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Repetitive Proximal Esophageal Contractions: A New Manometric Finding and a Possible Further Link Between Parkinson's Disease and Achalasia

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Abstract. Repetitive, spontaneous contractions of the proximal esophagus have recently been identified as a feature of achalasia. This article documents similar findings in six patients with Parkinson's disease. Parkinson's disease and achalasia share many common features neurologically. Both have Lewy bodies in the esophageal myenteric plexuses and the substantia nigra, in addition to evidence of degeneration of the dorsal motor nucleus of the vagus. The esophageal features radiologically and manometrically are also similar. Repetitive proximal esophageal contractions may represent another link between these diseases. They have also been reported in scleroderma. We speculate that the common link between all three disease processes may be poor distensibility of the esophagus.

Key words: Achalasia — Esophageal motor abnormalities — Parkinson's disease — Lewy bodies — Scleroderma — Deglutition — Deglutition disorders.

Repetitive proximal esophageal contractions (RPECS) have recently been identified as a feature of achalasia [1]. They were described as repetitive spontaneous contractions of the proximal esophagus and were noted in two-thirds of patients in whom achalasia was diagnosed. In this current study, similar features are reported in another condition, Parkinson's disease (PD). For many years it has been suggested that there is an association between PD and achalasia. The diseases have esophageal and neurological abnormalities in common. Repetitive proximal esophageal contractions may represent yet another link between the two conditions.

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Patients and Methods

As part of a research project investigating esophageal and oropharyngeal function in neurological disorders, 22 patients with PD have been assessed. The median age of the patients was 67 (range = 50–84) years and 17 were male. After an overnight fast, esophageal manometry was performed using a catheter comprising four solid-state intraluminal pressure transducers separated at 5-cm intervals (Konigsberg Instruments Inc., Pasadena, CA). Pressure data were collected and analyzed using Polygram equipment and software (Synectics Medical Inc., Irving, TX).

The lower esophageal sphincter (LES) was assessed using a station pull-through technique and its relaxation response to six wet swallows was measured. Esophageal body manometry was performed in two segments: The lower esophageal motility was recorded 2, 7, and 12 cm above the LES, and proximal esophageal motility was recorded 1, 6, and 11 cm below the upper esophageal sphincter (UES). Ten wet swallows were recorded in both positions.

Results

Repetitive, spontaneous contractions of the proximal esophagus were noted in 6 (27%) patients. In 2 patients, the contractions occurred simultaneously in all three proximal channels (Fig. 1). In the other

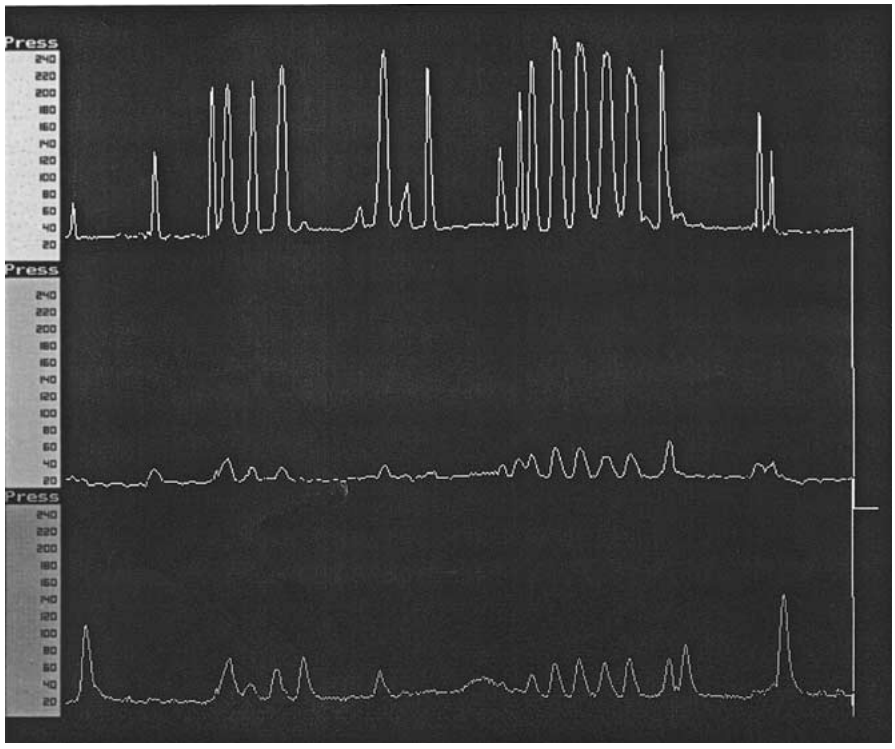

Fig. 1. Repetitive proximal esophageal contractions present at all three proximal transducers (1–11 cm below the UES).

