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Temporal variation in manometric findings in achalasia: A case report

Akalazyada manometrik bulguların değişikliği: Bir olgu sunumu

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Esophageal manometry is a valuable tool in the diagnosis of achalasia. The manometric features proposed for diagnosing classic achalasia are incomplete relaxation of the lower esophageal sphincter and aperistalsis in the body of the esophagus. Atypical achalasia cases have been reported that do not have the characteristic manometric features of classic achalasia. We report the clinical, radiological and manometric follow-up of a 45-year-old woman who presented with atypical manometric features of achalasia that have not been reported in the literature and who after a short period demonstrated the manometric features of classical achalasia.

Key words: Achalasia, manometry, achalasia variants, atypical achalasia

Özofagus manometrisi akalazya tanısı için değerli bir yöntemdir. Alt özofagus sfinkterinin inkomplet gevşemesi ve özofagus gövdesinde peristalsis yokluğu, klasik akalazya tanısı için gerekli manometrik özelliklerdir. Klasik akalazyanın karakteristik manometrik özelliklerini göstermeyen atipik akalazya olguları bildirilmiştir. Burada, literatürde bugüne kadar bildirilmeyen akalazyanın atipik manometrik özellikleriyle başvuran ve kısa bir süre sonra akalazyanın klasik manometrik özelliklerini gösteren 45 yaşında bir bayan hastanın klinik, radyolojik ve manometrik izlemini sunuyoruz.

Anahtar kelimeler: Akalazya, manometri, akalazya varyantları, atipik akalazya

INTRODUCTION

Achalasia is the best known and best understood motility disorder of the esophagus. It is an inflammatory disease with loss of inhibitory neurons in the myenteric plexus of unknown etiology (1, 2). The degenerative process appears to involve preferentially the nitric oxide-producing inhibitory neurons that affect relaxation of esophageal smooth muscle. Loss of inhibitory innervation in the lower esophageal sphincter (LES) can cause increase in basal sphincter pressure and can interfere with normal relaxation. Inhibitory influences are necessary for normal peristalsis and thus loss of inhibitory neurons in the body can result in aperistalsis (3).

Although upper gastrointestinal endoscopy and barium esophagogram can be suggestive of the diagnosis of achalasia, esophageal manometry is regarded as the 'gold' standard diagnostic test. The manometric features proposed for the diagnosis of classic achalasia are incomplete relaxation of the LES and aperistalsis in the body of the esophagus. Other manometric features that are not required for the diagnosis of achalasia but provide supportive evidence are elevated intraesophageal pressure, hypertensive LES and isobaric waveforms (4). Atypical achalasia cases have been reported that do not have the characteristic manometric features of classic achalasia (5, 6).

We present a case with esophageal motility disorder whose manometric features at initial examination were atypical for achalasia and changed during follow-up until the typical features appeared. Thus, we discuss and compare our case with various manometric presentations of esophageal motility disorders, particularly achalasia.

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CASE REPORT

A 45-year-old woman was evaluated with a two-month history of dysphagia and retrosternal pain. Dysphagia occurred with solid and liquid foods, but was more pronounced with liquids and cold swallow. Occasionally, the patient felt a sensation of food sticking in her chest that resolved spontaneously. The chest pain radiated to her back, which was related or unrelated with swallowing. She did not experience weight loss, regurgitation or vomiting. Her physical examination was normal.

The patient's laboratory examination, including complete blood count, routine biochemical tests, electrocardiogram and chest radiography, was normal. Barium esophagogram (Figure 1) demonstrated tertiary non-peristaltic contractions. The upper gastrointestinal endoscopy was normal.

Esophageal manometry was performed with a multi-channel water perfused recording system (Synetics PC Polygraph). Analyses were performed with 'Synetics PW Esophageal Manometry Analysis Module' software. An eight lumen catheter (Dent-Sleeve) was used for synchronous manometric recordings of the LES and body of the esophagus.

The initial manometry (Figure 2, Table 1) showed a LES pressure of 30 mmHg and complete relaxation of the sphincter with swallowing. Forty percent of wet swallows created non-peristaltic contractions in the esophageal body. Additionally, esophageal body contractions tended to have high contraction amplitudes (mean 280 mmHg) and long contraction durations (mean 10 sec). Multi-peaked contractions were also observed. With these manometric findings, the diagnosis of diffuse esophageal spasm (DES) was considered, and a calcium-channel blocker was prescribed to the patient.

