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Case Report

Resection of Esophageal Idiopathic Muscular Hypertrophy Complicated by Prominent Cicatricial Stenosis and Mucous Membrane Disorder : A Case Report

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ABSTRACT

A 44-year-old woman had had dysphagia since childhood. Because the symptoms worsened, she underwent diagnostic studies, which demonstrated degeneration of the mucous membrane, distension of the mid-esophagus, and an area of prominent cicatricial stenosis, approximately 4 cm long and of unknown etiology, in the lower part of esophagus. Because the symptoms were resistant to conservative medical treatment, transhiatal esophagectomy and reconstructive surgery were performed. Pathological examination of the resected specimen revealed thickening of the inner circular muscle layer in the lower part of the esophagus, leading to a diagnosis of esophageal idiopathic muscular hypertrophy (IMH). Esophageal IMH is a rare disease of unknown etiology. Most cases are asymptomatic and are diagnosed only at autopsy. Our patient had prominent cicatricial stenosis and mucosal damage in addition to the characteristic changes of esophageal IMH, and other conditions, such as gastroesophageal reflux, were suspected to be present. (Jikeikai Med J 2014 ; 61 : 21-8)

Key words : idiopathic muscular hypertrophy, esophagus, transhiatus esophagectomy

INTRODUCTION

Neoplastic lesions causing esophageal stenosis include malignant tumors, such as esophageal cancer, gastrointestinal stromal tumor, and benign tumors, such as leiomyoma, and neurogenic tumors[1]. On the other hand, esophageal stenosis may develop during the healing process of inflammatory lesions, such as reflux esophagitis and corrosive esophagitis[2].

Idiopathic muscular hypertrophy (IMH) of the esophagus is rarely reported, and its etiology remains poorly understood[3-18]. In addition, most reported cases are found at autopsy, and few cases have been treated with surgical resection. In the present paper we report on a 44-year-old women with esophageal IMH that exhibited prominent cicatricial stenosis and mucosal damage and was treated with surgical resection.

CASE REPORT

The patient was a 44-year-old woman whose chief complaints were dysphagia and vomiting. The patient had been noted to vomit milk for several months after birth and continued to have difficulty swallowing as an adult but had otherwise been well. To deal with this problem, she adjusted the amount of oral intake and types of meals. She

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consulted a physician at the age of 40 years, and an abnormal shadow in the lower esophagus was found with an upper gastrointestinal series. Because symptoms worsened, the patient consulted another physician at the age of 41 years. Esophageal achalasia was suspected. She underwent endoscopic dilatation, but her symptoms continued to worsen. Therefore, she was referred to our hospital for further evaluation and treatment.

The findings of physical examination were as follows : height : 149 cm ; weight : 38 kg ; body-mass index : 17.1 kg/m^2 ; body temperature : 36.8°C ; blood pressure : 108/60 mm Hg ; and heart rate : 64 beats per minute. The abdomen was flat, soft, and without apparent abnormalities.

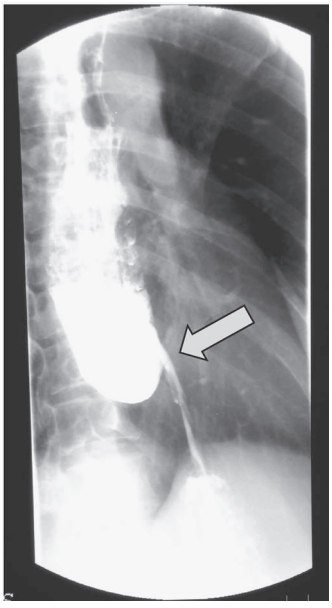

Fig. 1. Upper gastrointestinal series
Prominent stricture, approximately 5 cm in length with orad distension, was noted in the lower part of the esophagus (arrow).

Laboratory findings

An upper gastrointestinal series demonstrated a prominent 5-cm-long stricture of the lower esophagus oral to the esophagogastric junction, with orad distension. The contrast medium was poorly retained and slowly entered the stomach through the stricture (Fig. 1).

Upper gastrointestinal endoscopy revealed prominent cicatricial stenosis 32 to 36 cm from the incisors. Although an endoscope with an external diameter of 9 mm could not be passed through the stricture, a nasal endoscope with an external diameter of 5 mm could. Moreover, repeated areas of white scar-like degeneration of the mucous membrane, resembling the veins of a leaf, were observed from 20 cm from the incisors to the stricture. Findings of hiatus esophageal hernia and reflux esophagitis were not observed (Fig. 2).

