Clinical Vignettes

Unilateral Diaphragmatic Paralysis in a Diabetic Patient: A Case of Trepopnea

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Dyspnea is a common presenting complaint. Trepopnea, an under-recognized form of dyspnea, is difficult breathing in only one lateral decubitus position. One cause of trepopnea is unilateral diaphragmatic paralysis, which in itself is an uncommon diagnosis. We report a unique case of a 55-year-old diabetic man who presented with trepopnea and was found to have unilateral diaphragmatic paralysis secondary to isolated diabetic phrenic neuropathy. This case highlights the importance of recognizing trepopnea as an early clinical symptom of diaphragmatic paralysis and discusses diabetic phrenic neuropathy which can occur in the absence of peripheral neuropathy.

KEY WORDS: trepopnea; diabetes; neuropathy; phrenic; diaphragm; paralysis; dyspnea; sniff-test.

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INTRODUCTION

Shortness of breath is a common presenting symptom. In the spectrum of the "pneas," orthopnea, and platypnea are commonly recognized. An often forgotten type of dyspnea is trepopnea. Trepopnea depicts having difficulty breathing in one lateral recumbent position and originates from the Greek word 'trepo' meaning to twist or turn^{1,2}. Originally described by Francis Wood and initially known as "rotopnea," the phenomenon was later renamed to "trepopnea." This clinical sign was commonly described in patients with congestive heart failure (CHF) in the literature from the middle part of the 20th century but suffered from disuse over the years. Trepopnea reemerged recently in case reports of aneurysms of the sinus of valsalva, right-to-left interatrial shunts, recurrent lung cancer and cardiac tumors³⁻⁶. Hitherto, trepopnea has not been described in association with unilateral diaphragmatic paralysis, which in itself is an uncommon cause of respiratory complaints that may be caused by diverse etiologies. We present a unique case of a diabetic patient with unilateral diaphragmatic paralysis who presented with trepopnea.

CASE REPORT

A 55-year-old white man with diabetes mellitus, hypertension, and a history of cerebrovascular accident (CVA) presented to

the emergency room with a two month history of shortness of breath of an insidious onset. He had difficulty breathing only when he lay on his left side, with remarkable improvement when he shifted to the right lateral decubitus position. He also experienced shortness of breath after walking about half a block. When the patient slept on his left side, his wife observed some rapid exaggerated breathing movements on the right side of his chest waking him up at night. Snoring was not noted. He denied chest pain, dizziness, palpitations, pedal edema, cough, fever, weight loss or trauma. The patient was evaluated by his primary care physician and took furosemide without any symptomatic improvement.

He had had diabetes mellitus for 10 years with the latest hemoglobin A1c of 7.2% and was on oral hypoglycemic medications. His hypertension was controlled with metoprolol tartrate and enalapril. He had had a CVA with mild residual left-sided weakness about 10 years ago; a brain MRI done in 2005 did not reveal any abnormality. He stopped smoking 25 years ago and did not drink alcohol. On review of systems, he denied rashes, joint pain, tingling and numbness in hands and feet, new neurologic symptoms, or any other systemic complaints.

On examination, he was an overweight man lying in bed on his right side with mild respiratory distress. He was afebrile, with a pulse of 92 beats per minute, a blood pressure of 130/72 mmHg, a respiratory rate of 20 per minute, oxygen saturation of 96% on room air and a BMI of 41.60 kg/m². He had no jugular venous distension, thyromegaly or lymphadenopathy in the neck. Cardiac examination revealed regular rate and rhythm without S₃, S₄ gallops, murmurs or a rub. His trachea was midline, and there was no accessory muscle usage. He had reduced chest expansion and dullness to percussion on the right side and decreased breath sounds in the right posterior lower lung field. Abdominal exam showed a transient episode of paradoxical thoraco-abdominal breathing movement on the right side suggested by unilateral subcostal and intercostal retraction during inspiration. No hepatomegaly was noted. Distal pulses were intact, and there was no edema. On neurological exam, he had residual 4/5 muscle strength and 3+ reflexes in the upper and lower extremities on the left side from prior stroke with a normal exam on the right side. There was no sensory deficit.

His blood count, electrolytes, thyroid function tests, B-natriuretic peptide and cardiac enzymes were all normal. ECG showed an old left anterior fascicular block. A chest radiograph revealed an elevated right diaphragm (Fig. 1). An echocardiogram noted a normal ejection fraction with mild left ventricular hypertrophy without any chamber compression. A myocardial perfusion scan documented a normal ejection

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Figure 1. Chest radiograph showing elevated right hemidiaphragm (arrow).

fraction without evidence of ischemia. Computed tomography of the chest revealed an elevated right hemidiaphragm compressing the right atrium (Fig. 2). Bronchoscopy was normal, excluding any pulling effect on the diaphragm by an intrapulmonary pathology. Pulmonary function testing revealed a moderate restrictive lung disease with normal diffusion capacity. A fluoroscopic sniff test showed a paradoxical motion of the right diaphragm confirming the diagnosis of unilateral diaphragmatic paralysis which was attributed to phrenic neuropathy caused by diabetes. A surgical diaphragmatic plication was recommended due to the severity of his symptoms. Since the patient wanted to try medical management, continuous positive airway pressure

Figure 2. Coronal view of a computed tomography scan of the chest confirming the elevated diaphragm compressing the right atrium (arrow).

(CPAP) was offered; however, he failed the trial. Subsequently, the patient underwent surgical repair with complete resolution of all symptoms and remarkable functional recovery.

