Case Report

Idiopathic bilateral diaphragmatic dysfunction as a cause of dyspnea

D MacBruce[1,2], S Safdar[1,2], K Katpally[1,2], Hamid Shaaban[1,2], M Adelman[1,2]

Departments of [1]Internal Medicine and [2]Pulmonary and Critical Care Medicine, Saint Michael's Medical Center, Newark, NJ, An Affiliate of New York Medical College, NY, USA

ABSTRACT

Diaphragmatic paralysis is an unusual and often underrecognized cause of dyspnea. We present a case of bilateral diaphragmatic paralysis with no identifiable etiology. Our patient is a 73-year-old female with a history of smoking who presented with dyspnea and orthopnea. She was treated for obstructive lung disease with no improvement in dyspnea despite adequate therapy. She had pulmonary function tests (PFTs) that revealed marked decrease in vital capacity and was unable to perform lung volume maneuvers supine due to marked dyspnea. The maximal inspiratory pressure was 37 in the upright position and decreased to 27 in the supine position. She was given a presumptive diagnosis of idiopathic bilateral diaphragmatic dysfunction. Given the history, physical exam, and PFT findings, we felt that the patient did not need further invasive testing. The patient was treated with noninvasive mechanical ventilation due to hypercapnia and her symptoms improved.

KEY WORDS: Diaphragm, idiopathic, paralysis

Address for correspondence: Dr. Hamid Shaaban, 111 Central Avenue, Newark, NJ 07102, USA. E-mail: hamidshaaban@gmail.com

INTRODUCTION

Diaphragmatic paralysis is an unusual and often underrecognized cause of dyspnea. The diaphragm is the main muscle of respiration. It is a dome-shaped skeletal muscle dividing the trunk into thoracic and abdominal cavities. It is innervated by the phrenic nerve formed from the cervical C3, C4, and C5 nerve roots. Disease processes affecting the brain, the nerves, or the muscle may result in diaphragmatic dysfunction. Damage to the phrenic nerve due to injury during surgery or trauma or compression caused by bronchogenic or mediastinal tumors have been identified as the most common causes of diaphragmatic paralysis.[1-2] Other illnesses including Guillain–Barre syndrome, spinal cord injuries, myasthenia gravis, botulinum toxin, metabolic abnormalities such as hypophosphatemia, hypomagnesemia, hypokalemia, and hypocalcemia are some of the other causes of diaphragmatic weakness.[2-3] We present a case of bilateral diaphragmatic paralysis with no identifiable etiology.

Access this article online

Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/0970-2113.180919

CASE REPORT

The patient is a 73-year-old female, who was being evaluated for progressive dyspnea over a 1-year period. She stated that during the last 3 years she had been treated for a recurrent cough, wheezing and nasal stuffiness, with bronchodilators, inhaled steroids, leukotriene antagonists, and oral steroids with minimal improvement. Recently, she began sleeping with several pillows. She denied leg swelling. She was a former smoker, beginning at age 16, and smoked one pack of cigarettes a day until 20 years ago when she ceased smoking. She had no pertinent occupational history. There was a past medical history positive for hypertension, dyslipidemia, and hip surgery 3 years ago.

How to cite this article: MacBruce D, Safdar S, Katpally K, Shaaban H, Adelman M. Idiopathic bilateral diaphragmatic dysfunction as a cause of dyspnea. Lung India 2016;33:330-2.

Physical exam showed a blood pressure 114/70, pulse 87, and a respiratory rate of 17. The head and neck exam were normal. Lung sounds were diminished, and no wheezing was appreciated. The heart's rhythm was regular, and there were no gallops. The extremities were without edema.

Neurologically, the patient was alert and oriented. There were no cranial nerve abnormalities. The patient demonstrated a normal motor and sensory exam. Her deep tendon reflexes were normal in both upper and lower extremities.

An arterial blood gas (ABG) revealed a pH = 7.44, PCO_2 = 50, and PO_2 = 69. A chest X-ray showed poor inspiratory effort with no active cardiopulmonary disease. A noncontrast-enhanced computerized tomography (CT) of the lung demonstrated bibasilar segmental atelectasis. Pulmonary function testing [Table 1] showed a mixed picture with decreased flow rates, a severely reduced total lung capacity, and low DLCO. The maximal inspiratory pressure was 37 in the upright position and decreased to 27 on the supine position. Fluoroscopy of the diaphragm was performed, and this showed severely impaired diaphragmatic motion on normal tidal breathing as well as following deep inspiration [Figure 1a and b]. The following led us to a diagnosis of diaphragmatic dysfunction.

DISCUSSION

Dysfunction of the diaphragm can range from weakness to complete paralysis. The incidence of idiopathic bilateral paralysis of the diaphragm is unknown, and the disease may be underdiagnosed. A few cases have been reported in literature. In patient, no identifiable risk factors have been elucidated. There was no history of trauma, abdominal or thoracic surgery, or obvious neurologic or neuromuscular disorder.

