragm position. A CT neck and thorax revealed normal lung fields and no thymoma or

Address correspondence to: Dr Sruthi S Nair, Department of Neurology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Trivandrum 695011, Kerala, India. Tel: 91 471 2524488, Email: sruthisn@sctimst.ac.in, sruthisn@yahoo.com

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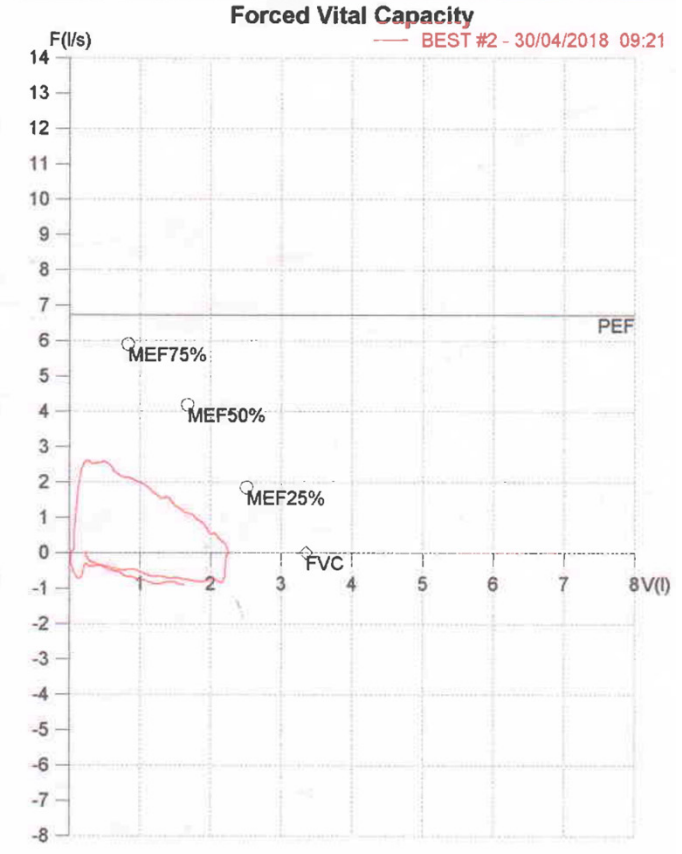
mediastinal adenopathy. Fluoroscopy showed normal diaphragm excursion. Spirometry showed a mild restrictive ventilatory defect (forced vital capacity of 67%) which could not explain the marked dyspnea. The spirometry flow volume loop showed a flat peak and flattening of the inspiratory portion which suggested an extrathoracic upper airway obstruction (Figure 1). This led to the suspicion of an upper airway disease and bilateral vocal cord abductor palsy was confirmed with fibreoptic videolaryngoscopy (Figure 2).

In the absence of a structural pathology to account for bilateral laryngeal abduction weakness, and sparing of other tenth nerve innervated structures, the vocal cord palsy was postulated to be due to neuromuscular junction transmission defect. The dose of prednisolone was hiked to 40 mg/day with which she attained remission of exertional dyspnea and nocturnal attacks. Immunomodulation was optimized with azathioprine, however she continued to remain steroid dependent, unable to reduce the dose of oral prednisolone to less than 10mg/day. At her last follow up in September 2021, she remained in remission.

DISCUSSION

Unusual presentations such as vocal cord paralysis have been described as the initial manifestation of both seropositive and seronegative myasthenia gravis patients.[1-4] In some patients, a precipitating



Age (years): 41, Sex: Female, Weight (kg): 74.0, Height (cm): 165.0				
Parameter	**Unit**	**Predicted**	**Test**	**% predicted**
Forced Vital capacity (FVC)	L (btps)	3.35	2.24	67
Forced Expiratory Volume in 1 second (FEV1)	L (btps)	2.89	1.81	62
FVC/FEV1	%	81.3	80.6	99
Peak Expiratory Flow (PEF)	L/s	6.73	2.60	39
Peak Inspiratory Flow (PIF)	L/s		0.84	

Figure 1. Spirometry flow volume loop of the patients shows flattening of the inspiratory loop suggestive extrathoracic upper airway obstruction.
Abbreviations: btps-body temperature, pressure, water vapor saturated, L-Litre, s-second

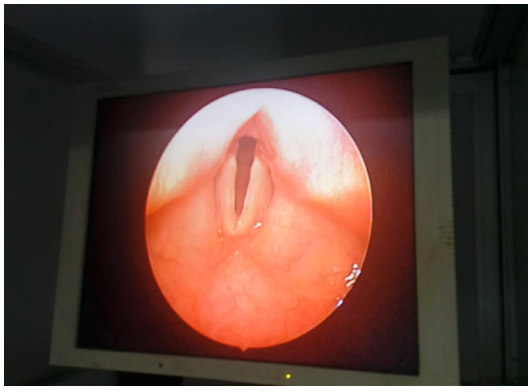


Figure 2. Snapshot of fibreoptic nasal endoscopy of the patient with myasthenia gravis showing incomplete bilateral vocal cord abductor movement. Vocal cord abduction was slow and minimal, and adduction was normal. Glottic space was just adequate.

factor such as surgery with use of depolarizing drugs could be identified as a trigger, whereas no precipitating cause could be ascertained in others. In all these reports, patients were not diagnosed and treated for myasthenia prior to the presentation of vocal cord paralysis.

On the contrary, our patient was in remission of the disease when she relapsed with dyspnea and no apparent triggers. The diagnostic pointers to determining vocal cord palsy as the etiology of dyspnea were the documentation of normal mobile vocal cords at her initial evaluation and confirmation of vocal cord paresis during the current symptomatic phase, recent onset dyspnea with no other contributing factors as shown by normal cardiac evaluation, CT chest and diaphragm function, and an inspiratory flat peak of flow volume loop on spirometry suggesting extrathoracic obstruction. Rapid and complete improvement of the symptoms with hiking the dose of steroid confirmed the diagnosis.

Respiratory distress in patients with myasthenia gravis could be due to weakness of the respiratory muscles themselves (predominantly diaphragm) or could be due to physiological airway obstruction secondary to vocal cord paralysis. Vocal cord paralysis leads to narrowing of the glottic area, and thereby increases airway resistance. The forced inspiration into a partially closed glottis causes an inspiratory stridor.[5] Most of the patients with this presentation required a tracheostomy as the disease was not recognized early at the onset. Timely recognition and intervention can avoid the need for tracheostomy in many patients.[6] The functional airway obstruction in our patient was the cause for intermittent dyspnea. The narrowed upper airways caused a flow limitation during exercise and may explain the exertion-induced dyspnea. During sleep, the physiological respiratory drives are diminished, leading to reduced tone of muscles further increasing airway resistance which could result in nocturnal dyspnea and awakening as reported in our patient. The absence of stridor is intriguing and may be explained by a milder vocal cord palsy. A more severe presentation may have been masked by the fluctuating weakness and the immunomodulators she was already on. Dyspnea in myasthenia can be multifactorial and is especially puzzling in situations when no fatiguability is demonstrable. A good clinicoradiological examination along with pulmonary function testing helped in narrowing down the cause of dyspnea. Recognizing flow volume loop patterns are important to get an idea of unexplained dyspnea, especially when the lung volumes are relatively normal as seen in our patient.

In conclusion, unexplained dyspnea in patients with myasthenia gravis should raise the suspicion of laryngeal weakness. Timely identification is critical in these patients to prevent emergency interventions.

DISCLOSURE

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Conflict of interest: None

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