

## Successful Therapeutic Combination of an Antispasmodic and Progressive Pneumatic Dilation in a Patient with Esophageal Motor Disorder: Importance of Differentiation of Classic and Vigorous Achalasia

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**ABSTRACT.** A 61-year-old Japanese woman who had been diagnosed as having classic achalasia was admitted to our hospital in April 199X because of dysphagia and chest pain of over 30 years' duration. Radiologic and manometric examinations resulted in a re-diagnosis of vigorous achalasia. Initial therapy with a pneumatic dilator relieved her dysphagia, but was ineffective for the chest pain. However, additional administration of an antispasmodic agent eliminated her esophageal symptoms completely. Although the concept of vigorous achalasia has been widely accepted in Western countries, to date only three cases of vigorous achalasia have been reported in Japan. When encountering a patient with dysphagia, not only should a diagnosis of achalasia be made, but also differentiation of its subtypes, classic or vigorous achalasia, should be done.

**Key words** ① vigorous achalasia ② pneumatic dilation ③ antispasmodic agent

Achalasia is a motor disorder of the esophageal smooth muscle in which the lower esophageal sphincter does not relax properly with swallowing. The term vigorous achalasia was first introduced by Olsen *et al*<sup>1)</sup> in 1957. Since repetitive, simultaneous large-amplitude contractions in patients with vigorous achalasia unlike classic achalasia, patients with this condition frequently complain of chest pain. Recently, however, Camacho-Lobato *et al*<sup>2)</sup> retrospectively analyzed the clinical and manometrical findings of 209 patients with achalasia, and concluded that chest pain was equally prevalent in both classic and vigorous achalasia.

We experienced a patient who had been followed up for over 30 years and who had been diagnosed with classic achalasia despite the presence of long-standing chest pain. This patient was successfully managed by a combination of pneumatic dilation and an antispasmodic agent after a re-diagnosis of vigorous achalasia was made. This case is herein described in detail.

## CASE REPORT

A 61-year-old Japanese woman was admitted to our hospital in April 199X, because of dysphagia and chest pain. In 1966, this patient was suffering from dysphagia and chest pain, and consulted her local clinic. After various examinations, a diagnosis of achalasia was made and medical treatment relieved her symptoms temporarily. However, such esophageal symptoms had repeatedly afflicted the patient thereafter.

On admission, a physical examination and laboratory data were unremarkable. A plain X-ray film of the chest disclosed an air-fluid level in a markedly widened mediastinum and the absence of a gastric air shadow.

A barium esophagram after intramuscular injection of an antispasmodic agent revealed a markedly dilated esophagus, barium stagnation and smooth tapering of the lower esophagus (Fig.1a). At its largest diameter, the esophagus measured 5.1 cm. These radiologic findings were compatible with those seen in classic achalasia. However, manometry disclosed not only elevated pressure of the lower esophageal sphincter (more than 20 mmHg) and failure of the lower sphincter to relax with swallowing, but also recurrent simultaneous contractions of high amplitude (Fig.2). These manometric findings suggested a diagnosis of vigorous achalasia. Reexamination of barium esophagram was thus indicated. Before intramuscular administration of an antispasmodic agent, an esophagram disclosed marked tortuosity and segmental spasm, with a corkscrew pattern (Fig.1b). Based on both radiologic and manometric features, a diagnosis of vigorous achalasia was confirmed.

