

Progressive supranuclear palsy complicated with Lambert-Eaton myasthenic syndrome : report of a case

Ai Tsukamoto^{#1}, Hiroaki Osaki^{#2}, Yuishin Izumi^{#2}, Ryuji Kaji^{#2}, Takao Mitsui^{#1}

#1. Department of Neurology, Tokushima National Hospital, National Hospital Organization, 1354 Shikiji, Kamojima, Yoshinogawa, Tokushima 776-8585 Japan

#2. Department of Neurology, Tokushima University Hospital, 2-50-1 Kuramoto-cho, Tokushima 770-8503 Japan

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Abstract

The case was an 83-year-old man. Prostate cancer had been diagnosed at 76 years of age and he had received hormonal therapy. Cognitive impairment, slow movement, gait disturbance and dysuria developed at 81 years old. He had a diagnosis of progressive supranuclear palsy. When he was hospitalized in Tokushima National Hospital, in addition to the neurologic symptoms mentioned above, there were blepharoptosis and a decrease of deep tendon reflexes. The waxing phenomenon was detected by an examination for high-frequency repetitive stimulation in the hypotherner muscle. Blood antiP/Q type VGCC antibody was positive. Lambert-Eaton myasthenic syndrome was diagnosed from the above-mentioned findings. The disorder is regarded as Lambert-Eaton myasthenic syndrome with prostate cancer. The mechanism of the association between this disorder and progressive supranuclear palsy is not clear.

Key words: Lambert-Eaton myasthenic syndrome, paraneoplastic syndrome, prostate cancer, anti-P/Q type VGCC antibody

Introduction

Lambert-Eaton myasthenic syndrome (Lambert-Eaton myasthenic syndrome: LEMS) is one of the paraneoplastic syndromes, and is frequently complicated with small cell lung cancer. We had a case of LEMS with prostate cancer complicated with progressive supranuclear palsy (PSP). There were few reports that LEMS merged for prostate cancer [1,2]. Furthermore, no report of a case in which LEMS and PSP merged was found.

Case Report

The patient, an 83-year-old man, suffered from dysphagia, anarthria and gait disturbance. He had been given a diagnosis of prostate cancer in 2010 and had received hormonal therapy. His oldest brother and a younger brother had colorectal cancer. There had been no consanguineous marriage. He

could not use a cell-phone and was not able to calculate from November 2015. He was unable to speak clearly from the same period, and affective incontinence was found, too. In about March 2016, it became hard for him to open his eyes, and gait disturbance and urinary retention developed. Because slow movement, cognitive functional decline and gait disturbance were detected, he was transferred to the University of Tokushima Hospital and had a medical examination with a diagnosis of progressive supranuclear palsy (PSP). Because home treatment was difficult, he transferred to Tokushima National Hospital for long-term medical treatment on November 1, 2016. His height was 165 cm, the weight was 61.8 kg, the blood pressure was 125/51mmHg, and the pulse was 62 a minute. He showed deorsumduction restrictions, dysphagia, anarthria, slow movement, a decrease of deep tendon reflexes, a severe decrease from

bilateral feet malleolus medialis moderate degree, posture reflex disturbance, dysuria on bilateral ptosis, ocular motility neurologically. There was no clear muscle weakness, but several steps were available for the walk if there was assistance. Laboratory findings demonstrated that hemoglobin had decreased to 11.3 g/dl, platelet had decreased to 124,000 / μ l. Serum albumin and total cholesterol had decreased to 3.2 g/dl and 102 mg/dl, respectively. Creatine kinase and C-reactive protein were 49IU/l and 0.17mg/dl, respectively. Human cerebral natriuretic peptides (BNP) had increased to 385.2pg/ml. The tumor markers including PSA, ProGRP, NSE, CEA, CA19-9, and soluble IL-2 receptor were negative. The anti-acetylcholine receptor antibody, the anti-Musk antibody, anti-nuclear antibody, and rheumatoid factor were negative. The antiP/Q type VGCC antibody had risen to 267.0pmol/l (normal, <20). The electrocardiogram accepted ventricular three steps of pulses, negative T wave in I, II, III, aVF, V3-6. There was no neoplastic lesion other than prostate cancer by trunkal CT. Echocardiography showed a left ventricle relaxation abnormality pattern and expansion of the left atrium. Ventricular extrasystole occurred frequently all day long as shown by the Holter electrocardiogram. SNAP decreased to 2 μ V in both sural nerve in the examination of nerve conduction. CMAP of the left tibial nerve decreased to 1.17mV. The high-frequency stimulation test of 30Hz of the right ulnar nerve at rest showed the waxing phenomenon (0.24 \rightarrow 1.2mV) (Figure 1). There was mild atrophy of the tegmentum mesencephali in the brain MRI (Figure 2). In the examination of cognitive function, replies were slow and MMSE, HDS-R22 and FAB were 26/30, 22/30, and 13/18.

Discussion

This patient did not have clear muscle weakness in the four limbs, but dysphagia matched blepharoptosis. The deep tendon reflexes decreased, and there was dysuria. A high-frequency repetitive stimulation study showed waxing of the amplitude.

Furthermore, antiP/Q type VGCC antibody was positive. LEMS was diagnosed than the above. In LEMS, antiP/Q type VGCC antibody is detected in approximately 85% of patients. LEMS is a paraneoplastic syndrome complicated with small cell lung cancer in approximately 60 [3].

