中文題目：一個患有喘、吞嚥困難、雙下肢無力的男性

英文題目：A man with dyspnea, dysphagia and bilateral lower limbs weakness

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Introduction:

Dyspnea, dysphagia and bilateral lower limbs weakness, each has its distinct set of differential diagnoses in their workup. However, when these chief complaints accumulate on one patient, they should hint us of diseases with systemic involvement in nature.

Case presentation:

A 46-year-old man was admitted to MICU due to aspiration pneumonia with respiratory failure needing NIPPV, and later, intubation with mechanical ventilation during hospitalization.

4 months ago, he started experiencing bilateral lower limbs weakness, lower back pain, exertional dyspnea and dysphagia especially on swallowing fluid. He visited cardiologist due to dyspnea and was to have fair cardiac function on heart sonography and ECG. His dyspnea was thought to be due to obesity and bilateral lower limbs weakness, thus he was suggested to visit a neurosurgeon. For his lower back pain with bilateral lower limbs weakness, MRI was done which showed L4-5 moderate degree thecal sac narrowing, L1-2, L2-3 and L3-4 mild thecal sac narrowing. Spinal surgery was arranged. Meanwhile, he also visited a pulmonologist where lung function test arranged showed restrictive breathing pattern. At the same time, he presented to an otolaryngologist, who later referred him to a gastroenterologist, both who performed evaluation on him but with no definitive diagnosis. He went to a neurologist for his dysphagia. The neurologist found no definite focal weakness, but symmetrically decreased deep tendon reflexes, and equivocal Babinski sign. Brain MRI showed no relevant lesion. Before receiving repetitive stimulation test and nerve conduction test arranged by the neurologist, he underwent L4/5/S1 laminectomy, discectomy with interbody fusion with cage and TPS fixation.

Unfortunately, the surgery did not improve his lower back pain nor his bilateral lower limbs weakness. His bilateral lower limbs weakness and dyspnea worsen after the surgery. 2 days before presenting to our emergency department, he experienced fever and dyspnea. He was diagnosed with aspiration pneumonia and was admitted to our MICU due to desaturation down to 70%. He was treated with Augmentin for his pneumonia and was placed on High

Frequency Chest Wall Oscillation (HFCWO) to facilitate sputum expectoration. His respiratory failure gradually worsen needing intubation and mechanical ventilation 1 week later. To survey his subacute generalized weakness, spinal cord MRI, CSF study, repetitive stimulating test and nerve conduction test done showed no definitive findings. Acetylcholine antibody later showed negative result. Myopathies antibodies done showed anti-Ro52 (2+) and anti-SRP (3+). Rheumatologist was consulted for further treatment.
On inspection, heliotrope eruption, shawl sign, periungual erythema, Holster sign, Mechanic's hand, were negative, equivocal Gottron's papules was found on bilateral knuckles area. Creatinine kinase was 1601 IU/L. EMG was not accessible during his stay in MICU. MRI of bilateral thighs showed diffusely edema and mild enhancement of bilateral internal, external obturator muscles, adductor group and left gluteal maximus muscles, compatible with inflammatory myopathies. Due to concurrent aspiration lobar pneumonia with respiratory failure, low dose methylprednisolone (40mg BID) was given, along with plasma exchange for 5 days. The patient showed daily improvement in muscle power of his 4 limbs since the first day of plasma exchange. 1 week later he was extubated with HFCWO support, but his nasogastric tube was kept in use. As his pneumonia improved, bedside rehabilitation was started and he was transferred to general ward on the 6^{th} week of hospitalization. After intense rehabilitation training for another 1.5 month, he was able to stand briefly with support. He was then discharged to nursing home to continue on rehabilitation training.

Discussion:

Polymyositis usually present with proximal motor weakness, most severely around the shoulder/pelvic girdles and neck flexors. The patient usually has elevated serum muscle enzymes, abnormal neuro-diagnostic studies especially EMG, positive myositis-associated autoantibodies. Muscle biopsy, if accessible, should be performed to confirm the diagnosis. Myositis-associated autoantibodies and muscle biopsy are both helpful in prognostication. Anti-SRP antibodies are associated with necrotizing autoimmune myopathy, which commonly present with rapid progression of proximal muscle weakness, lower-extremity weakness, myalgia, dysphagia, dyspnea, persistently elevated CK, resistance to glucocorticoid and overall, a poor prognostic factor. These features are all found in our case, complicating his recovery from bedridden status. Patients with polymyositis need corticosteroids and immunosuppressive agents early. Immunosuppressive agents include methotrexate, azathioprine, mycophenolate mofetil, cyclophosphamide, IVIG, plasma

exchange or rituximab. Our case has a painstaking experience in looking for a right diagnosis for his discomfort, which we, as medical service providers, could improve with a careful history taking and a general knowledge of inflammatory myopathies.

Conclusion:

This is a case of polymyositis initially presenting with dyspnea, dysphagia and bilateral lower limbs, whose condition rapidly worsened to the state of respiratory failure needing intubation and mechanical ventilation. A careful history taking and a general concept of polymyositis could have prompted further investigation or, at least, appropriate referral earlier, and could even save our patients from unnecessary surgeries.