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A CASE OF LAMBERT-EATON MYASTHENIC SYNDROME WITH PROMINENT AUTONOMIC SYMPTOMS AFTER TRAVELLING TO CHINA

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Introduction

Lambert-Eaton myasthenic syndrome (LEMS) is a rare neuromuscular disorder characterized by fluctuating limb muscle weakness and various autonomic symptoms. Its incidence and prevalence are highly variable in different population. One explanation for this variability is underdiagnosis due to coexisting cancer symptoms.

Case Report : Here we report a 66-year-old male patient who developed severe autonomic symptoms such as postural hypotension (80/50), xerostomia and xerophthalmia after returning from a travel to China. Some episodes of dropped head were present at the beginning of the disease. A fluctuating weakness of the proximal limbs was also present but for almost 3 years postural hypotension and depression were considered the cause of the weakness and fatigability. The weakness was prevalent in the lower limbs and was highly variable from day to day. Ocular symptoms has never been present. He didn't complained any bulbar symptom but a mild dysarthria and an hypophonic voice after prolonged conversation. An immunological screening was negative and a Sjogren syndrome was excluded. Past medical history showed a long standing depression treated with paroxetine and mitral valve replacement for an endocarditis 10 years before. Hypercholesterolemia treated with statin and hypoacusia were also present. After 3 years an EMG with repetitive nerve stimulation showed an abnormal decremental response with low-rate stimulation and a myasthenic syndrome was suspected. Neurological examination was normal including fatigability test exempt a slightly ataxic gait and absent deep tendon reflexes that reappeared weakly after isometric contraction.

At this point antibodies against neuromuscular junction were tested. Anti-acetylcholine receptor antibodies were negative while anti voltage-gated calcium channels antibodies were positive at high titre (300 pmol/L; normal range 0.0–24.5 pmol/L). A new electrophysiologic study demonstrated an increase in compound motor action potentials in several nerve distributions after isometric contraction. A PET total body was negative and considering the time from onset the patient seems to have a non paraneoplatic LEMS. Symptomatic and corticosteroid treatment rapidly improved weakness, fatigability, xerostomia and xerophthalmia.

Conclusion

This case is peculiar because its onset after travelling to China and for the prominent autonomic symptoms. This latter feature together with moderate fluctuating weakness and normal neurological examination delayed the diagnosis for almost 3 years. LEMS is a rare dissease but probably is underdiagnosed and antibodies against voltage-gated calcium channel should be part of screening when symptoms of autonomic failure are present.