Malignant lymphoma, leukemia, prostate cancer, rectal adenocarcinoma, laryngeal cancer, and bladder cancer are complicated malignant tumors other than lung cancer [4]. This patient received hormonal therapy for prostate cancer that has been well controlled. We found only two reports that merged LEMS for prostate cancer. As for one of these cases, amyosthenia developed five years after prostate cancer was diagnosed. When amyosthenia developed, adenocarcinoma and small cell carcinoma coexisted. It is thought that it is necessary for this case to follow up whether these changes do not appear carefully. Because cases of merger between LEMS and PSP have not been reported, it is more likely to be an accidental merger. However, there will be room for examination of this possibility in the future.

References

1. Têtu B, Ro JY, Ayala AG, Ordóñez NG, Logothetis CJ, von Eschenbach AC. Small cell carcinoma of prostate associated with myasthenic (Eaton-Lambert) syndrome. *Urology* 33; 148-52, 1989
2. Agarawal SK, Birch BR, Abercrombie GF. Adenocarcinoma of the prostate and Eaton-Lambert syndrome. A previously unreported association. *Scand J Urol Nephrol* 29; 351-3, 1995
3. Motomura M. Clinical condition and treatment of <education lecture 15> autoimmunization-related myoneural junction disease. *Clinical neurology* 51; 872-876, 2011/
4. Motomura M, Fukuda T. Lambert-Eaton myasthenic syndrome. *Brain and Nerve* 63, 745-754, 2011

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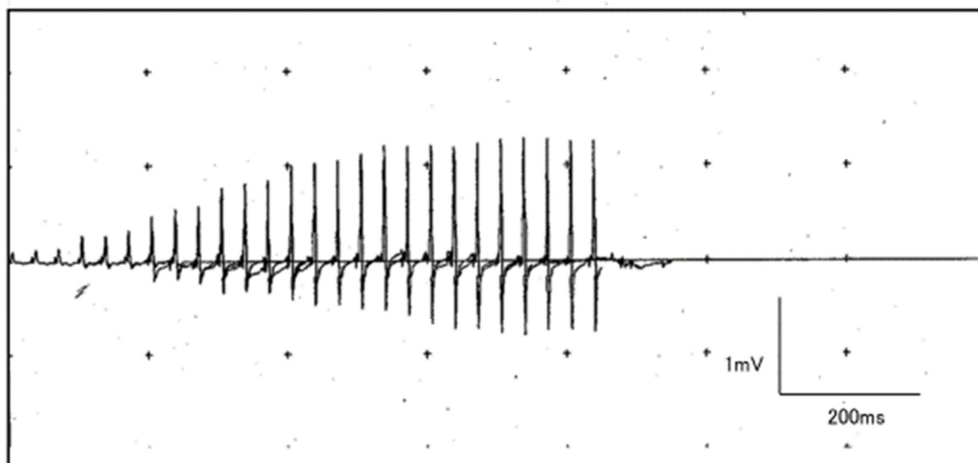


Figure 1. High-frequency stimulation test of 30Hz of the right ulnar nerve at rest showed the waxing phenomenon.

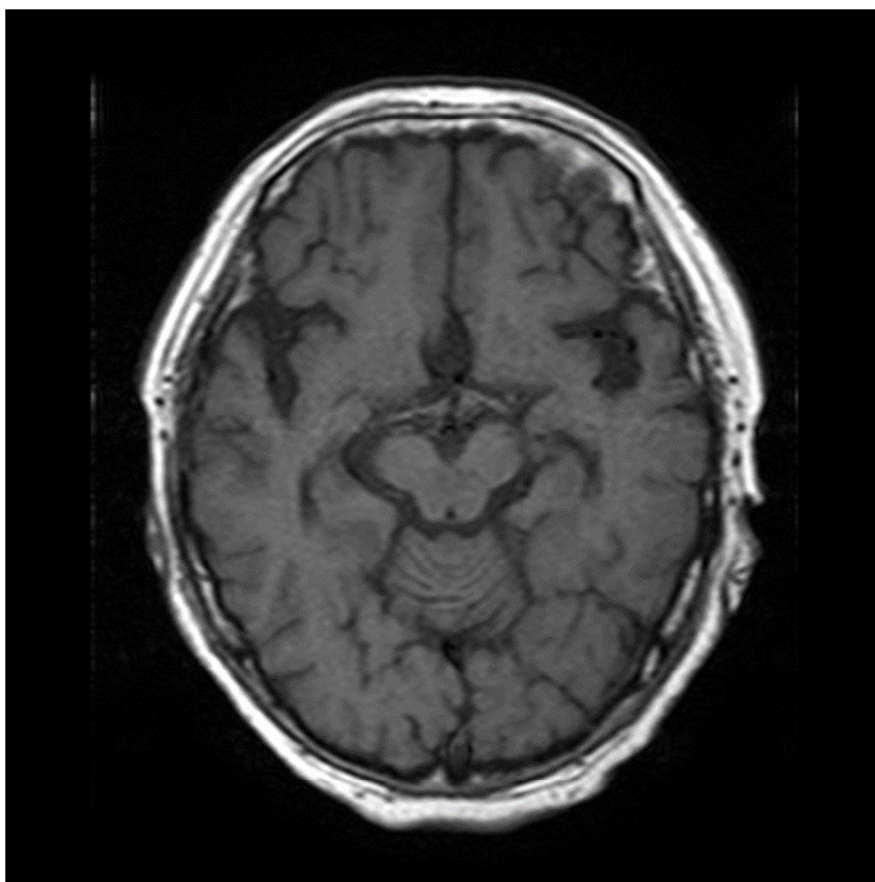


Figure 2. Brain MRI. Axial T1-weighted image showing midbrain atrophy.