A 58 year-old woman visited our department with a 2-month history of dysphagia on liquids and solids. Barium esophagogram showed pseudodiverticula suggesting simultaneous esophageal contractions and tapering at the gastroesophageal junction (Fig. 1). Esophageal manometry was performed in a standardized manner using an 8-channel water-perfused manometry catheter (Synectics Medtronic, Stockholm, Sweden). We diagnosed her with diffuse esophageal spasm (DES) because the manometric features showed simultaneous contractions associated with >10% of wet swallows, mean simultaneous contraction amplitude of >30 mmHg and multiple peak contraction (Fig. 2A). She was advised to take diltiazem 30 mg 3 times a day and her symptoms improved. However, this motility disorder progressed to classic achalasia within 8 months (Fig. 2B). After pneumatic balloon dilatation, her dysphagia improved.

Several studies suggest that the motility disorder such as DES, nutcracker esophagus and achalasia share a common pathophysiologic mechanism, which involves the alteration in nitric oxide synthesis/degradation or loss of nitric oxide containing inhibitory neurons in the lower esophageal sphincter. The loss of intramural inhibitory neurons leads to the loss of normal peristalsis and inability of the lower esophageal sphincter to relax properly during swallowing. Some reports support the notion of progression of DES to achalasia. However, recent prospective cohort study showed that progression from DES to achalasia was uncommon (8%) and did not identify predictors of progression to achalasia based on the initial manometry parameters. So, longer follow-up diagnostic tool such as high resolution manometry may help assess more accurately the pathophysiologic mechanism and true rate of progression from DES to achalasia.

Figure 1. Barium esophagogram shows pseudodiverticula suggesting simultaneous esophageal contractions and tapering at the gastro- esophageal junction.
Progression of Diffuse Esophageal Spasm to Achalasia

Figure 2. Esophageal manometry. (A) Manometric findings show simultaneous contractions associated with > 10% of wet swallows, mean simultaneous contraction amplitude of > 30 mmHg and multiple peaked contractions. (B) After 8 months, follow-up manometric features show that lower esophageal sphincter relaxation become incomplete and contractions become to have low contraction amplitudes, which are the typical findings of classic achalasia. LES, lower esophageal sphincter.

References