Journal of the Canadian Association of Gastroenterology, 2018, 1(1), 2–3
doi: 10.1093/jcag/gwy002
Image of the Month
Advance Access publication 19 March 2018

OXFORD

Image of the Month

A Case Illustrating the Natural Progression of Type III to Type II Achalasia

Achalasia is characterized by loss of esophageal peristalsis and impaired lower esophageal sphincter (LES) relaxation. Progressive loss of nitrergic inhibitory innervation in the esophagus likely plays a major role in the progression of achalasia (1). This hypothesis is supported by histological studies in patients diagnosed with achalasia, in whom there is a decrease of neurons containing nitric oxide synthase and interstitial cells of Cajal, a unit that likely modulates nitrergic nerve response (2, 3). Given that the normal physiology of esophageal peristalsis and LES relaxation is dependent on nitrergic innervation (4), it has been hypothesized that achalasia naturally progresses from spastic type III, to pan-pressurization seen in type II, and finally, to loss of esophageal pressurization in advanced type I achalasia (5). To date, there is no data supporting the hypothesis of disease progression between achalasia subtypes.

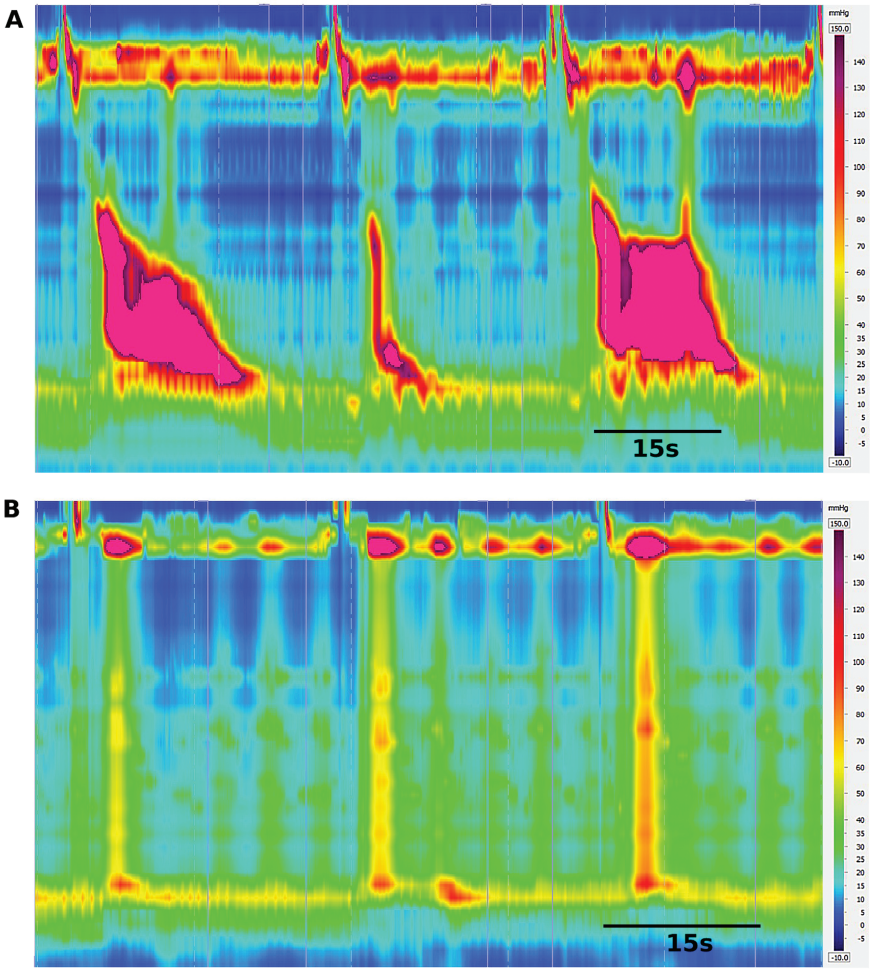

Figure 1. High-resolution esophageal manometry study of the index case: (A) demonstrating type III achalasia at the initial visit; (B) progressing into type II achalasia in the subsequent study two years later.

We report a case of a previously healthy 60-year-old male who presented with dysphagia and was subsequently diagnosed with type III achalasia by high-resolution esophageal manometry study (EMS) (Figure 1A). Following the initial diagnosis, he underwent two pneumatic dilatations without any significant clinical improvement of his dysphagia. Two years later, a repeat EMS demonstrated type II achalasia (Figure 1B).

This is the first longitudinal clinical case that demonstrates the progression from type III to type II achalasia. It not only strengthens our current understanding of the natural history of achalasia but also supports the hypothesis of the loss of nitrergic inhibitory innervation in the pathogenesis of achalasia.

Ann T. Ma, MD[1],
Louis W. C. Liu, BEng, MEng, PhD[1,2]

[1]Division of Gastroenterology, Department of Medicine, University of Toronto, 200 Elizabeth Street, Toronto, Ontario, M5G 2C4, Canada; [2]Department of Medicine, Toronto Western Hospital, University Health Network, 399 Bathurst St, 8MP-329. Toronto, Ontario, M5T 2S8, Toronto, Ontario, Canada

***Correspondence:** Louis WC Liu, Division of Gastroenterology, Toronto Western Hospital, University Health Network, 399 Bathurst Street, 8MP-329, Toronto, Ontario, Canada, M5T 2S8, e-mail louis.liu@uhn.ca*

References

1. Ates F, Vaezi MF. The pathogenesis and management of Achalasia: current status and future directions. Gut Liver 2015;9(4):449–63.
2. Gockel I, Bohl JR, Eckardt VF, Junginger T. Reduction of interstitial cells of Cajal (ICC) associated with neuronal nitric oxide synthase (n-NOS) in patients with achalasia. Am J Gastroenterol 2008;103(4):856–64.
3. Chen JH, Wang XY, Liu LW, et al. On the origin of rhythmic contractile activity of the esophagus in early achalasia, a clinical case study. Front Neurosci 2013;7:77.
4. Paterson W, Mayrand S, Mercer C. Chapter 2: The Esophagus. In: Thomson ABR, and Shaffer EA, ed. First Principles of Gastroenterology and Hepatology. CAPstone Academic Publishers Ltd. 2012:45–82.
5. Kahrilas PJ, Bredenoord AJ, Fox M, et al.; International Working Group for Disorders of Gastrointestinal Motility and Function. Expert consensus document: Advances in the management of oesophageal motility disorders in the era of high-resolution manometry: a focus on achalasia syndromes. Nat Rev Gastroenterol Hepatol 2017;14(11):677–88.