One month later, the second esophagus manometry demonstrated decrease in body contraction amplitudes and LES pressure as compared with the initial findings (Table 1). The patient's complaints did not improve and we began to follow her without any treatment. Her dysphagia became more severe three months later. A third manometry was performed (Figure 3, Table 1). The LES pressure was elevated to 52 mmHg with incomplete relaxation. Esophageal body contractions were aperistaltic in 90% of wet swallows, in which some had high amplitudes. A new esophagogram was done. The second esophagogram (Figure 4) showed narrowed ending of the esophagus like a bird's beak and a minimally dilated esophagus with mildly delayed emptying of barium into the stomach. At this time, with the diagnosis of achalasia, pneumatic balloon dilatation (30 mm achalasia ballo-

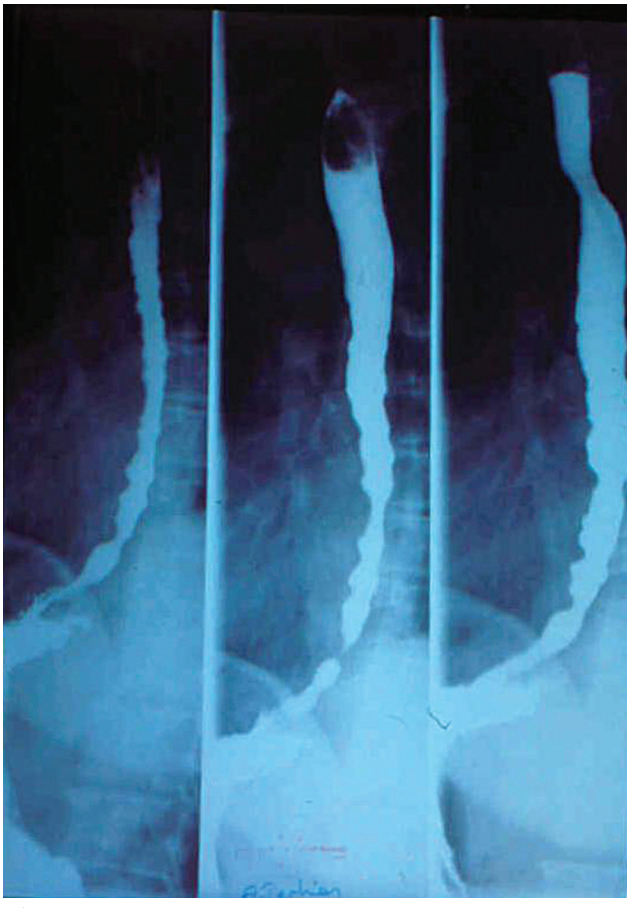

Figure 1. Initial esophagogram in January 2000 showed tertiary contractions with normal emptying.

Table 1. Esophageal manometric characteristics of the patient

	First Manometry	Second Manometry	Third Manometry	After Balloon Dilation
LES Pressure	30 mmHg	22 mmHg	52 mmHg	7 mmHg
Contraction amplitude	280 mmHg	90 mmHg	100 mmHg	28 mmHg
Contraction duration	10 sec	6 sec	8 sec	5 sec
Abnormal peristalsis	40%	50%	90%	100%
LES relaxation	Complete	Complete	Incomplete/absent	Incompetent LES

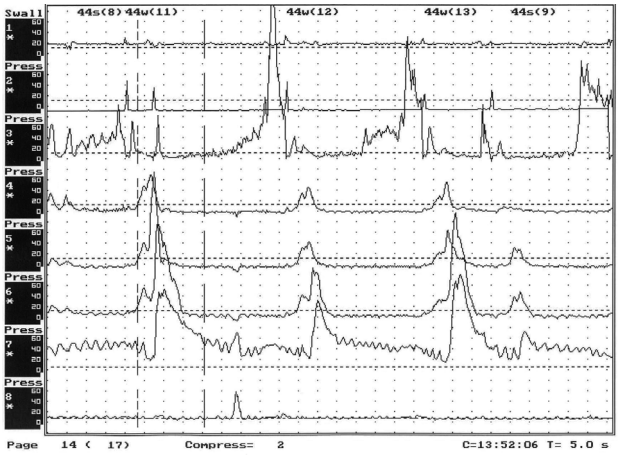

Figure 2. First esophageal manometry in January 2000 demonstrated non-peristaltic contractions in the body of the esophagus (channel 3-4-5-6) with normal LES function (channel 7). Some of the esophageal contractions were of high amplitude.

on dilator, Rigiflex, Microvasive, USA) was performed. Her symptoms resolved rapidly after balloon dilatation. Six months later, manometry (Figure 5, Table 1) showed low–amplitude aperistaltic body contractions and low LES pressure (mean 7 mmHg).

