

Bilateral Phrenic Nerve Palsy in a Diabetic Causing Respiratory Failure

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Abstract

Neuropathy is one of the major reasons of morbidity in diabetes mellitus (DM). We presented a 64-year-old male who was a case of type 2 DM for >6 years. He presented with orthopnea and respiratory failure secondary to bilateral phrenic neuropathy and resultant diaphragmatic palsy. Clinical examination, ultrasound, and nerve conduction studies confirmed the bilateral involvement of the phrenic nerves. Phrenic neuropathy may be an important, albeit a rare complication of diabetes, and hence, diaphragmatic dysfunction associated with diabetic phrenic neuropathy should be considered in any patient with unexplained breathlessness, orthopnea, and respiratory failure.

Keywords: Diabetes neuropathy, phrenic nerve, respiratory failure

INTRODUCTION

Diabetes mellitus (DM) is a major health problem for the population because of its high mortality and morbidity and costs of therapy.^[1] DM is the most common cause of peripheral neuropathy in the developed world. The prevalence of diabetic neuropathy varies between 5% and 60% in different studies.^[1,2] Bilateral symmetrical sensorimotor distal polyneuropathy is the most frequent type.^[1,2] Although rare, diabetes can affect phrenic nerves causing unilateral or bilateral diaphragmatic paralysis with subsequent respiratory compromise.^[3-5] Other causes of diaphragmatic palsy need to be investigated and excluded before making a provisional diagnosis of diabetic phrenic nerve neuropathy. Although diabetic mononeuropathy is common and usually recovers spontaneously, bilateral phrenic nerve neuropathy appears to be uncommon and difficult to treat, resulting in the need for long-term respiratory assistance.^[3,4]

CASE REPORT

A 64-year-old male, known to have type 2 DM for more than 6 years, presented to the emergency department with respiratory distress. He had a history of burning sensation in his feet in the past 1 year and gradual decrease in power of distal lower limb muscles (flopper coming out of his feet while walking), which progressed to proximal muscle

weakness. He was under treatment in the outpatient for the past 6 months for a progressive diabetic neuropathy on duloxetine, gabapentin, and nortriptyline. Despite his blood sugar being well controlled (Hb1Ac 7.7) on oral hypoglycemic agents, he continued to have increasing symptoms of weakness and inability to walk due to proximal muscle involvement. He started using a walker at home for the past 2 months and for the past 2 weeks before admission, complained of inability to lie down supine and sleep. He would prop himself up with pillows to avoid breathlessness. He had no history of smoking or substance abuse. He was admitted to the intensive care unit of a tertiary center with respiratory distress in the form accessory use of respiratory muscles, tachypnea, and fall in oxygen saturation. And therefore, put on noninvasive ventilation. On examination, air entry into his lungs was reduced bilaterally with no wheeze or crepitations. He was more comfortable in propped up position compared to supine. His plantar reflexes were absent bilaterally with reduced knee jerks 1+ bilaterally. Power in his lower limb was 3/5 in the proximal and distal muscle groups. The rest of the examination was normal. Over

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the next 2 weeks, his dyspnea worsened and he developed desaturation with altered mental status, with arterial blood gas showing pH of 7.28, PO₂ 81 mmHg, HCO₃ of 31 mEq/L, and PCO₂ of 78 mmHg, developing hypercarbic respiratory failure requiring endotracheal intubation and mechanical ventilation. His mentation and PCO₂ improved after starting mechanical ventilation.

His blood count, electrolytes, thyroid function tests, B-natriuretic peptide, cardiac enzymes, serum protein electrophoresis, and anti-acetylcholine receptor antibodies were all normal. Chest radiogram revealed elevated right dome of diaphragm [Figure 1]. An echocardiogram was normal. Computed tomography (CT) of the chest revealed bilateral posterior lower zone atelectasis [Figure 2]. No other mass or lesion was present in lungs or mediastinum. CT pulmonary angiography did not reveal any evidence of pulmonary embolism. Ultrasound suggested bilateral diaphragmatic palsy, with right hemidiaphragm being totally immobile and partial mobility of the left hemidiaphragm. Since the investigations mentioned above were inconclusive, a differential diagnosis of Lambert–Eaton syndrome, Guillain–Barre syndrome, and bilateral diabetic phrenic neuropathy was considered. Therefore, he was treated empirically with intravenous immunoglobulin and other supportive treatment. As it did not improve, nerve conduction studies (NCSs) of peripheral nerves were done. Tibial nerve conduction parameters were distal motor latency of 3.18 ms, distal motor amplitude of 9.31 mV, motor conduction velocity 52.12 m/s, and F-wave of 49.44 ms. Sural nerve conduction parameters were distal sensory latency of 2.15 ms, sensory amplitude of 19.90 μ V, and sensory velocity of 41.19 m/s. These conduction parameters were suggestive of severe sensorimotor degeneration with axonal damage of peripheral nerves, mainly involving sural and tibia nerves bilaterally. Phrenic nerve was transcutaneously stimulated at the posterior border of the sternocleidomastoid muscle in the supraclavicular fossa, just above the clavicle, using bipolar surface bar electrodes with the cathode placed

caudally. A constant-current stimulator delivering rectangular pulses of 0.2-ms duration was used. NCS of phrenic nerves showed right phrenic nerve latency and amplitude as 8.9 ms and 0.3 mV, respectively, and left phrenic nerve latency and amplitude as 8.4 ms and 0.35 mV. These conduction parameters were suggestive of bilateral phrenic nerve neuropathy.

