Symptomatic Right Diaphragm Hemiparesis Causing Severe Compression of the Right Atrium and Right Ventricle

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Introduction
Although a number of muscles support the mechanics of respiration, the diaphragm is the most important muscle driving respiratory forces in the thoracic cavity and enabling proper ventilation and oxygenation. Originating from the C3-C5 cervical nerve roots, the phrenic nerve extends into the thoracic cavity to provide sensory and motor innervation to the right and left hemidiaphragm. Traumatic injury to the nerve is the most common cause of diaphragmatic weakness. Due to the long course of the left phrenic nerve in the thorax, left-sided diaphragmatic weakness is more common compared to the right. As a result, unilateral diaphragmatic paralysis is more common than bilateral disease and is often an asymptomatic incidental finding on imaging. However, clinical manifestations such as dyspnea on exertion, decreased exercise performance, and sleep disturbances are common. Rarely, increased thoracic pressure from diaphragm elevation can cause cardiac compression. We present a case of significant right diaphragmatic hemiparesis causing severe compression of the right atrium and partial compression of the right ventricle.

Case
A 43-year-old female presented to our clinic complaining of shortness of breath that began suddenly four months prior. Past medical history is significant only for seasonal allergies and prior smoking for 12 pack-years, quitting 13 years ago. Initially the shortness of breath was associated with a sharp, pleuritic chest pain. She was seen by her primary care physician and was prescribed steroids and antibiotics, which improved but did not resolve the chest discomfort. Dyspnea with exertion remained persistent. Her BMI was 38 with a blood pressure of 132/99 mmHg, heart rate 107 bpm, and oxygen saturation 98% on room air. Chest x-ray revealed right hemidiaphragm elevation, which was also seen on chest CTA. Fluoroscopic sniff test confirmed elevated right hemidiaphragm with slight paradoxical movement of approximately one rib length, while the left diaphragm moved freely. Pulmonary function testing revealed restrictive lung physiology. Transthoracic echocardiogram revealed preserved LVEF, RV dilatation, paradoxical intraventricular septal motion consistent with RV volume overload, mildly reduced RVSF due to severe extrinsic compression of the right atrium in both systole and diastole by the diaphragm and liver. There was no evidence of pericardial effusion or solid mass lesion. Inflammatory workup including CCP, ESR, and CRP were normal.

Due to the severity and persistence of symptoms, she was ultimately referred for surgical correction.

Results
AP chest x-ray

TTE apical 4-chamber view

Discussion
Hemidiaphragm paralysis occurs due to direct trauma to the phrenic nerve. Surgical trauma, most commonly due to cardiac bypass and cold cardioplegia, cryoablation for atrial fibrillation, or intra-abdominal surgery, such as liver transplantation due to inferior vena cava clamping, occurs at an incidence of 10-30%. However, our patient had no prior history of intrahepatic or abdominal surgery. Other common causes due to congenital disorder, viral infection, nerve disorder, or compression due to malignancy.

There are only a few reported cases of cardiac sequelae resulting from elevation and dysfunction of the diaphragm. These reports include of asymptomatic right atrial compression seen incidentally on echocardiogram, one reported from evagination of the diaphragm after liver transplant, and another from central evagination causing left ventricular compression and arrhythmias. One case was reported as an incidental finding of bilateral diaphragmatic dysfunction on imaging after a cardiac workup, and three cases of polycystic kidneys with liver cysts causing diaphragm elevation and cardiac compression.

Generally, these patients remain asymptomatic and findings are often incidentally found on imaging studies such as x-ray, CT, MRI, or echocardiogram. As noted by previous case reports, patients with cardiac and pulmonary compression will often present with exertional dyspnea and fatigue. Pulmonary function tests will show restrictive physiology. Fluoroscopic sniff test remains the most reliable test to detect diaphragm paralysis. Treatment is based on severity of the obstructive process and symptoms. Diaphragmatic plication decreases intrathoracic pressure, compression force on the heart and lung, as well as allowing for increased lung function and capacity.

References