CASE REPORT

A PUZZLING CASE OF SEVERE PULMONARY HYPERTENSION - A CASE REPORT

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Pulmonary hypertension (PH) is a life-threatening cardiopulmonary disorder marked by elevated pressure in the pulmonary arteries. We report a 51-year-old male, post-pericardectomy, presenting with breathlessness. Diagnosis revealed pulmonary hypertension with depressed right ventricle function. Managed with non-invasive ventilation (NIV) and oxygen, he was slated for right heart angiography but couldn't lie flat. Fluoroscopy identified bilateral diaphragm paralysis. Due to a lack of diaphragm-related procedures, he continued non-invasive ventilation, oxygen, and pulmonary vasodilators. With these measures, the functional status of the patient improved and he was able to carry out his routine activities.

Keyword: Pulmonary hypertension; Diaphragm paralysis; Non-invasive ventilation; Pericardectomy; Constrictive pericarditis; Tuberculosis

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INTRODUCTION

The diaphragm is the chief muscle of inspiration. Diaphragmatic paralysis, whether unilateral or bilateral, can cause paradoxical motion of the lower rib cage with inspiration, leading to severe respiratory impairment causing dyspnoea, paroxysmal nocturnal dyspnoea, hypoventilation, hypoxia, hypoxemia, and respiratory failure.[1,2] These circumstances, ignored for years, led to severe pulmonary hypertension. The diagnosis is difficult to reach due to the rarity of this condition and the low index of suspicion.[1–3]

We report a case of pulmonary hypertension caused by idiopathic bilateral diaphragm dysfunction, presented to the pulmonary hypertension clinic as a diagnostic challenge, emphasizing the significance of early detection and intervention to prevent the progression of pulmonary hypertension.

CASE REPORT

A 51-year-old man, with history of pericardectomy 20 years ago due to constrictive pericarditis secondary to tuberculosis, presented in the emergency with complains of severe dyspnoea (New York Heart Association Class IV) and orthopnoea. He had previously visited multiple physicians and specialists for these symptoms but had not found relief. On examination, he had oedema in both lower extremities and was unable to lie down flat due to breathlessness. His blood pressure was 110/60mmHg, and his heart rate was 60/min. His oxygen saturation (SpO2) was 89% on room air while sitting, and decreased to 70% while walking.

Initial lab investigations showed a haemoglobin and haematocrit of 15.5 gm/dl and 47.7, with a normal value of creatinine and electrolytes. A chest X-ray showed vascular congestion and both hemi-diaphragms were at the same level (Figure 1-A). Echocardiography revealed severe pulmonary artery hypertension (PASP=110 mmHg) with moderate tricuspid regurgitation, dilated right chambers, and D-shaped left ventricle with normal left ventricular function (Figure 1 –B and C).

A CT scan of his chest showed dilated pulmonary arteries (MPA=41 mm, RPA=25 mm and LPA=25) without any evidence of pulmonary embolism. An Anti-nuclear Antibody (ANA) profile was negative. A pulmonary function test revealed a restrictive pathology. The patient was started on oxygen therapy and pulmonary vasodilators. Right heart catheterization could not be done due to the patient's inability to lie down flat. Fluoroscopy confirmed bilateral diminished diaphragm movement with respiration. To address the patient's condition, non-invasive ventilation (NIV) was initiated at home, along with oxygen inhalation therapy, especially during sleep. The patient's condition improved after a few days, and he was able to return to work with the use of oxygen therapy during exertion. Ultimately, the patient was diagnosed with idiopathic bilateral diaphragm dysfunction, which led to pulmonary hypertension and right heart dysfunction.

