Alpha-synuclein Accumulation in a Patient with Auerbach’s Plexus of Pure Autonomic Failure

Taeko Kurashiki-Osaka¹, Tadashi Adachi¹,², Hiroyuki Nakayasu¹ and Kenji Nakashima²

Key words: Lewy body, alpha-synuclein, pure autonomic failure, Auerbach’s plexus, gastrointestinal tract, peripheral nerve

(A Intern Med 53: 2261, 2014)
(DOI: 10.2169/internalmedicine.53.2294)

A 76-year-old man complained of orthostatic hypotension and a disturbance of micturition. Six months later, gastric carcinoma was diagnosed and partial gastrectomy was performed. Two months after the operation, the patient was referred to our neurology department due to repeated syncope. A neurological examination revealed urinary retention and orthostatic hypotension; however, no gastrointestinal symptoms, cardiac dysfunction or extrapyramidal signs were observed. A head-up tilt test showed severe hypotension (baseline, 160/105 mmHg; at 60°, 112/77 mmHg), and the level of plasma catecholamine secretion was decreased. Furthermore, a decreased heart/mediastinum ratio (1.58) was evident on ¹²³I-metaiodobenzylguanidine scintigraphy. Pure autonomic failure (PAF) was diagnosed according to the diagnostic criteria of the American Autonomic Society and American Academy of Neurology (1). Alpha-synuclein immunostaining demonstrated intraneuritic alpha-synuclein deposits in Auerbach’s plexus of the resected stomach (Picture). These findings support the concept that PAF represents an alpha-synucleinopathy (2).

The authors state that they have no Conflict of Interest (COI).

Acknowledgement
The authors wish to thank Dr. Shu Nakamoto for his kind preparation of the pathological samples.

References

¹Department of Neurology, Tottori Prefectural Central Hospital, Japan and ²Department of Brain and Neurosciences, Division of Neurology, Faculty of Medicine, Tottori University, Japan
Received for publication December 10, 2013; Accepted for publication April 29, 2014
Correspondence to Dr. Hiroyuki Nakayasu, nakayasuhi@gmail.com
© 2014 The Japanese Society of Internal Medicine Journal Website: http://www.naika.or.jp/imonline/index.html