Hence, a diagnosis of diabetic phrenic nerve neuropathy leading to diaphragm palsy was considered after ruling out other causes of diaphragmatic palsy such as viral infections, Guillain–Barre syndrome, myasthenia gravis, spinal cord injuries, or tumors of neck and mediastinum based on patient's history, progression of disease, and investigations. Multiple trials to wean the patient off ventilator support failed over a period of 2 months, and he was ventilator dependent. He was discharged with home ventilator support in a stable condition. A professional team now looks after him at home.

DISCUSSION

Neuropathy, one of the major complications of DM, is an important cause of morbidity and mortality. Injury during cardiac surgery is the most commonly reported cause of phrenic nerve palsy, although surgery involving spine, shoulder, head, and neck can also be causative.^[6] Other etiologies include tumors of the neck and mediastinum, viral infections, brachial plexus neuropathy, vasculitis, neuropathies, and spinal cord injuries. However, in a significant number of cases, no cause is found.^[7] Patients receiving duloxetine and gabapentin have reported to have complaints of nonspecific muscle weakness and fatigability. However, the pathogenesis of muscle weakness with the use of these drugs is unknown. Diabetic phrenic neuropathy was the only identifiable etiology in our patient. Paradoxical respiratory breathing suggested by paradoxical movement of the abdominal wall during inspiration, as seen in our patient, can be an important clue in the presentation. Once physical examination and chest imaging, with subsequent verification by CT, confirmed

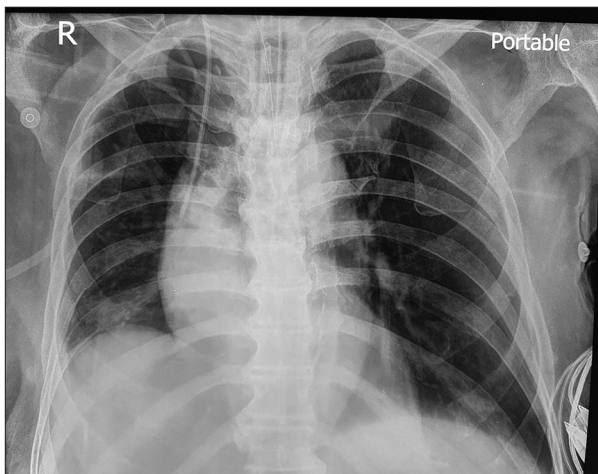


Figure 1: Chest X-ray anteroposterior view on admission showing elevated right hemidiaphragm

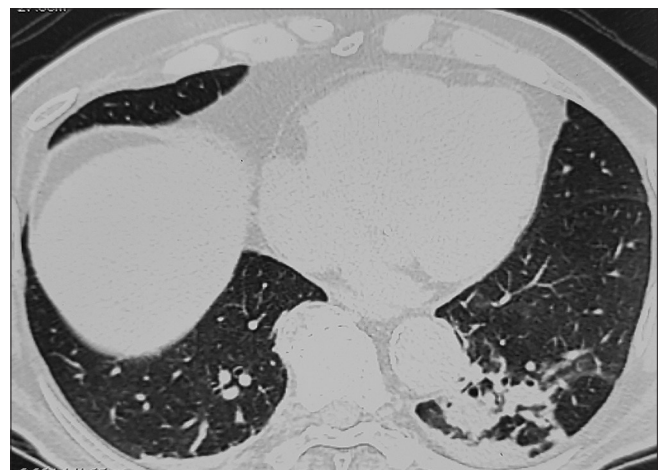


Figure 2: Computed tomography of the chest revealed bilateral lower posterior lobe atelectasis which was not marked as the patient was on positive pressure ventilation

elevated diaphragm, ultrasonography characteristics have been used to detect and monitor diaphragmatic paralysis.^[3,7] There is no correlation between the presence of phrenic neuropathy and peripheral neuropathy or the degree of glycemic control.^[8] In a study by Yesil *et al.*, bilateral phrenic and peripheral NCSs were performed in a total of 37 diabetic, 40 prediabetic patients, and 18 healthy controls, which showed that the amplitudes of phrenic nerve were lowest in diabetic patients and highest in control group; latencies of phrenic nerve were longer in prediabetic and diabetic patients whereas conduction velocities of all sensory nerves except ulnar nerve were prolonged and statistically significant.^[9] Most cases resolve spontaneously and do not require treatment, although spontaneous recovery is minimal after 2 years.^[10] When there is a functional limitation, surgical plication remains the gold standard.^[7] Diaphragm resections and video-assisted minimally invasive surgery have also been effective.^[7] As for the medical treatment, one case report described successful use of topiramate over 26 weeks for diabetic phrenic nerve palsy.^[5] Continuous positive airway pressure (CPAP) or in worse cases, mechanical ventilation may be used to prevent ventilatory failure in patients with bilateral paralysis, in pediatric patients and can be used as a bridge for patients awaiting surgical correction.^[11,12]

We suggest that diabetic patients with orthopnea and unexplained breathlessness should be considered for diaphragmatic dysfunction due to phrenic neuropathy, and a review of patients' medication list is essential to analyze any drugs aggravating muscle weakness.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published

and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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