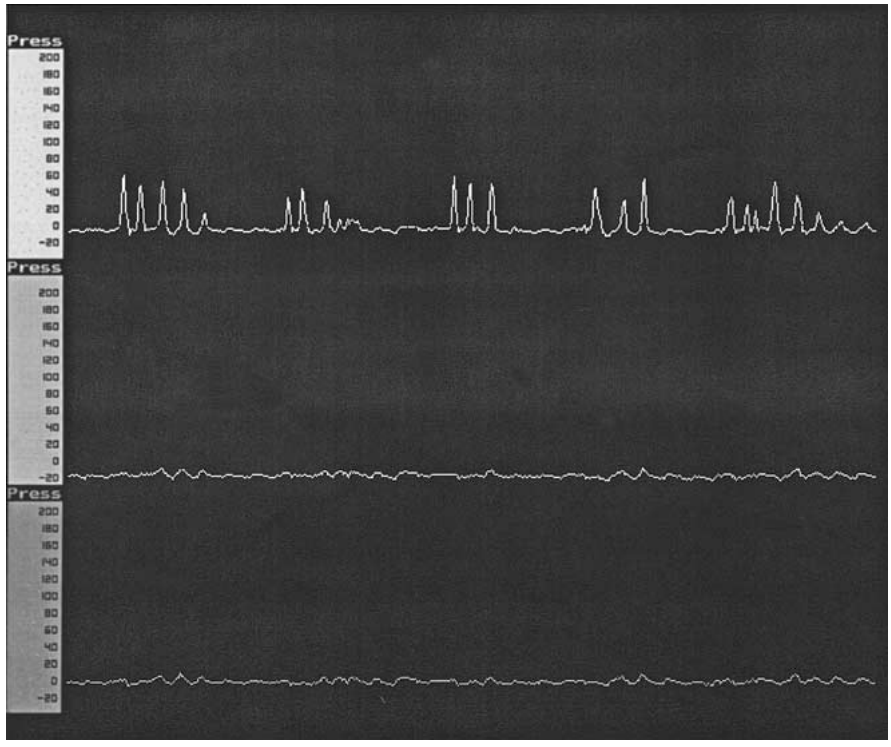

Fig. 2. Repetitive proximal esophageal contractions confined to the most proximal segment of the upper esophagus. Repetitive proximal esophageal contractions begin in response to a swallow.

4 patients, repetitive activity was confined to the most proximal transducer (Fig. 2). The repetitive activity had a frequency of 0.22–0.30 Hz and occurred both spontaneously and in response to swallowing. In none of these 6 patients did the contractions also occur in the distal esophagus, and in only one of the other 16 subjects was there spontaneous, repetitive activity in the distal esophagus. However, in 5 of the 6 cases of RPECS, abnormalities of distal esophageal motility were recorded. In 3 patients, simultaneous contractions (60%–90%) occurred in response to water swallows, the other peristaltic waves being of low amplitude in one of these three. In 2 patients, there was complete absence of peristalsis. LES pressure and relaxation were normal in both patients.

Comparing subjects with and without RPECS, there was no significant difference in duration of parkinsonian symptoms (median = 6.0 vs. 9.5 years), Hoehn and Yahr severity scale (1.5 vs. 2.0), or self-rated dysphagia (3.5 vs. 5.0 on a scale of 7 to 1, where 7 represents normal swallowing and 1 an inability to consume anything orally).

Discussion

In our study, repetitive proximal esophageal contractions have been identified in six (27%) patients with PD. This abnormality has also recently been reported as occurring in achalasia. On the basis of our study, RPECS do not appear to be a major factor influencing each patient's degree of dysphagia. Nor are they related to prolonged or severe PD. This contrasts with the findings of Weber et al. [2] who suggested a correlation of severity with the Hoehn and Yahr scale. However, their findings were based on only six patients.