A thoracoabdominal computed tomographic scan demonstrated a 4-cm-long stricture in the lower esophagus and proximal distension. Swelling of the mediastinal or intra-abdominal lymph nodes was not observed (Fig. 3).

Although esophageal manometry was performed with multichannel impedance pH monitoring to evaluate the possibility of esophageal achalasia and reflux esophagitis, the internal pressure catheter was unable to be passed through the stricture, and, therefore, esophageal motor function and the lower esophagus sphincter muscle could not be evaluated. In the distended part of the esophagus, no atypical constrictions or spasms were observed. Moreover, the

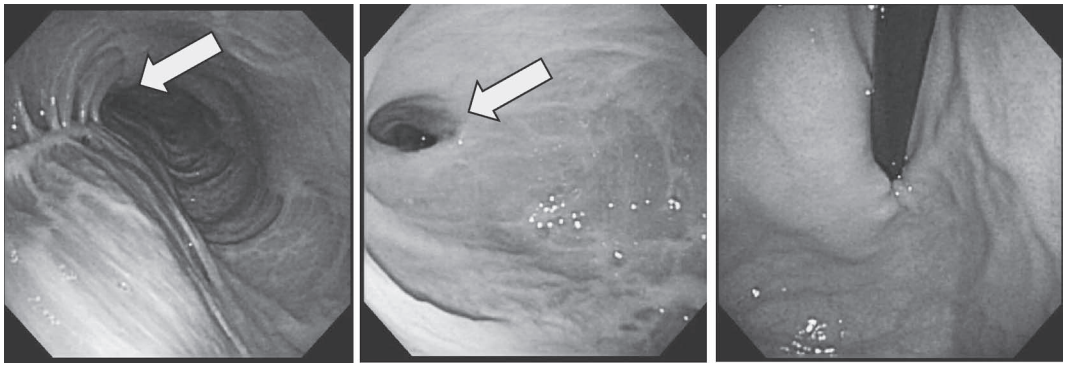

Fig. 2. Upper gastrointestinal endoscopy
Repeated areas of scar-like degeneration of the mucous membrane, resembling the veins of a leaf, were observed 20 cm from the incisor. Prominent stricture was observed 32 to 36 cm from the incisors (arrows). Neither hiatus esophageal hernia nor reflux esophagitis was observed.

