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Variant achalasia, the hole of the original classification

ESOPHAGUS

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Original Article

ABSTRACT

Background/Aims: High-resolution manometry (HRM) is a useful tool for classifying esophageal motility disorders. However, there remain findings that cannot be classified in clinical fields. Recently, an updated classification system was announced. The purpose of this study was to evaluate whether originally unclassifiable groups can now be classified according to the updated Chicago Classification system.

Materials and Methods: We reviewed the results of HRM studies performed from January 2008 to December 2010 on 150 consecutive patients (75 men, age 17-76) referred to the Gospel Hospital manometry laboratory for evaluation. We found originally unclassified results and re-categorized them according to the updated Chicago Classification system.

Results: Thirty-seven of 150 patients were originally unclassified cases. Patients from the unclassified and classified groups had similar distributions of age and sex. All unclassified patients were re-diagnosed as having variant achalasia according to the updated Chicago Classification system.

Conclusion: The updated Chicago Classification can categorize originally unclassified groups of esophageal motility disorder.

Keywords: Esophagus, achalasia, manometry, classification

INTRODUCTION

Solid-state high-resolution manometry (HRM) is capable of simultaneously monitoring the entire axial pressure profile from the pharynx to the proximal stomach and has sophisticated topographic plotting algorithms, representing an unquestionable evolution in esophageal manometry (1). A large clinical study with 400 consecutive patients and 75 control subjects provided a platform to develop a systematic approach to analyzing esophageal motility using HRM and pressure topography plots (2). The resulting classification scheme has been named the Chicago Classification Criteria of Esophageal Motility Disorders. In our hospital, we started to diagnose patients according to the Chicago Classification system after 2008 and acknowledged that HRM can be a useful tool to classify esophageal motility disorders. However, we found some cases that could not be classified according to the Chicago Classification system. Therefore, we needed to separate our cases into two groups: a classified group that could be categorized by the Chicago Classification system and an unclassified group that could not be assigned to a category. The most recent meeting of the HRM Working Group was in Ascona, Switzerland, in conjunction with an international congress focused on the clinical evaluation of esophageal disease, at which the HRM Working Group announced a updated classification system (3). The purpose of this study was to review patient records and HRM data and see how many cases could not be assigned to a certain category defined by the original Chicago Classification system used in 2008 and 2010 and to determine whether these unclassified findings could be categorized according to the updated Chicago Classification system (3).

MATERIALS AND METHODS

Patients

We reviewed HRM results performed from January 2008 to December 2010 on 150 consecutive patients

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(75 men, age 17-76) who were referred to the Gospel Hospital manometry laboratory. These patients presented with a diverse set of pathological conditions to a tertiary-care practice specializing in the management of esophageal disease. Patients were interviewed and examined to assess symptoms and to perform anthropometric measurements. Patients were initially categorized by their dominant clinical symptom. Thirty patients were undergoing evaluation for dysphagia and 120 for predominantly gastroesophageal reflux disease (GERD) symptoms (heartburn, regurgitation). These are described in Table 1. No patients underwent surgical treatment for their condition such as pneumatic dilation, Heller myotomy, or both for achalasia or fundoplication for GERD. The Gospel Hospital Institutional Review Board approved the study protocol.

Getting data from HRM

We used a solid-state HRM assembly (Mano scan 360™, Sierra Scientific Instruments Inc., Los Angeles, CA) with 36 solid-state sensors spaced at 1-cm intervals (O.D. 4.2 mm). The calibration, and poststudy thermal correction was performed at each test (4). In brief, each sensor is circumferentially sensitive, accurate to within 1 mmHg, capable of recording transient pressure changes in excess of 6,000 mmHg/s, and zeroed to atmospheric pressure. Studies were performed with the patient in a supine position after at least a six-hour fast. The HRM assembly was passed transnasally and positioned to record from the hypopharynx to the stomach. After 5 min period of resting period to assess basal sphincter pressure, and ten 5 mL water swallows obtained in a supine posture. Subsequently, the data were analyzed using ManoView™ analysis software (Sierra Scientific Instruments Inc.) for topographic pressure plotting and analysis.