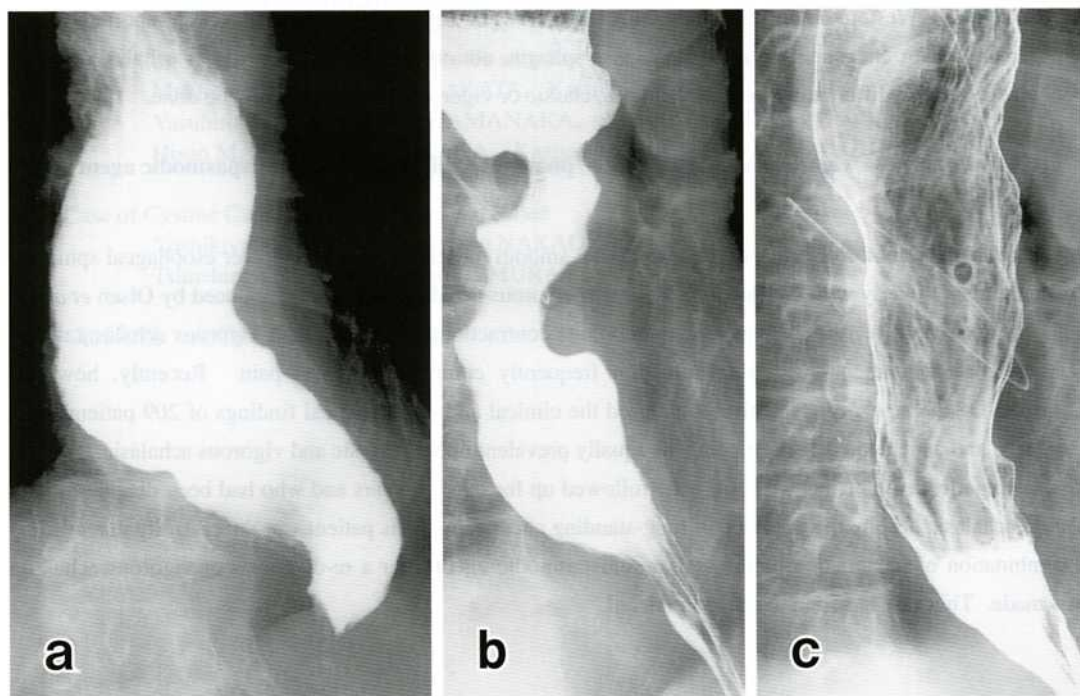


Fig. 1. A barium esophagram after injection of an antispasmodic demonstrated marked dilatation of the esophagus and smooth tapering of the lower esophagus (a). Before injection, however, a marked tortuosity and segmental spasms of the esophagus were observed (b). Following a combination of progressive pneumatic dilation and administration of an antispasmodic agent, barium stagnation and segmental spasms nearly disappeared (c).

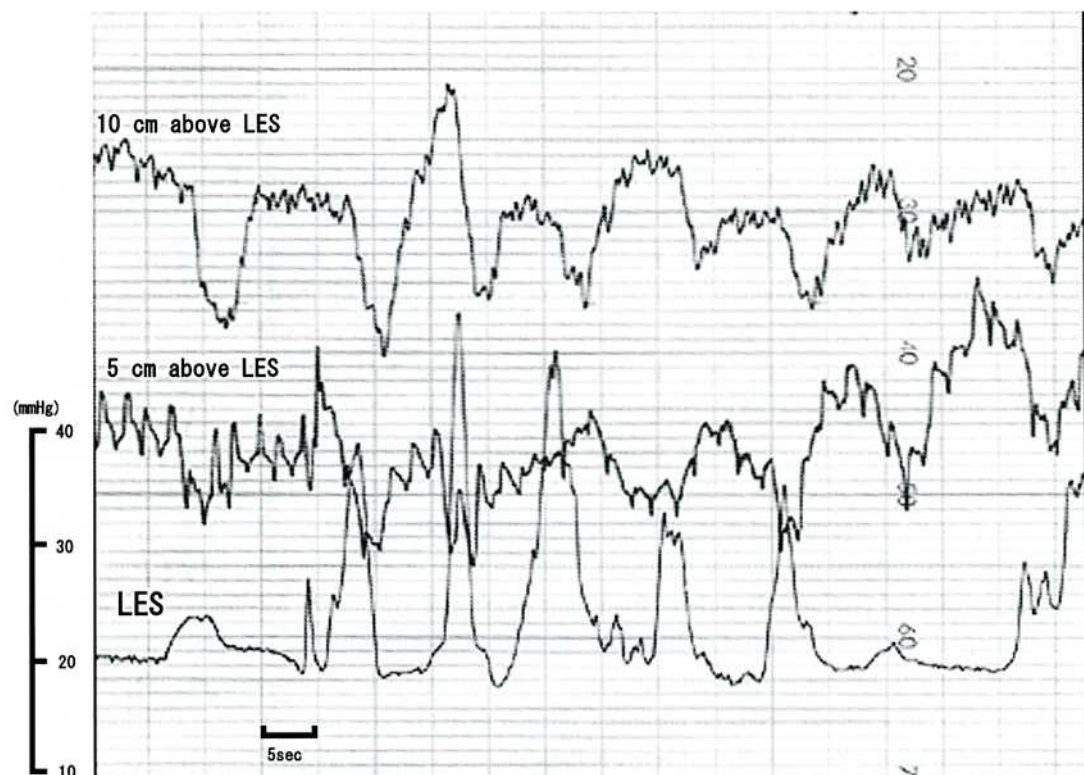


Fig. 2. Manometry revealed characteristic repetitive simultaneous contractions of high amplitude.

