CLINICAL VIGNETTE

A Case of Bilateral Diaphragmatic Paralysis

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Bilateral diaphragmatic paralysis is an uncommon and underdiagnosed condition. It should be considered in the differential diagnosis for unexplained dyspnea and hypoxemia. We present a patient with slowly progressive dyspnea and respiratory failure requiring mechanical ventilation without predisposing risk factors. The diagnosis of idiopathic bilateral diaphragmatic paralysis was made based on chest x-ray and diaphragmatic ultrasound in the absence of other etiologies.

Case Report

A 78-year-old male with hypertension, hyperlipidemia, and paroxysmal atrial tachycardia presented with progressive shortness of breath and chest tightness for six months. Symptoms initially started two years prior with paroxysmal nocturnal dyspnea, which progressed to the point where he had to sleep sitting upright. He remembers being told that his diaphragm was not working well but could not recall any other details. Further questioning revealed nocturnal awakenings with severe morning headaches and daytime somnolence.

Physical examination was remarkable for paradoxical abdominal movement during inspiration. Chest x-ray revealed bilateral hemi-diaphragmatic elevation, and ABG showed a pH of 7.38, pO2 of 30, and pCO2 of 70. Complete blood count, thyroid function tests, comprehensive metabolic panel, and rheumatologic testing were within normal limits as were nerve conduction studies, serial spinal MRIs, and spinal fluid analysis. Polysomnography revealed moderate obstructive sleep apnea, hypoventilation, and hypoxemia. Pulmonary function tests showed severe restrictive impairment with a marked decrease in the diffusing capacity. Diaphragmatic ultrasound demonstrated bilateral diaphragmatic paralysis. A diaphragmatic electromyography could not be obtained.

There was no preceding trauma or surgeries nor underlying heart or lung disease, mediastinal masses, myopathies, or neuropathies. He was diagnosed with idiopathic bilateral phrenic nerve paralysis and placed on supplemental oxygen at 3 L/min with nasal CPAP at night.

Six months later, the patient presented in acute respiratory distress requiring intubation with mechanical ventilation. He was unable to be successfully weaned necessitating tracheostomy. The patient was able to wean while upright, so the ventilator was removed during the day but resumed at night while recumbent. The patient was offered evaluation for diaphragm pacing, which he refused. He also declined Bipap and continues with this current regimen.

Discussion

The diaphragm is the chief muscle of inspiration separating the thorax from the abdomen. During inspiration, the diaphragm moves inferiorly to increase the volume of the thorax, so that the lungs can expand with air. The contraction of the diaphragm pushes the abdomen and rib cage outwards.[1,2]

One of the key signs of diaphragmatic dysfunction on physical examination is abdominal paradox. Instead of moving outwards during inspiration, the abdomen moves inward. The accessory muscles lift the rib cage outward to lower intrathoracic pressure, forcing the paralyzed diaphragm to move superiorly, which is best observed with the patient supine. Tachypnea, accessory muscle use, and decreased diaphragmatic excursion can also be seen.[3]

Diaphragm dysfunction can either be unilateral or bilateral. Unilateral paralysis is more common with less severe symptoms. They can present with mild dyspnea on exertion, dyspnea when supine, and/or decreased exercise tolerance.[1,3] In contrast, bilateral diaphragmatic paralysis presents with unexplained dyspnea at rest, when supine, and/or exertion. They can have significant orthopnea and recurrent respiratory failure. Patients can experience nocturnal hypoxemia resulting in hypersomnolence, morning headaches, fatigue, and confusion.[3]

Most of the causes of diaphragm paralysis can be categorized into the following five areas:

1. Traumatic: cardiac, pulmonary, esophageal, or mediastinal surgeries;
2. Compression: cervical osteoarthritis or tumors;
3. Inflammatory: vasculitis, pneumonia, or pleurisy;
4. Neuropathic: polio, myositis, multiple sclerosis, diabetic neuropathy, etc; or
5. Idiopathic.[1,3]

Diaphragmatic paralysis can be confirmed by several tests. Chest x-ray or chest CT scan will show diaphragm elevation on the affected side(s).[1,3] While being 90% sensitive for paralysis, it is only 44% specific. A sniff test is more useful for unilateral paralysis.[3] During the sniff test, the patient is

asked to breathe in and out through their nostrils while the diaphragms are monitored with fluoroscopy. In unilateral paralysis, the diaphragm either moves upward or does not move at all. However, the test is not as accurate for bilateral diaphragm paralysis.[1,3] Diaphragm ultrasound can be used to measure changes in the diaphragm thickness during inspiration. A lack of thickening, usually less than 2 mm, is diagnostic of diaphragmatic paralysis.[3] Ultrasound can monitor recovery of the paralyzed diaphragm, if reversible.[1] Pulmonary function tests (PFTs) are important in the diagnosis of diaphragmatic dysfunction.[4] Bilateral diaphragm paralysis produces a moderate to severe restriction with 30-50% of the predicted value for total lung capacity and worsens when the patient is supine. With unilateral paralysis, there is only a 10-30% decrease in the vital capacity.[3] Transdiaphragmatic pressure (Pdi) is the standard for diagnosis of bilateral diaphragmatic paralysis; however, it is invasive. This test requires transnasal placement of balloon catheters in the lower esophagus and stomach to measure the difference between the pressures. Pdi less than 80 cm of water in men and less than 70 cm in women is diagnostic of diaphragmatic paralysis.[3,4] Diaphragm electromyography stimulates the phrenic nerve to determine if neuropathy or myopathy is responsible for diaphragm dysfunction.[5]

Once the diagnosis of unilateral or bilateral diaphragm dysfunction is made, treatment options are considered. Treatment will depend on whether an underlying, correctable cause has been identified. Most times, unilateral diaphragm paralysis does not need treatment and can be monitored.[1] If patients have a pCO2 greater than 45 during the day, oxygen saturation less than or equal to 88% for 5 consecutive minutes at night, or progressive neuromuscular disease with a maximal static inspiratory pressure less than 60 cm of water or a forced vital capacity less than 50% of the predicted value, then they should be started on non-invasive nocturnal ventilation.[3] After 6-12 months without signs of recovery, diaphragm plication can be attempted. The procedure flattens the diaphragm to allow the lung to expand.[1,3] This may lead to up to 20% improvement in measured pulmonary function by lessening the paradoxical movement.[3] This is unlikely to be helpful in bilateral diaphragmatic paralysis. However, diaphragmatic pacing may be an option for these patients. It requires intact phrenic nerves to provide full ventilatory support for ventilator-dependent patients.[1,2]

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