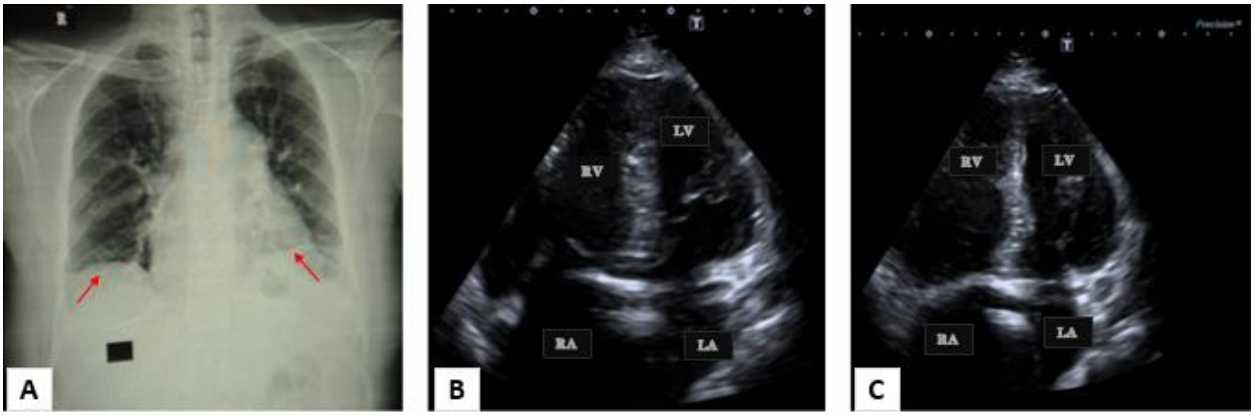

Figure 1: (A): The chest x-ray showed signs of vascular congestion and both hemi-diaphragms were aligned at the same level (indicated by red arrows). (B and C): Echocardiography revealed severe pulmonary artery hypertension with elevated pulmonary artery systolic pressure (PASP-110 mmHg), along with moderate tricuspid regurgitation. The right-sided cardiac chambers appeared dilated, while the left ventricle showed a "D-shaped" configuration, indicating increased pressure in the right ventricle.

DISCUSSION

Diaphragm weakness is an uncommon cause of orthopnoea that can lead to pulmonary hypertension and right heart failure. Patients who present with dyspnoea, orthopnoea, and paradoxical thoracoabdominal breathing when lying down should be investigated for the possibility of diaphragm paralysis.[4,5]

The causes of diaphragm paralysis are diverse and include traumatic, compression-related, inflammatory, neuropathic, and idiopathic factors. Trauma to diaphragm can be a result of surgeries like open-heart procedures, compression can be due to cervical spine manipulation or tumour.[6,7] Inflammatory processes, neuropathic conditions such as diabetic neuropathy and multiple sclerosis, infections, and systemic conditions can also contribute to diaphragmatic paralysis. In some cases, the cause of diaphragm paralysis is unknown (idiopathic).[2,8]

The pathophysiology that causes pulmonary hypertension in patients with diaphragm weakness is intricate. Diaphragm weakness results in a lower total lung capacity leading to alveolar hypoventilation. This condition worsens when lying down and improves to some extent when upright. Alveolar hypoventilation results in chronic hypoxemia, which, if left untreated, can lead to the development of pulmonary hypertension and eventually right heart failure.[9,10]
Diagnosis of diaphragm paralysis requires a multifaceted approach, including a clinical examination, radiological tests, and non-invasive diagnostic tools like the sniff test, thoracic ultrasound, and magnetic resonance navigator echo. Pulmonary function tests may show a restrictive pattern, with a reduced forced expiratory volume in 1 second (FEV1) and vital capacity.[2,10] Confirmation through phrenic nerve stimulation and diaphragmatic electromyography is pivotal.[2]

The mainstay of treatment of pulmonary hypertension is the treatment of the underlying cause. For the management of unilateral diaphragm paralysis, a conservative approach is usually recommended in asymptomatic patients. The role of surgery is controversial, though transthoracic diaphragmatic plication may provide relief for respiratory insufficiency in cases of irreversible diaphragmatic paralysis. Minimally invasive approaches like video-assisted thoracoscopic diaphragm plication have significantly improved functional status and Spirometry. Diaphragmatic pacing and phrenic nerve reconstruction presents an alternative, but are costly to perform, not without limitations, and needs surgical expertise to perform.[7,11,12]

Non-invasive ventilation options including nasal continuous positive airway pressure/bilateral positive airway pressure or intermittent positive pressure ventilation improve total lung capacity and are recommended for bilateral diaphragmatic paralysis along with ventilation and oxygenation leading to improvement in the overall condition.[9,13]

Nasal continuous positive airway pressure/bilateral positive airway pressure or intermittent positive pressure ventilation is recommended for bilateral diaphragmatic paralysis. Severe cases may require tracheostomy and mechanical ventilation. Treatment choice depends on individual patient characteristics, underlying causes, and the severity of respiratory compromise.[9,13]

Unilateral diaphragmatic paralysis has a good prognosis, and compensatory mechanisms can help with adequate ventilation, but the quality of life of

patients can be significantly affected due to paradoxical respiratory movements.[2,4] Bilateral diaphragmatic paralysis prognosis varies depending on the underlying cause, with patients with myopathies or chronic demyelinating conditions having a poor prognosis. Individuals with a combination of diaphragm paralysis and underlying respiratory diseases often require mechanical ventilatory support, indicating a less favourable outcome.[14]

In our patient diaphragm weakness leading to a state of long-standing alveolar hypoventilation was considered the main cause for pulmonary hypertension. No obvious cause was found for his diaphragmatic weakness. A similar case was reported by Meysman *et al*, in which diaphragm weakness was also identified as the primary factor in the development of pulmonary hypertension.[5]

This case underscores the critical importance of early recognition of diaphragm weakness to prevent the development of pulmonary hypertension. The multifaceted approach to diagnosis and management, including non-invasive ventilation, contributes to improved outcomes in patients with idiopathic bilateral diaphragm dysfunction

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Limitations: One study limitation was that we could not perform phrenic nerve stimulation and diaphragmatic electromyography in our patient. This limitation is significant as it could have provided further insights into the aetiology and extent of diaphragm paralysis.

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