Initially, the patient underwent progressive pneumatic dilation therapy. After this forceful dilation of the esophagocardiac junction, the patient's dysphagia disappeared immediately. However, the chest pain continued thereafter. A follow-up esophagram showed that repeated segmental spasms remained. Therefore, an antispasmodic agent, scopolamine butylbromide, was orally administered. Fortunately, the patient was free from chest pain after administration of this antispasmodic. Disappearance of barium stagnation and segmental spasms of the esophagus was confirmed by an esophagram (Fig.1c). Three years after this combined treatment, the patient has demonstrated a good course.

## DISCUSSION

Various reports have described the characteristic radiologic features of achalasia to be esophageal dilatation and distal smooth tapering, a so-called "bird's beak appearance"<sup>3,4</sup>. In vigorous achalasia, uncoordinated simultaneous contractions, which produce multiple ripples or curling of the esophagus, termed a "corkscrew appearance", have also been seen under X-ray fluoroscopy<sup>5</sup>. However, it should be noted that this appearance was recognized only before injection of an antispasmodic agent. The esophageal diameter in vigorous achalasia is smaller than that in classic achalasia. Goldenberg *et al*<sup>6</sup> calculated the ratio of the esophagus measured at its largest caliber to the height of a thoracic vertebrae, and reported that it was less than 1.7 in 71 % of patients with vigorous achalasia. In classic achalasia, the ratio was 1.7 or more in 81 % of the patients. In our patient, the esophagovertebral ratio was 2.2. This high value and negligence in

fluoroscopic observation before administration of an antispasmodic agent may have led to a misdiagnosis in our case, resulting in the patient's long-standing esophageal symptoms.

Manometry is the most effective diagnostic procedure for achalasia. The basal lower esophageal sphincter pressure is normal or elevated, and relaxation following swallowing either does not occur or is reduced in degree, duration, and consistency. In the esophageal body, however, the resting pressure is elevated. Simultaneous contraction is a characteristic manometric finding. Whereas these contractions are of low amplitude in classic achalasia, they are of high amplitude and long duration in vigorous achalasia. However, Todorczuk *et al*<sup>7)</sup> doubted the existence of vigorous achalasia itself as a separate condition, because their clinicomanometric data was statistically similar in both patients with vigorous achalasia and those with nonvigorous achalasia. Nevertheless, we would stress that a different condition from classic achalasia certainly exists, based on the clinical course in our patient.

Achalasia is frequently resistant to medical treatment. Pneumatic dilation is the most effective nonsurgical treatment of achalasia. Eckardt *et al*<sup>8)</sup> analyzed the clinical data in 54 patients with achalasia, and found that patients of 40 years old or more responded better to pneumatic dilation than those of less than 40 years. They also reported that clinical symptoms improved in 36 of 41 symptomatic patients, but that 7 of these 36 patients had continuously suffered from some esophageal symptoms. It is important to determine why esophageal symptoms continued in these seven patients. Unfortunately, detailed clinical information on these seven patients was not provided in their paper. Such unresponsive patients may be, like our patient, ones with vigorous achalasia. Forceful pneumatic dilation would be effective in treating symptoms due to cardiac stenosis, but may be logically ineffective in those due to esophageal spasms.

To date, only three cases of vigorous achalasia have been previously reported in the Japanese literature, excluding those documented in abstracts<sup>9)~15)</sup>. Those cases and our case of vigorous achalasia are summarized in the Table. The ages of the patients ranged from 25 to 72 years old, with the mean age being 55.3 years old. Included our patient, and the female was only 2 cases. All patients complained of dysphagia. Chest pain, which seems to be the most characteristic symptom in vigorous achalasia, developed in half of all

Table. Clinical Characteristics of Patients with Vigorous Achalasia in Japan

Author	(Reported Yr)	Age	Sex	Symptoms	Disease duration** (yr)	Treatments of Vigorous Achalasia
1 Yakabi	(1982)	41	M	Dysphagia, Vomiting	13	Pneumatic dilation
2 Furuya	(1995)	66	M	Dysphagia	60***	ISDN****
3 Yamada	(1996)	72	M	Dysphagia, Weight loss	52	Pneumatic dilation
4 Yoshida	(1999)	61	M	Dysphagia, Vomiting	8	Pneumatic dilation
5* Morizaki	(2002)	49	M	Dysphagia, Chest pain	1	non
6 Yoneyama	(2004)	25	F	Dysphagia, Chest pain	2	Pneumatic dilation, ISDN
7* Hamamoto	(2004)	68	M	Dysphagia, Chest pain	40	Pneumatic dilation
8 Our case		61	F	Dysphagia, Chest pain	32	Pneumatic dilation, anticholinergic

\* Concomitant with esophageal cancer.

\*\* Period during which patients suffered from symptoms till diagnosis.

\*\*\* Approximate value.

