P-93

Idiopathic bilateral phrenic neuropathy : A Case Report

Sang Woo Kim, MD, Yu Jin Im, MD, Duk Hyun Sung, MD, PhD

Department of Physical Medicine and Rehabilitation, Sungkyunkwan University School of Medicine, Samsung Medical Center, Seoul, Korea

Introduction

- Diaphragm is dome shaped structure innervated by the phrenic nerve.
- Patients with unilateral diaphragm weakness are usually asymptomatic but patients with bilateral diaphragm weakness are more likely to have orthopnea and dyspnea with exertion.
- There are various causes of diaphragmatic dysfunction including multiple sclerosis, Guillain-Barre syndrome, myasthenia gravis and muscular dystrophy.
- Here, we report on a patient with bilateral phrenic neuropathy who presented with clinical manifestation of the bilateral idiopathic neuralgic amyotrophy of phrenic nerve.

Case report

- A 68-year-old female patient presented to the Emergency Medicine with persistent orthopnea.
- Six months prior to her first visit to our hospital, she experienced a clear onset of orthopnea and had visited another emergency medicine department at that time.
- A Chest computed tomography (CT) revealed evidence of pulmonary thromboembolism, and she was prescribed anticoagulants.
- However, the patient's dyspnea worsened during sleep despite the prescribed anticoagulants and she was subsequently admitted to the Pulmonology department for further management.

- A follow-up Chest CT performed at our hospital did not reveal any evidence of pulmonary embolism.

- The possibility of the patient's orthopnea having a pulmonary or cardiac origin was considered low, and therefore, a consultation with the Department of Physical Medicine and Rehabilitation (PMR) was requested for further management.
- On physical examination, bilateral distal upper arm weakness was noted, along with both elbow flexion contractures of 45 degrees(Table 1).
- While in a supine position, she reported dyspnea within 30 seconds. Accessory muscle contraction and abdominal wall retraction were observed during inspiration, indicating paradoxical breathing.

- Compared to the sitting position, forced vital capacity (FVC) was reduced by over 75% in the supine position(Table 2).
- EMG/nerve conduction study (NCS) revealed bilateral ulnar entrapment neuropathy at the elbow level, but there was no widespread denervation to suggest motor neuron disease.
- Needle electromyography (EMG) and ultrasound evaluation of the diaphragm were difficult to perform due to orthopnea and obesity.

- Given the sudden onset of orthopnea and the absence of physical findings suggestive of cervical myelopathy, the patient was diagnosed with idiopathic neuralgic amyotrophy of bilateral phrenic nerves.

- The patient was provided with non-invasive ventilation via Bilevel positive airway pressure (BiPAP) and was advised to continue using it upon discharge.
- During a follow-up visit after nine months from the onset of orthopnea, the patient reported improvements in orthopnea and was no longer using BiPAP.

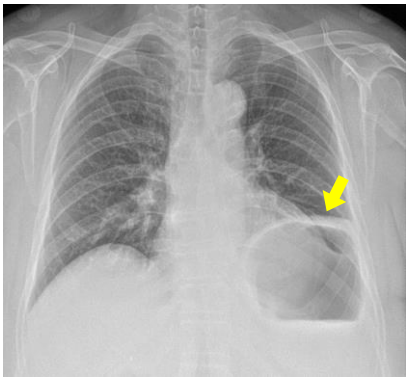

Figure1. Left diaphragm elevation (arrow) shown in chest X-ray

Table 1. The results of neurological examination and electrophysiologic study

	Category	Result	
N/Ex	Motor strength (MRC grade)	Anterior neck flexor 5 Shoulder flexor 5/5 Elbow flexor 5/5 Wrist extensor 5/5 Finger flexor 5/5 Finger abductor 2+/2+ Extensor indicis proprius 4+/4+ Extensor digitorum communis 4+/4 Abductor pollicis brevis 5/5-	
	Sensory	Intact on both side	
	Muscle atrophy	Both FDI atrophy(Rt>Lt)	
	DTR	Biceps jerk 2/2 Triceps jerk 2/2	
EP study	SNAP	No SNAP in both ulnar nerve	
	CMAP	No CMAP in Lt. ulnar nerve Reduced CMAP amplitude in Rt. ulnar nerve	
	Needle EMG	Spontaneous activity	Denervation potential in Rt. FDI, Rt. Mid lumbar PSP, Lt. lower lumbar PSP
		MUAP	Neuropathic potential in Rt. FDI

Abbreviations: MRC; N/Ex; Neurologic examination, EP; Electrophysiologic, Medical Research Council, DTR; Deep Tendon Reflex, NCS; Nerve Conduction Study, EMG; Electromyography, CMAP; Compound Muscle Action Potential, MUAP; Motor Unit Action Potential, FDI; first dorsal interossei, PSP; paraspinalis

Table 2. The results of PFT, MIP/MEP, ABGA

Test	Position	Results
PFT	Sitting	FEV1 1.56L, FVC 1.74L
	Supine	FEV1 0.26L, FVC 0.32L
ABGA	Sitting	pH 7.38, pO2 84.0mmHg, pCO2 45.0mmHg
	Supine	pH 7.388, pO2 48.1mmHg, pCO2 45.4mmHg
MIP/MEP	Sitting	MIP 31cmH2O / MEP 92cmH2O

Abbreviation : PFT; Pulmonary function test, ABGA; Arterial blood gas analysis, MIP; maximal inspiratory pressure, MEP; maximal expiratory pressure, ABGA; arterial blood gas analysis, FEV; Forced expiratory volume, FVC; Forced vital capacity

Conclusion

- The key points in distinguishing bilateral diaphragmatic paralysis from unilateral diaphragmatic paralysis are orthopnea and paradoxical breathing on physical examination.
- To make a diagnosis, PFT in the sitting and supine positions, as well as USG of diaphragm thickness during expiration and inspiration, can be performed.
- We observed, as in previous studies, that improvement of symptoms occurs within 1-2 years in cases where idiopathic neuralgic amyotrophy affects the phrenic nerve.