DISCUSSION

Currently, manometric definition of achalasia has evolved to include different features other than the classical findings of high LES pressure, incomplete relaxation and low amplitude waves of the body of the esophagus (5,6). Atypical cases have been reported that have manometric features not characteristic of classic achalasia (4,5). Vigorous achalasia is the best known atypical entity.

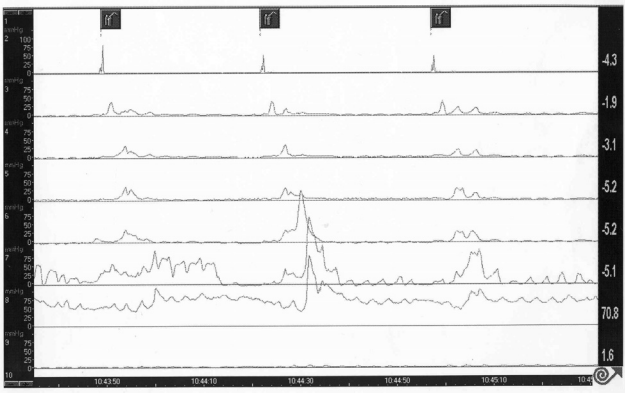

Figure 3. Third esophageal manometry in March 2000 showed approximately complete loss of peristalsis in the esophageal body and hypertensive LES with incomplete relaxation (channel 8).

Vantrappen et al. (7) suggested that primary esophageal motility disorders constitute a spectrum of motor disorders composed of achalasia, DES and related or intermediate types. In their study, 24% of patients with severe esophageal motility disorders constituted an intermediate form. This classification may be outdated because manometry techniques have evolved since the publication of the above report.

Recently, Hirano et al. (5) investigated manometric heterogeneity in patients with idiopathic achalasia and identified four manometrically distinct variants of achalasia: 1- the presence of high amplitude esophageal body contractions together with incomplete LES relaxation (vigorous achalasia), 2- a short segment of esophageal body aperistalsis together with incomplete LES relaxation, 3- complete LES relaxation together with esophageal body aperistalsis, and 4- impaired LES relaxation to wet swallows and aperistalsis in the body of the esophagus with transient complete LES relaxation.

Hirano et al. (5) demonstrated that manometric features of achalasia are quite heterogeneous. We think that our case demonstrates a variant of achalasia not reported by Hirano et al., although we did not obtain esophageal tissue for histological examination as the above study did. Our case had non-peristaltic repetitive high amplitude contractions similar to the features of Hirano's vigorous achalasia type. However, our case had complete LES relaxation in her first study, whereas Hirano's vigorous achalasia type patient lacked LES relaxation. Since Hirano's cases were not followed up, it is intriguing to speculate that some of their achalasia variants are in fact just manometric findings during the course of the disorder. Our case demonstrates different manometric presentations of achalasia over a period of time. Therefore, the importance of longitudinal follow-up of patients with atypical manometric features is evident as demonstrated by our case.

There is scanty information on the long-term behavior of these variants. There are a few case reports of patients with esophageal motility disorders whose initial manometric findings were reminiscent of DES, while manometric features of achalasia appeared several months or a few years later (7-13). Thus, the authors considered that transition of DES to achalasia is possible. However, in these studies, manometry was not utilized as frequently as in our study. Khatami et al. (14) pros-