\*\*\*\* Isosorbide dinitrate

patients. Two of eight the patients developed esophageal cancer : one had the esophagus removed , and the other had endoscopic resection. The disease duration, which was the period from the appearance of esophageal symptoms to the diagnosis, was average 26 years. Such a long disease duration may present difficulty in making a diagnosis of vigorous achalasia. One patient, who had been suffering from only dysphagia, had been managed with isosorbide dinitrate alone. Three patients, who had complained of vomiting or weight loss in addition to dysphagia, but who had not suffered from chest pain, required pneumatic dilation therapy. Other three patients, who had been suffering from not only dysphagia, but also from chest pain, required the combination of an antispasmodic agent and pneumatic dilation therapy. Therefore, the difference in the therapeutic modality required may be due to the severity of the condition.

## REFERENCES

- 1) Olsen AM, Ellis FH Jr, Creamer B : Cardiospasm (achalasia of the cardia). *Am J Surg* 93 : 299-307, 1957
- 2) Camacho-Lobato L, Katz PO, Eveland J, Vela M, Castell DO : Vigorous achalasia. Original description requires minor change. *J Clin Gastroenterol* 33 : 375-377, 2001
- 3) Stewart ET : Radiographic evaluation of the esophagus and its motor disorders. *Med Clin North Am* 65 : 1173-1194, 1981
- 4) Reynolds JC, Parkman HP : Achalasia. *Gastroenterol Clin North Am* 18 : 223-255, 1989
- 5) Sanderson DR, Ellis FH Jr, Schlegel JF, Olsen AM : Syndrome of vigorous achalasia: clinical and physiological observation. *Dis Chest* 52 : 508-517, 1967
- 6) Goldenberg SP, Burrell M, Fette GG, Vos C, Traube M : Classic and vigorous achalasia: a comparison of manometric, radiographic, and clinical findings. *Gastroenterology* 101 : 743-748, 1991
- 7) Todorczuk JR, Aliperti G, Staiano A, Clouse RE : Reevaluation of manometric criteria for vigorous achalasia. Is this a distinct clinical disorder? *Dig Dis Sci* 36 : 274-278, 1991
- 8) Eckardt VF, Aignherr C, Bernhard G : Predictors of outcome in patients with achalasia treated by pneumatic dilation. *Gastroenterology* 103 : 1732-1738, 1992
- 9) Yakabi K, Izumi T, Ashida E, Saito E, Kitamura T, Mori H, Seki A, Matsuo H, Kosaka J, Takenaka E : A case of vigorous achalasia successfully treated with pneumatic dilatation. *Nippon Shokakibyo Gakkai Zasshi (Jpn J Gastroenterol)* 79 : 872-875, 1982 (in Japanese with English abstract).
- 10) Furuya R, Kunimi M, Ihara F, Ishihara M, Mori K, Yoshiya K, Sakakibara K, Iida K, Nishino M, Yoshitoshi K : A case of vigorous achalasia successfully treated with isosorbide dinitrate spray. *Nippon Shokakibyo Gakkai Zasshi (Jpn J Gastroenterol)* 92 : 1765-1769, 1995 (in Japanese with English abstract)
- 11) Yamada H, Iwakiri K, Yoshizawa M, Kotoyori M, Sugiura T, Futagami S, Nakagawa Y, Yonezawa M, Sueoka N, Kagawa T, Kawakami A, Hirakawa T, Sesoko M, Kobayashi M : Transition from diffuse esophageal spasm to vigorous achalasia. *Shokakinaishikyo No Shinpo (Prog Dig Endosc)* 48 : 89-92, 1996 (in Japanese with English abstract)
- 12) Yoshida T, Nishiaki M, Takeo Y, Harada T, Okita K, Abe T, Hayashi H, Tangoku A, Oka M : A case of vigorous achalasia. *Shokaki No Rinsho* 2 : 98-100, 1999 (in Japanese)
- 13) Morisaki Y, Kondo N, Matsuyama T, Yoshizumi Y, Maehara M : A case of esophageal carcinoma concomitant with vigorous achalasia. *J. Natl. Def. Med. Coll* 27 : 157-162, 2002 (in Japanese with English abstract)
- 14) Yoneyama F, Yamada T, Komori K, Hattori T : Vigorous achalasia with high-amplitude esophageal body contraction: a case report. *Esophagus* 1 : 95-97, 2004
- 15) Hamamoto T, Maeta S, Noguchi M, Mirura N, Ohmura H, Hori T, Tsuruhara I : A case of esophageal early cancer concomitant with vigorous achalasia. *Nippon Shokakibyo Gakkai Zasshi (Jpn J Gastroenterol)* 101: 983-988, 2004 (in Japanese)