

Myasthenia Gravis: A Rare Cause of Orthopnoea due to Bilateral Diaphragmatic Paralysis

K.M.C. Nishantha and D. Madegedara

Respiratory Disease Research and Treatment Unit, Teaching Hospital, Kandy, Sri Lanka

ABSTRACT

A case of a 68-year-old patient with bronchial asthma who presented with orthopnoea and respiratory failure in supine position is presented. [Indian J Chest Dis Allied Sci 2011;53:189-190]

Key words: *Myasthenia gravis*, Diaphragmatic paralysis, Orthopnoea.

INTRODUCTION

We describe case of a 68-year-old patient with controlled bronchial asthma who presented with respiratory failure in supine position following bilateral diaphragmatic paralysis due to *Myasthenia gravis*. He complained of progressive orthopnoea of eight months duration. Neurological examination was normal. Fluoroscopy in both the standing and supine positions revealed bilateral diaphragmatic paralysis that accentuated in the supine position. Electromyographic studies suggested a diagnosis of *M. gravis*. *To the best of our knowledge*, this is the first case of isolated diaphragmatic paralysis associated with *M. gravis* in adults presenting with supine respiratory failure.

CASE REPORTS

A 68-year-old businessman presented with insidious onset progressive shortness of breath that markedly increased in the latter part of the day for eight months. During the last three months of his illness, he became markedly short of breath on lying down. There was no cough, wheezing or ankle oedema.

On examination, he was tachypnoeic with diminished breath sounds bilaterally in the lower zones. In upright position, the respiration was normal. However, in the supine position, the patient immediately complained of severe dyspnoea and exhibited thoraco-abdominal paradoxical breathing with the use of accessory muscles of respiration. There were no other remarkable signs on general and systematic examination, including a

comprehensive neurological work-up. Chest radiograph demonstrated elevation of the both hemidiaphragms with bilateral basal atelectasis. Subsequent fluoroscopy of the diaphragms demonstrated minimal excursion of both the left and right diaphragm, with no paradoxical motion identified on the sniff test. Arterial blood gas values obtained in the sitting position on presentation were: a pH of 7.38; arterial carbon dioxide tension (PaCO₂), 42 mmHg; arterial oxygen tension (PaO₂), 94 mmHg; and HCO₃, 23 mEq/L. Pulse oxymetry showed an oxygen saturation of 94% in the sitting position and that it dropped to 88% in the supine position. Spirometry in the sitting position showed a forced vital capacity (FVC) of 2.03 L (74% of predicted), forced expiratory volume in one second (FEV₁) of 1.03 (48%), and FEV₁/FVC of 92% consistent with a restrictive ventilator impairment. His maximum voluntary ventilation (MVV) was 30% of the predicted value. The diffusion capacity (DLCO) was normal. A cervical spine radiograph was unremarkable. High resolution computerised tomography of the chest revealed bi-basal atelectasis. Phrenic nerve stimulation at neck showed an absent response of the diaphragm bilaterally with preserved conduction through phrenic nerves. Repetitive nerve stimulation study of the diaphragm revealed significant decremental potentials suggestive of *M. gravis*. The Tensilon test was performed to assess the improvement in the MVV and showed a 4% rise. Acetylcholine receptor antibody level was also within the normal range (<0.07nmol/L). Results of chemical profile, haematological profile, cardiologic assessment, serial creatinine phosphokinase and thyroid-stimulating hormone were in the normal range.

[Received: March 3, 2011; accepted: May 25, 2011]

Correspondence and reprint requests: Dr Dushantha Madegedara, Consultant Respiratory Physician, Teaching Hospital, Kandy, Sri Lanka; Phone/Fax: 0094812384374; E-mail: dmadegedara@yahoo.com

DISCUSSION

Among neuromuscular disease and conditions, bilateral diaphragmatic paralysis has been recognised as a cause of respiratory failure since the decline of poliomyelitis and diphtheritic poly-neuropathy.¹

We investigated the occurrence of orthopnoea in a patient with well controlled bronchial asthma. Lung function tests of the patient revealed a restrictive ventilatory defect with a very low MVV. We excluded parenchymal lung disease and extra-thoracic pathology. Since the echocardiogram was normal, neuro-muscular or muscle diseases were considered as the cause of weakness of respiratory muscles, and thereby, causing orthopnoea and a restrictive lung defect. Significant postural desaturation of oxygen favoured a diagnosis of bilateral diaphragmatic weakness.

Causes of diaphragmatic weakness are diverse and include, motor neuron disease,² post-polio syndrome, thoracic trauma,³ multiple sclerosis, myopathies,⁴ Lyme disease⁵ and muscular dystrophy (acid maltase deficiency) etc.

Elevated hemi-diaphragms on chest radiography, compared to a normal chest film taken two years back and significant decremental responses on accessory muscles, deltoid and diaphragms following repetitive nerve stimulation confirmed muscle weakness. The MVV was recorded before and after the administration of Tensilon (edrophonium). Although there was no significant improvement of MVV, the duration of the effort of the patient increased significantly after the injection of Tensilon.

The patient was started on neostigmine, azathioprin and prednesolone. On follow-up at four weeks, he reported improvement and was able to sleep without arousal. The supine oxygenation improved significantly from 88% to 94%.

This case highlights the importance of a good clinical history that pointed to other possible causes of the shortness of breath, besides being an asthmatic. The critical analysis of the spirometry findings together with the MVV pointed towards a diaphragmatic weakness. Meticulous search for the cause of bilateral diaphragmatic weakness resulted in diagnosing this rare but treatable aetiology.

REFERENCES

1. Luce JM, Huseby JS, Marini JJ. Bilateral diaphragmatic paralysis as a cause of respiratory failure. *West J Med* 1980;132:456-60.
2. The prognosis for patients with BDP is uncertain and depends on the cause of the disorder. Respiratory failure due to bilateral diaphragm palsy as an early manifestation of ALS. 2003;9:CS34-6.
3. Spitzer SA, Korozyn AD, Kalaci J. Transient bilateral diaphragmatic paralysis. *Chest* 1973;64:355-7.
4. Abbott RA, Hammans S, Margarson M, Aji BM. Diaphragmatic paralysis and respiratory failure as a complication of Lyme disease. *J Neuro Neurosurg Psychiatr* 2005;76:1306-7.
5. Lin MC, Liaw MY, Huang CC, Chuang ML, Tsai YH. Bilateral diaphragmatic paralysis: a rare cause of acute respiratory failure managed with nasal mask bilevel positive airway pressure (BiPAP) ventilation. *Eur Respir J* 1997;10:1922-4.