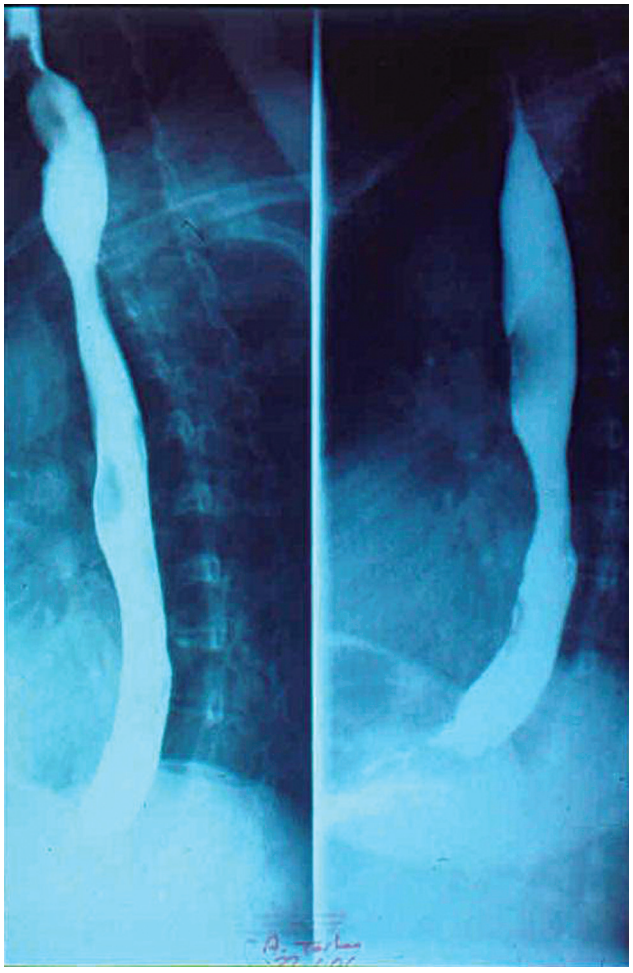

Figure 4. Second esophagogram in March 2000 demonstrated mild dilation of the esophageal body with narrowed ending of the esophagus similar to a bird's beak.

pectively investigated whether DES progresses to achalasia. They revealed that progression from DES to achalasia was uncommon and that during follow-up, only one of the 12 patients was diagnosed to have gained manometric features of achalasia. Furthermore, DES currently appears to be a very rare entity using stringent manometric criteria, i.e. normal LES relaxation and esophageal contractions of excessive amplitude and duration, temporally associated with chest pain and dysphagia (4,15). Under these circumstances, we suggest that those patients with DES reported to undergo alteration of their manometric features during follow-up had early phase achalasia because at the time of the reporting of these studies, the heterogeneity of manometric features of achalasia was not robustly defined.

Furthermore, prolonged manometric recordings provide additional information about the heterogeneous manometric features of achalasia. Prolonged recordings in patients with achalasia revealed the occurrence of complete LES relaxation, transient LES relaxations (TLESRs), variations in LES pressure associated with a meal or phase 3 and high amplitude and retrograde esophageal pressure waves (16). In some patients with classic achalasia, after balloon dilatation or Heller myotomy, return of peristalsis in the esophageal body and complete LES relaxation were demonstrated (17, 18).

Generally, the diagnosis of achalasia is delayed; the duration of symptoms at presentation averages about two years (19). Since most patients consult physicians after their symptoms are well established, it is possible that the early manometric features of the disease are under-recognized. At the time of presentation of our case, barium swallow and upper gastrointestinal endoscopy were normal. Furthermore, initial manometry showed 40% non-peristaltic high amplitude contractions with long duration in the esophageal body, which was typical of DES, and thus the diagnosis of DES was considered. However, on follow-up, the patient developed classical features of achalasia. It is conceivable that in the early stages of achalasia, some myenteric ganglia of the esophageal body are preserved (20).

It is important to recognize that many patients with dysphagia are found to have minor manometric abnormalities, such as occasional repetitive contractions, spontaneous contractions, simultaneous contractions, or impaired LES relaxation, but do not have a pattern that is diagnostic of eit-

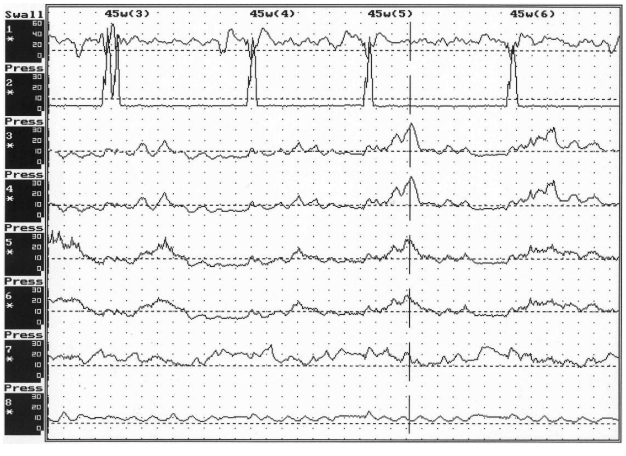

Figure 5. After balloon dilatation, the patient still presented manometric features of classical achalasia in the esophageal body and incompetent LES.

her DES or achalasia (8). Considering our case and the cases cited in the literature, we think that it is important to follow such indeterminate patients manometrically, because it is possible that temporal changes occur in the motility disorders of the esophagus, particularly in the case of achalasia.

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