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Special Edition
"Revisiting Physical Diagnosis in Respiratory Medicine"

Case Report

Diaphragmatic Dysfunction without Paradoxical Breathing: A Case of Nemaline Myopathy

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KEYWORDS: Ultrasonography; Diaphragmatic dysfunction; Paradoxical respiration.

A 35-year-old woman was admitted to this hospital because of worsening shortness of breath and productive cough for 3 days. Her past medical history was remarkable for Namaland sympathy, which was diagnosed at the age of 4 by muscle biopsy as she developed muscle weakness with Gower's sign. For 3 years prior to this admission, she had been suffering from recurrent pneumonia, likely due to aspiration due to impaired bulbar functions along with her muscle weakness. Obstructive Sleep Apnea (OSA) was suspected as she complained of morning headache, daytime fatigue and night sweats and the diagnosis was confirmed by overnight polysomnography with severe obstructive sleep apnea (OSA) with AHI of 46.2 events per hour 2 years ago, which had been treated by Bi-level Positive Airway Pressure (BIPAP). Echocardiography was normal except for mild elevation in pulmonary arterial mean pressure.

On examination, she was in respiratory distress. The temperature was 37 ºC, the blood pressure 112/64 mmHg, the pulse 107 beats per minute, the respiratory rate 22 breaths per minute, and the oxygen saturation 93% while the patient was breathing with supplemental oxygen of 1.5 L per minute through nasal cannula. Auscultation of the chest revealed bilateral holoinspiratory crackles. Computed Tomography of the chest revealed consolidation in right middle and left lower lobe, concerning for aspiration pneumonia, particularly by bacterial origin.

On one night, her oxygen saturation dropped down to 64% and she became lethargic. Her arterial blood gas revealed elevation of partial pressure of carbon dioxide ($PaCO_2$) of 108 mmHg. This acute hyper apnea was thought to be due to hypoventilation due to unfitted mask. As the mask was re-adjusted, and FiO_2 was lowered to keep her SpO_2 above 85%, her mental status and $PaCO_2$ level improved. Her overall condition including breathing dramatically improved as we performed respiratory physiotherapy along with frequent sputum suctioning. Given this, we assumed her pneumonia is exacerbated by hypoventilation and by inability to cough up sputum due to respiratory muscle weakness, including diaphragm dysfunction. Diaphragm ultrasonography revealed thin diaphragm thickness (tdi) along with minimal change of thickness of diaphragm (Δ tdi%: Δ tdi / tdi end-expiration) during inspiration and expiration to support our hypothesis (tdi on end-inspiration: 1.76 mm (mean), tdi on end-expiration: 1.73 mm, Δ tdi%:1.7 %) (Figure 1A, 1B, Video 1). Despite the presence of diaphragm dysfunction according to the definition by Maccool[1] and Minami,[2] movement of rib cage and abdomen were normal (Video 2). Ryan et al reported that patients with typical congenital hemline sympathy often present with respiratory distress by diaphragm dysfunction with slow progress, as seen in our case.[3] This case is unique as she did not develop paradoxical breathing, a common finding for bilateral diaphragm dysfunction. This finding supports Tobin's finding that paradoxical breathing is triggered by respiratory high load, rather from respiratory muscle weakness.[4,5]

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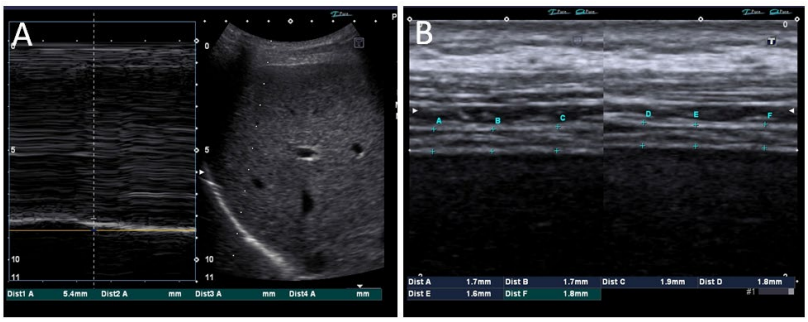
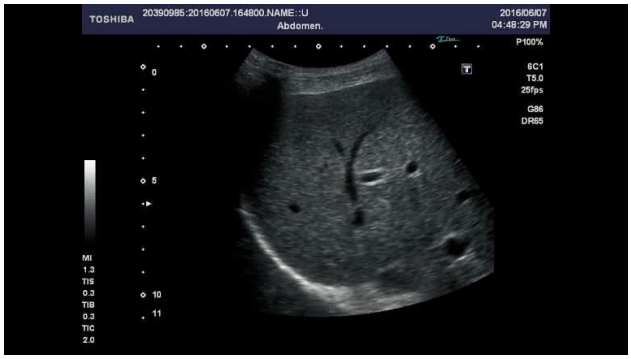


Figure 1: Ultrasonography of the diaphragmusing M-mode (A) showed the motion of the diphragmatic dome seemed to be quite limited, suggesting diaphragmatic dysfunction. Thickening of the diaphragm was almost equal at end-inspiration (B, left column, 1.76 mm) and end-expiratoin (B, right column, 1.73 mm).

Video 1: Diaphragm ultrasonography during inspiration and expiration.



Video 2: Movement of rib cage and abdomen.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

CONSENT

The authors obtained written informed consent from the patient for submission of this manuscript for publication.

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