Esophageal spasms have been documented radiologically in PD as tertiary contractions along the body of the esophagus [3,4]. No study reported the spasms as being restricted to the proximal esophagus. There have been few systematic studies of esophageal manometric findings in PD. In 1965, Fisher et al. [5] studied seven PD patients and described both spontaneous, simultaneous contractions of the esophageal body and complete absence of peristalsis. Bramble et al. [6] reported their manometric findings in 20 patients, noting normal baseline motility but a significant increase in nonperistaltic activity after the administration of atropine. More recently, repetitive contractions of the esophageal body were noted in 4 of 18 PD patients [7]. No study has described repetitive, spontaneous activity confined to the proximal esophagus.

The restriction of these contractions to the proximal esophagus in our subjects, with absence of similar dysmotility distally, suggests that it may be related to striated muscle or its neurologic control. Several possible explanations must be considered. First, RPECS may be a manifestation of the pill-rolling tremor seen in other striated muscle in PD. However, the rate of a pill-rolling tremor is classically described as 3.5–7 cycles/s and has a uniform frequency for all muscle groups, including the tongue [8–10]. This rate is faster than these repetitive esophageal cycles which have a very consistent frequency of 0.2–0.3 Hz. Second, the upper transducer might be slipping briefly in and out of a tremulous UES. Arguing against this is the constant esophageal baseline pressure on either side of the contractions, even during swallowing, indicating that the UES is not being entered. It would also not explain the RPECS recorded by all three transducers in two patients.

A third hypothesis to be considered is that RPECS are a drug side effect. All 6 patients had taken their medication within two hours of the study. However, 2 patients were also studied 15 hours off medication, in addition to the manometry one hour after medication; in one of these patients, RPECS were present in both tracings. Dopamine has been documented to induce simultaneous contractions of the esophagus in chickens [11] and opossums [12]. However, in no case were the contractions confined to the proximal esophagus, and the hypothesized mechanism of action involved stimulation of dopaminergic receptors present in both esophageal smooth muscle and the LES [12]. It would be of interest to study both upper and lower esophageal responses of normal human subjects to administrations of L-dopa.

Repetitive proximal esophageal contractions were recently reported in achalasia, and there is growing evidence of a link between PD and achalasia. Neuropathologic studies have identified Lewy bodies in achalasia patients. They occur predominantly in the esophageal myenteric plexuses, especially of the lower esophagus. They have also been found in the substantia nigra, the classical site for PD Lewy bodies [13]. Similar to achalasia, Lewy bodies have been noted in the esophageal myenteric plexuses in PD patients with dysphagia [14]. It is suggested that the presence of Lewy bodies outside the nigrostriatal tract reflects the sites of greatest PD involvement [15]. Other neurological findings common to both diseases include degeneration of the dorsal motor nucleus of the vagus, the origin of esophageal smooth muscle

efferents [16], and generalized autonomic nervous system dysfunction [17–19].

Radiologically and manometrically, both PD and achalasia have the following characteristics in common: dilated esophagus [20], aperistalsis [5], and poor LES relaxation [21]. Our study presents evidence that the abnormality of RPECS, only recently described in achalasia, is also present in PD. Repetitive proximal esophageal contractions may be confined to the segment immediately distal to the UES or involve the whole of the proximal esophagus. They may be swallow-related or spontaneous. All of these features are similar to the phenomenon reported in achalasia. This linkage with achalasia may also suggest a possible pathophysiologic mechanism for the phenomenon. Zhang and Diamant [1] speculated that the proximal contractions are a protective response to increased intraesophageal pressure, reducing the potential for aspiration. They suggested that the increased intraesophageal pressure is a result of higher muscle tone and reduced ability of the esophagus to distend. They further supported this hypothesis by noting the similar findings in response to balloon distension in the proximal esophagus [22].

Interestingly, scleroderma has also been reported to display similar contractions in the proximal esophagus [23], possibly also as a result of poor esophageal distensibility resulting from fibrotic infiltration of the musculature. Our study demonstrates that PD, which could limit esophageal distensibility by muscular rigidity, also has similar findings.

In conclusion, our study reports the finding of RPECS in Parkinson's disease. RPECS was previously described in achalasia and scleroderma. Neuropathologic similarities between PD and achalasia were discussed and a possible unifying feature of poor esophageal distensibility was outlined for all three disease processes.

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