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【 LETTERS TO THE EDITOR 】

Secondary Achalasia in Myotonic Dystrophy May Have a Different Pathology and Management

Key words: myotonic dystrophy, skeletal muscle, smooth muscle, achalasia, myotomy, fundoplication

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The Authors Reply We read with interest the response to our article in *Internal Medicine,* "Affection of the gastrointestinal smooth muscles in myotonic dystrophy is not usual," authored by Finsterer et al. As the title suggests, we agree that the diagnosis of secondary achalasia in myotonic dystrophy is transient, and on follow-up examinations, the diagnosis changes to aperistalsis. Furthermore, symptoms arising from damaged cricopharyngeal striated muscle appear more severe than those arising from the esophageal smooth muscle, which is the usual focus in the clinical setting.

Our patient had myotonic dystrophy-1 with CTG expansions in dystrophia myotonica-protein kinase (*DMPK*). She had skeletal muscle myotonia when myotonic dystrophy was first diagnosed, and her cricopharyngeal symptoms appeared later than the other skeletal muscle symptoms. Early achalasia has no subjective symptoms; therefore, whether or not she had objective findings of achalasia at the onset of myotonic dystrophy is unclear. Other gastrointestinal involvement, including megacolon and chronic constipation, and cardiac diseases such as AV-block were not observed. In older reports of myotonic dystrophy, gastrointestinal involvement has been reported (1); further studies using advanced techniques, such as high-resolution manometry, are necessary to clarify the concomitant rate of gastrointestinal motility disorders. Gastrointestinal symptoms are common, and myotonic dystrophy may also cause gastrointestinal symptoms through neuropathy or endocrine system disturbances other than motility disturbance.

Achalasia is a rare acquired esophageal motility disorder of an unknown etiology and may be associated with autoimmune mechanisms, viral infection, and genetic susceptibility (2, 3). In achalasia, the esophageal branch of the vagal nerve is damaged, and the Auerbach plexus in the esophagus is histologically degenerated. However, other gastrointestinal involvement is rare (4), and cardiac involvement in such cases tends to not be more than that of the general population. In contrast, myotonic dystrophy is an inherited disorder and the most common form of muscular dystrophy. Furthermore, myotonic dystrophy is associated with various complications that are different from those of achalasia.

For cricopharyngeal and esophageal symptoms in myotonic dystrophy, interventions should be considered depending on the general condition and prognosis of the patients. In our patient, reflux esophagitis was not observed; therefore, minimally invasive treatment, such as balloon dilation and peroral endoscopic myotomy (5), might be considered in order to target the loss of relaxation of the lower esophageal sphincter due to secondary achalasia. However, cricopharyngeal achalasia, which is more common and severe in cases of myotonic dystrophy than secondary achalasia, cannot be treated by techniques that focus on the lower esophageal sphincter. Thus, the efficacy of such esophageal interventions for alleviating dysphagia and preventing aspiration is unclear and may be limited.

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

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