DISCUSSION

The descriptive term “trepopnea” has an interesting history. In 1937, Wood et al. observed that some patients with CHF preferred to sleep in the right lateral decubitus position^{1,2}. They attributed this preference to different reasons: due to choice or awareness of heart beating in a certain position; tolerable shortness of breath in one lateral position labeled as trepopnea of choice; and complete intolerance of one lateral position known as trepopnea of necessity^{1,2,7}. Recently, there has been a rekindling of interest in this clinical symptom in patients with CHF; the proposed mechanism is mainly due to an elevated sympathetic input and pulmonary venous pressure in the decubitus position^{7,8}. In our patient with a right unilateral diaphragmatic paralysis, trepopnea occurred in the left decubitus position perhaps as a result of the compression of the volume in the normal left lung via the displacement of the mediastinum towards the left side, thus further reducing the overall capacity for gas exchange.

Unilateral diaphragmatic paralysis is uncommon, and its incidence is not well known with only hospital based studies and case reports found in the literature. The presentation, in contrast to bilateral diaphragmatic paralysis, is subtle. It may manifest with dyspnea on exertion, but more often, is incidentally found on chest radiography. Injury during cardiac surgery is the most commonly reported cause, although surgery involving spine, shoulder, head and neck can also be causative⁹. Other etiologies include tumors of the neck and mediastinum, viral infections, brachial plexus neuropathy, vasculitis, neuropathies, spinal cord injuries; in a significant number of cases, no cause is found¹⁰. Diabetic phrenic neuropathy was the only identifiable etiology in our patient.

Although rare, diabetes can affect phrenic nerves causing unilateral or bilateral diaphragmatic paralysis with subsequent respiratory compromise^{11–13}. In one study, prolonged phrenic nerve latencies were found in 23% of diabetic patients with exertional dyspnea¹⁴. There is no correlation between the presence of phrenic and peripheral neuropathy or the degree of glycemic control^{15,16}, although studies in diabetic mice have shown that phrenic neuropathy can be induced by elevated blood sugar and may improve with insulin¹⁷.

Presentation and Workup

In a patient presenting with dyspnea, inquiring about the symptom of trepopnea would be prudent. This part of history is even more crucial in the setting of an elevated diaphragm. Paradoxical respiratory breathing suggested by unilateral subcostal and intercostal retraction during inspiration, as seen in our patient, can be an important clue. Although of limited utility in obese patients, a bedside assessment by percussion for diaphragmatic excursion can serve as a valuable tool to assess diaphragmatic dysfunction. Once history and chest imaging, with subsequent verification by computed tomography, confirm elevated diaphragm, a fluoroscopic sniff test can then show paradoxical cranial movement of the diaphragm during rapid inspiration. After that, spirometry

Fig. 3. Suggested algorithm for evaluating unilateral diaphragm paralysis.

should be performed¹⁰. In unilateral diaphragmatic paralysis, the test will reveal a decrease of $\geq 10\%$ of the vital capacity on lying supine, which is greater than the normal variation of 3–5%¹⁸. Spirometry not only helps with the diagnosis but also provides objective data to follow the progress of the patients. Though mostly useful in this regard, sometimes clinical symptoms do not correlate well with spirometric data¹⁹. Electromyogram (EMG) may also be used for diagnosis and help differentiate between myopathic and neuropathic causes when there is diagnostic uncertainty; however, the test has its limitations. Owing to technical issues with the surface and needle electrodes, the most reliable method is based on esophageal catheters with the inherent risks of an invasive procedure²⁰.

Recently, ultrasonography and plain radiographic characteristics have also been used to detect unilateral diaphragmatic paralysis and differentiate it from eventration^{10,11,21}. A suggested workup is recommended in Figure 3; however, there is variation based on institutional practices and experiences.

Treatment and Prognosis

Asymptomatic unilateral diaphragmatic paralysis does not require treatment. Spontaneous recovery is minimal after two years²². When there is functional limitation, surgical plication remains the gold standard⁹. Diaphragm resections and video assisted minimally invasive surgery have also been effective^{9,23}. In one series of 41 patients, 30 had the video assisted procedure and the rest had thoracotomy. Mean stay for the entire group was 3 days, and complications included two patients with pneumonia, two with atrial fibrillation, and one had paralytic ileus, and one with a deep venous thrombosis. The study had a mean long term follow up of 57 months, and all patients except for four showed clinical improvement. The non-responders were obese or had had the paralysis for more than 4 years²³. Another study of 22 patients with surgical plication also showed an excellent response at a mean follow up of 4.9 years. Mean hospital stay was

5.5 days, and five patients developed paralytic ileus after surgery. There were three deaths due to an acute coronary events, pulmonary embolism, and right heart failure. These were likely not related to the surgical intervention itself²⁴. The long-term prognosis of unilateral diaphragmatic paralysis is excellent except in patients with an underlying lung disease. As for the medical treatment, one case report described successful use of topiramate as treatment of diabetic phrenic nerve palsy¹³. CPAP may be used to prevent ventilatory failure in patients with bilateral paralysis, in pediatric patients and as a bridge for patients waiting for surgical correction^{24,25}.

CONCLUSION

Trepopnea is an important clinical symptom. Dyspnea can be a common complaint in diabetic patients particularly in the context of coexisting coronary artery disease, congestive heart failure and/or obstructive sleep apnea, to name a few. However, when no etiology is identifiable for the complaint, eliciting a history of trepopnea in patients presenting with dyspnea is valuable. This can be particularly true in a diabetic patient who has decreased diaphragmatic excursion on physical examination and an elevated diaphragm on imaging. In the correct clinical context, a workup for diaphragmatic paralysis with fluoroscopic sniff test, and spirometry should be pursued. Once confirmed, patient should be referred for surgical opinion based on the duration and intensity of symptoms.

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