Table 1: Pulmonary function test

PFT parameter	Observed	Predicted (%)
FVC (L)	0.89	31
FEV_1 (L)	0.51	24
FEV_1/FVC (%)	57	70
VC (L)	1.01	36
TLC (L)	2.16	43
RV (L)	1.15	56
DLCO	7.9	35
PI max	37	55

FVC: Forced vital capacity, FEV_1: Forced expiratory volume in 1 s, VC: Vital capacity, TLC: Total lung capacity, DLCO: Diffusing capacity of the lungs for carbon monoxide, PI max: Maximal inspiratory pressure, RV: Residual volume, PFT: Pulmonary function test

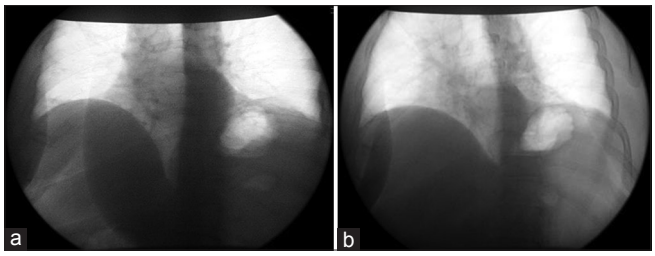

Figure 1: Fluoroscopy of the diaphragm was performed and this showed (a) severely impaired diaphragmatic motion on normal tidal breathing as well as (b) following deep inspiration

This patient presented with dyspnea out of proportion to the underlying medical illness, including obesity and obstructive airway disease. As is often the case, marked orthopnea was present, leading the patient to sleep in recliners. The paralyzed diaphragms are pushed up by abdominal content in the supine position, but in the upright position moved caudally with the help of gravity. As diaphragm function deteriorates, patients may be unable to generate a transdiaphragmatic pressure of more than 30 cm against a closed airway, and abdominal paradox may be seen on physical examination.[4] Our patient did not improve in spite of adequate treatment for underlying obstructive airway disease, warranting further evaluation of dyspnea.

On chest radiographs, elevation of the hemidiaphragms with small lung volumes were present but can be misinterpreted as a poor inspiratory effort not specific for diaphragmatic paralysis. A CT scan of the chest revealed bibasilar atelectasis which would be unlikely in patients with chronic obstructive pulmonary disease or congestive heart failure. While our patient demonstrated no diaphragmatic movement on the "sniff" test, it is no longer considered the test of choice in diagnosing bilateral diaphragmatic paralysis. Up to 6% of normal patients may have false positive test of cephalad displacement, or <2 cm caudal excursion of the diaphragm on inspiration. Contraction of the abdomen during inspiration in severely ill patients may result in a false negative test as the diaphragm may appear to move caudally on expiration as the abdominal muscles relax.[2-4]

Decrease between 30% and 50% of predicted forced vital capacity measured in the upright position on pulmonary function test (PFT) may help support the diagnosis, is readily available, relatively inexpensive and noninvasive.[5] Our patient demonstrated severe restriction with 36% of predicted vital capacity in the upright position. Although our patient was unable to perform the PFT in the supine position, a further decrease in the forced vital capacity is expected. The maximal static inspiratory pressure can be reduced to about 30% of predicted in severe bilateral diaphragm paralysis. Our patient demonstrated a reduction of approximately 50% in the upright position which was further reduced in the supine position. This test is effort dependent and variable. Maximum expiratory pressure, residual volume, and functional residual capacity should remain preserved as is expected in disease affecting the diaphragm while sparing the muscles of active expiration. Arterial blood gas determinations may show hypoxemia with underlying ventilation/perfusion mismatch and progressive hypercapnia as respiratory failure develops.

Measurement of diaphragmatic thickness by ultrasonography at the zone of apposition with the rib cage has been used in a small series to confirm and assess recovery of diaphragmatic paralysis. M-mode ultrasonography is the latest method to evaluate a paralyzed diaphragm, from which the paralyzed hemidiaphragm shows no active caudal movement with inspiration. Electromyography of the diaphragm during quiet breathing or stimulation of the phrenic nerve is considered the gold standard but is of limited use in clinical practice. The test is invasive, difficult to interpret and can be technically difficult to perform.[5-7]

While idiopathic diaphragmatic paralysis may improve spontaneously over a long course, it may persist or progress, and patients may require ventilator support particularly at night. Patients with diaphragmatic paralysis can eventually develop progressive irreversible ventilatory failure resulting from the fatigue of the accessory muscles. Patients are started on ventilator support at PCO_2 ≥45 and/or oxygen saturation ≤88%.[6] Patients with cor pulmonale also may manifest improvement in function and the correction of blood gas abnormalities with nighttime or intermittent daytime noninvasive ventilation. A tracheostomy with positive-pressure intermittent or permanent ventilation is reserved for patients with life-threatening disease. Plication of the diaphragm has been used in severe symptomatic unilateral disease but is contraindicated in obese patients and bilateral disease.[5-7]

Phrenic nerve pacing is being evaluated for the treatment of diaphragmatic dysfunction.

CONCLUSION

Diaphragmatic dysfunction, although rare, should be considered in the evaluation of dyspnea.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Kumar N, Folger WN, Bolton CF. Dyspnea as the predominant manifestation of bilateral phrenic neuropathy. Mayo Clin Proc 2004;79:1563-5.
2. Brander PE, Järvinen V, Lohela P, Salmi T. Bilateral diaphragmatic weakness: A late complication of radiotherapy. Thorax 1997;52:829-31.
3. Davis J, Goldman M, Loh L, Casson M. Diaphragm function and alveolar hypoventilation. Q J Med 1976;45:87-100.
4. Gierada DS, Slone RM, Fleishman MJ. Imaging evaluation of the diaphragm. Chest Surg Clin N Am 1998;8:237-80.
5. Murray JF, Nadel JA, editors. Textbook of Respiratory Medicine. Philadelphia, Pa: WB Saunders; 1997.
6. McCool FD, Tzelepis GE. Dysfunction of the diaphragm. N Engl J Med 2012;366:932-42.
7. Onders RP, Elmo M, Kaplan C, Katirji B, Schilz R. Extended use of diaphragm pacing in patients with unilateral or bilateral diaphragm dysfunction: A new therapeutic option. Surgery 2014;156:776-84.