中文題目：以急性呼吸衰竭同時伴隨巨大縱膈腔胸腺瘤及章魚壺心肌症為初始臨床表現之重症肌無力症病例: 一個案報告及文獻回顧

英文題目：Myasthenia gravis with initial presentation of acute respiratory failure concomitant with huge mediastinal thymoma and takotsubo syndrome: a case report and literature review

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Introduction: Myasthenia gravis (MG) is the most common disorder of neuromuscular transmission. In generalized disease, the weakness may affect ocular muscles, bulbar, limb, and respiratory muscles. MG crisis is a life-threatening exacerbation of myasthenia gravis that is defined as worsening of myasthenic weakness requiring ventilation. About 50% patients with thymoma develop MG and cardiac events were reported in 16 % of patients with MG. We reported a patient presenting initially with acute respiratory failure, a huge mediastinal mass, and takotsubo syndrome. MG was diagnosed and managed soon later.

Case presentation: A 49-year-old female without underlying disease presented to our emergency department with dyspnea for 1 day. She was intubated with ventilator under the diagnosis of acute respiratory failure. A huge mediastinal mass was found in chest X-ray. Computed tomography scan of the thorax showed a huge anterior mediastinal tumor with diameter about 10 centimeters and heterogeneous enhancement at right upper chest. On the second day after admission, she complained chest pain and dyspnea and elevated cardiac enzyme with ST-T change on electrocardiogram developed. Takotsubo cardiomyopathy was diagnosed after coronary angiography. At the same time, she had left ptosis, muscle weakness of four limbs especially in lower limbs, and easily falling down in past two years. In recent months before admission, she even couldn’ t walk normally. She didn’ t see a doctor due to fear of pandemic COVID-19. Neurologist was consulted and myasthenic crisis was highly suspected. After surgical removal of the huge right mediastinal tumor, she received 5-course plasmapheresis. Her muscle power recovered well after plasmapheresis. Ventilator weaning and extubation were successful 10 days after admission. Pathology of the excisional huge mediastinal tumor reported an encapsulated type AB thymoma, modified Masaoka stage I. Her serum anti-acetylcholine receptor antibody was positive and confirmed the diagnosis of MG. Her weakness had good response to treatment and rehabilitation was performed thereafter. She was discharged when she could walk shortly without assist.

Discussion: Myasthenia gravis (MG) crisis is a life-threatening exacerbation of myasthenia gravis that is worsening of myasthenic weakness requiring ventilation. Plasmapheresis is one of the main managements in myasthenic crisis. It’ s a short-term treatment for impending and manifest myasthenic crisis in those who with significant respiratory or bulbar dysfunction. A systematic review and meta-analysis showed that the clinical MG remission rate after plasmapheresis was significantly higher than that in the control group. The pathogenesis of takotsubo cardiomyopathy may be secondary to a primary metabolic disturbance in cardiomyocytes. The stress-induced catecholamine release may lead to increased calcium, which activates cAMP, causing damage to the cardiac cells especially affecting the vulnerable apical region. Previous study showed that particularly stress from myasthenic crisis may

trigger Takotsubo cardiomyopathy and myasthenic crisis-triggered takotsubo syndrome particularly affects female patients. Thymomas can be separated into several types including simply A, B (B1, B2, B3) and mixed AB type according to the WHO classification. Management for thymoma is multiple strategy for different stages as diagnosed, complete surgical resection is recommended for Masaoka stages I and II by computed tomography imaging. Based on previous studies, thymoma-associated MG patients have higher probability to develop generalized symptoms and higher incidence of myasthenic crisis or treatment refractoriness. Randomized clinical trials provided improving clinical outcomes and reducing the need for immunosuppressive therapy in myasthenia gravis patients who received thymectomy.

Conclusion: We reported a case who had undiagnosed myasthenia gravis with initial presentation of myasthenic crisis and a huge mediastinal thymoma followed by takotsubo syndrome. In the era of pandemic COVID-19, diagnosis and management of some diseases may be changed and more difficult.