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Neuromuscular Junction Disorder with Progressive Cerebellar Dysfunction in A Lung Cancer Patient

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Introduction

Paraneoplastic neurologic syndromes (PNS) can affect the central nervous system (eg, paraneoplastic cerebellar degeneration [PCD]) and the neuromuscular junction (eg, Lambert-Eaton myasthenia syndrome [LEMS] and myasthenia gravis [MG]). A small percentage of patients with small cell lung cancer (SCLC) have a PNS, of which the most frequent is LEMS. The symptoms of PNS could arise before the diagnosis of SCLC. And we need to pay attention to these patients. The screening recommendation for the patient with motor weakness due to neuromuscular junction disorder (LEMS type) would be repeated cancer screening tests including CT or positron emission tomography (PET) scans. Early diagnosis can advance initiation of anti-tumor therapy and thereby improve survival. We experienced a patient, who presented initially LEMS, MG and PCD, was finally diagnosed with PNS by SCLC.

CASE REPORT

A patient, 60-year-old man, admitted because of ataxia, dysarthria, ptosis, dizziness and weakness of proximal limbs with difficulties in walking over 14-days period. On physical examination, he had nystagmus with balance disturbance on standing, and tandem gait was impossible. Brain MRI showed no specific abnormal findings. In serologic study, AChR-Ab, anti-MuSK Ab and paraneoplastic antibodies were negative. Thyroid fuction test was normal. In motor nerve conduction study (NCS), the findings were delayed latencies, low amplitudes but conduction velocities were preserved. The sensory NCS was unrevealing. On electromyography, proximal and distal muscle of limbs showed motor units changes with short duration and variable amplitudes. Repeated electrical stimulation (RNS) showed decremental response in ADQ by 22% on low-frequency (3Hz) stimulation.(Fig 1a) However, in high rate stimulation (20Hz), there were incremental responses in APB by 40.8%. (Fig 1b). The initial chest CT showed a mass in anterior mediastinum and PET scan showed no remarkable lesion in thorax (Fig. 2a). He got thymectomy but cytology revealed thymic cyst only. He was administered oral prednisolone and pyridostigmine. However, the effect of medical treatment was not clear as well as additional IVIG therapy. Considering of progressive ataxia, dysarthria, and motor weakness, we strongly suspected the possibility of PCD with neuromuscular junction disorders caused by lung cancer. Follow up after 3 months chest CT and PET-CT showed lymphadenopathy in right upper paratracheal area and the biopsy confirmed metastatic SCLC (Fig. 2b). Bone scan shows no evidence of metastasis (Fig. 3).

Conclusion

We have experienced a case with progressive weakness, ataxia and dysarthria as initial symptoms of lung cancer without respiratory symptoms. These changes were diagnosed as a PNS (LEMS, MG and PCD). Early recognition of PNS and repeated screenings would improve the diagnosis and treatment of these cancers.

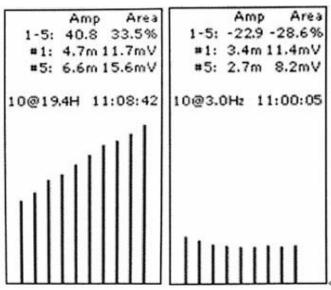


Fig. 1a : MG pattern, marked decrement of CMAP amplitude in ADM on 3Hz stimulation Fig. 1b : LEMS pattern, marked increment of CMAP amplitude after exercise in APB on 20Hz stimulation

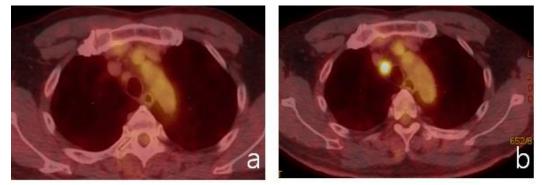


Fig. 2a: Initial PET-CT showed no abnormal lesion Fig. 2b: Follow up after 3 months PET-CT showed lymphadenopathy in right upper paratracheal area.



Fig. 3: Bone scan shows no evidence of metastasis