Analyzing HRM

We analyzed HRM findings based on the original Chicago Classification system and investigated the unclassified group (1). After then, we classified the HRM findings according to the updated Chicago Classification system published in 2012 (3).

Table 1. Baseline characteristics of patients

	n=150
Age, year, mean±SD	54.8±11.9
Age group	
11-40, n (%)	13 (8.6)
41-60, n (%)	87 (58.1)
61-80, n (%)	50 (33.3)
Sex, male, n (%)	75 (50)
Symptoms	
GERD, n (%)	30 (20)
Dysphagia, n (%)	120 (80)

SD: standard deviation; GERD: gastroesophageal reflux disease

Statistical analysis

Statistical analysis was performed using the Statistical Package for the Social Sciences (SPSS) software (version 16.0, SPSS, Chicago, IL, USA). Correlations of the proportion of unclassified manometric findings and age grouping or sex were analyzed by a two-sided Fisher exact test. Two-tailed p values less than 0.05 were regarded as statistically significant.

RESULTS

Baseline characteristics (Table 1)

The average age of all patients was 54.8 years old. When we separated patients into three groups according to age, patients between 41 and 60 years were prevalent. Male patients made up 50% (75 patients) of investigated patients. Twenty percent of patients had dysphagia and 80% of patients had GERD.

Classification according to the result of HRM (Table 2)

Among the 150 patients, 119 (79.9%) showed abnormal findings. Eighty-two patients were classified according to the Chicago Classification. Each classified finding and its number of patients are as follows: absent peristalsis, 3; intermittent hypotensive peristalsis, 17; frequent hypotensive peristalsis, 28; distal esophageal spasm, 6; classic achalasia, 3; spastic achalasia, 5; functional EGJ obstruction, 20. Thirty-seven patients are categorized as unclassified.

Age and sex distribution of unclassified group (Table 3)

When we analyzed the distribution of classified and unclassified patients according to age group and sex, no significant difference was seen.

Unclassified findings according to symptoms (Table 4)

There were four findings in unclassified patients. All patients demonstrated elevated integrated relaxation pressure (IRP) with specific following manometric presentations; ≥30% of

Table 2. Diagnosis according to high-resolution manometry results

	n=150
Normal findings, n (%)	31 (20.7)
Abnormal findings, n (%)	119 (79.3)
Classified findings, n (%)	82 (68.9)
Absent peristalsis, n	3
Intermittent hypotensive peristalsis, n	17
Frequent hypotensive peristalsis, n	28
Distal esophageal spasm, n	6
Classic achalasia, n	3
Spastic achalasia, n	5
Functional EGJ obstruction, n	20
Unclassified findings, n (%)	37 (31.1)

EGJ: esophagogastric junction

swallows with hypotensive or absent peristalsis, 10 patients with GERD and 6 patients with dysphagia; ≥70% of swallows with hypotensive or absent peristalsis, 2 patients with GERD; ≥30% of swallows with hypotensive or absent peristalsis and spasm (CFV>8 cm/s) with ≥20% of swallows, 10 patients with GERD and 6 patients with dysphagia; normal CFV, mean DCI>5000 and <8000 mmHg.s.cm, 3 patients with GERD. All of these unclassified findings were categorized as variant achalasia based on the updated Chicago Classification system (3).

DISCUSSION

Solid-state HRM is capable of simultaneously monitoring the entire axial pressure profile from the pharynx to the proximal stomach and has sophisticated topographic plotting algorithms, representing an unquestionable evolution in esophageal manometry (1). The application of HRM with topographic plots for assessment of esophageal motor function was first described by Clouse and Staiano in 1991 along with the observation that the propagation of esophageal peristalsis was not seamless (2). More recently, Fox et al. (5) combined HRM pressure topography plots with simultaneous videofluoroscopy to establish the correspondence between specific pressure topography signatures and impaired bolus transits. They concluded that HRM with pressure topography plotting was more accurate than conventional manometry in identifying impaired bolus transit attributable to either focal breaks in the peristaltic wave front or impaired esophagogastric junction (EGJ) relaxation. Moreover, according to a large scale study with 400 consecutive patients and 75 control subjects, resultant schemes were consistent with conventional classifications with the following caveats: (a) hypercontractile conditions are more specifically defined, (b) distinctions are made between simultaneous contractions attributable to rapidly propagated contractions and those attributable to compartmentalized esophageal pressurization, and (c) there is no "nonspecific esophageal motor disorder" classification (2).

High-resolution manometry has been proved to be an efficient tool to investigate esophageal motility disorders. However, we found that several conditions could not be categorized by the Chicago Classification. While we were considering the holes of the Chicago Classification, the HRM Working Group meeting proposed a updated Chicago Classification (3). According to our investigation, the originally unclassified 37 group is now categorized as variant achalasia. All unclassified results in the originally classification showed elevated IRP and some instances of intact peristalsis or weak peristalsis. In the updated Chicago Classification, the category of these findings was variant achalasia (3). The majority of patients (25 of 37) did not receive any specific treatment directed at their motility disorder. Ten patients received calcium channel blocker or nitroglycerine for their symptoms. Two of the 37 patients underwent laparoscopic distal esophageal myotomy.

The esophageal pressure topographic metric developed to optimally distinguish normal from impaired EGJ relaxation is the IRP (6,7). The upper limit of normal for the IRP using the HRM assembly is 15 mmHg (6). Therefore, elevated IRP is a key factor in diagnosing achalasia. One of the novel features of the updated Chicago Classification system compared to the previous one is the differentiation of achalasia into three known and variant types (3,8-10). In the originally classification, we could not categorize results with elevated IRB in some instances of intact or weak peristalsis (8-10). However, the updated classification system enabled us to diagnose this group as variant achalasia.

Table 3. Distribution comparison between classified and unclassified groups in age and sex according to high-resolution manometry findings

	Unclassified n=37	Classified n=82	p value
Male, n (%)	12	40	0.538
Age, year, mean±SD	51.2±14.3	55.6±11.0	0.174
Age group			0.376
11-40, n (%)	2	4	
41-60, n (%)	18	40	
61-80, n (%)	17	38	

SD: standard deviation

Table 4. Unclassified findings of originally high-resolution manometry matching with updated classification

	Unclassified in original classification		
Symptoms, n (%)	**GERD 25 (67.6)**	**Dysphagia 12 (32.4)**	**updated classification**
†Elevated IRP and following findings with			
≥30% of swallows with hypotensive peristalsis or absent peristalsis, n	10	6	Variant achalasia
≥70% of swallows with hypotensive peristalsis or absent peristalsis, n	2	0	Variant achalasia
≥30% of swallows with hypotensive peristalsis or spasm (CFV >8 cm/s) with ≥20% of swallows, n	10	6	Variant achalasia
Normal CFV, 8000 mmHg/s.cm>mean DCI >5000 mmHg/s.cm, n	3	0	Variant achalasia

GERD: gastroesophageal reflux disease; IRP: integrated relaxation pressure; CFV: contractile front velocity; DCI: distal contractile integral
†These findings did not match with the diagnostic criteria of three subtypes of achalasia.

This study has several limitations. First, it is a retrospective study, so we did not have enough information to characterize variant achalasia compared with other types of achalasia. Therefore, we can only investigate the HRM findings. Moreover, we did not fully differentiate variant achalasia from primary or secondary hypercontractibility disorders. Third, we cannot be sure that our data represent general situations because it is a single center study, and our hospital is a tertiary referral center.

Up until now, we did not know much about variant achalasia; it is a somewhat mysterious disease. Variant achalaisa might be the the early stage of overt achalasia or may be an independent disease. This problem should be the focus of future research. However, we now can name these findings as variant achalasia and have basic data to advance our research.

Ethics Committee Approval: Ethics committee approval was received for this study from the ethics committee of The Gospel Hospital Institutional Review Board.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

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