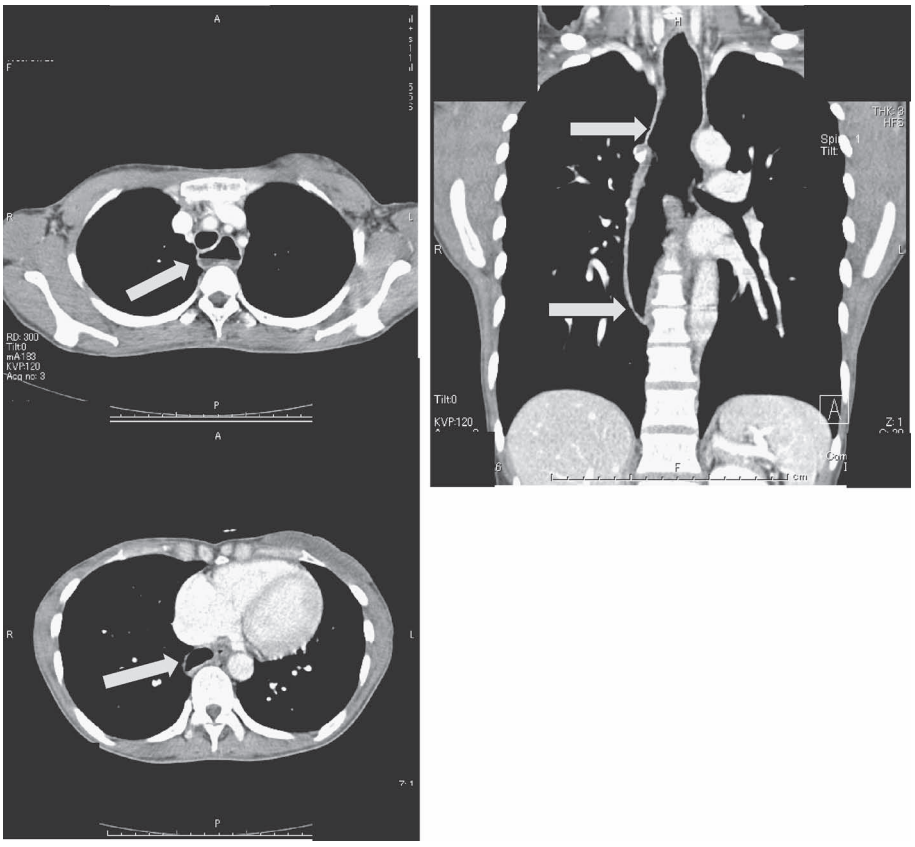

Fig. 3. Thoracicoabdominal computed tomographic scan
Slight wall thickening (arrows) was observed in the lower part of the esophagus with proximal distension.

number of episodes of reflux into the esophagus was 11 within 24 hours, which was within the normal range.

Course of treatment

The morphological changes in the esophagus seen on upper gastrointestinal endoscopy/contrast radiography are shown in Fig. 4. The scar from the upper part of the chest to the central part of the esophagus seen on upper gastrointestinal endoscopy resembled that after burns or after treatment for corrosive esophagitis. A slightly normal esophageal mucosa was observed in the abdominal esophagus, and the esophagocardiac junction and the squamocolumnar junction were in the same location.

The cause of esophageal stenosis was investigated on the basis of these findings, but no cause could be identified. The patient had dysphagia at the time of the first visit ; however, because she had had similar symptoms for many years, she did not desire invasive treatment. The symptoms did not improve thereafter, and the vomiting worsened after she had visited the hospital as an outpatient for approximately 6 months. Therefore, esophagectomy and reconstruction surgery were performed under laparoscopic assistance for diagnostic treatment.

Intraoperative findings

Surgery was performed with the patient in the supine position under general anesthesia. In addition to the camera port in the lower abdomen, 5-mm ports were inserted in both the left and right sides of the abdomen, and a 12-mm port was inserted in the left hypochondriac area. The esophageal hiatus was opened, a camera was inserted into the mediastinum, and the mediastinal esophagus was detached from the surrounding tissue. Periesophageal adhesion was severe near the stricture, and the esophagus was freed cephalad and resected. Reconstruction was performed with a gastric tube of the greater curvature side, which was lifted through the retrosternal route and anastomosed with the cervical esophagus by means of the triangle

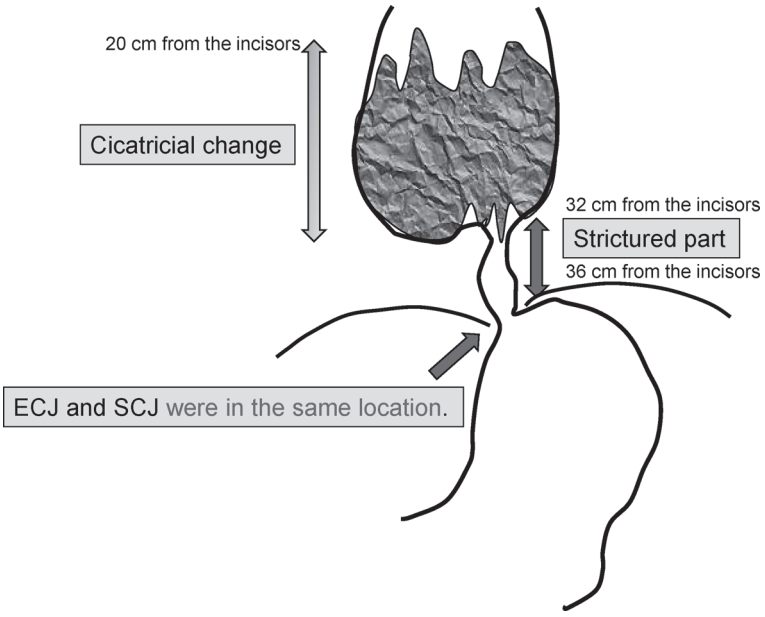

Fig. 4. The morphologic schema of the esophagus in the present case.

method. The operation time was 419 minutes, and blood loss was 230 g.

Postoperative course

Delivery of enteral nutrition was started the day after surgery, and oral intake was started 7 days after surgery. No major postoperative complications developed, and the patient was discharged from the hospital 27 days after surgery. As of 12 months after surgery, the patient has good oral intake and no difficulty in swallowing and has gained 5 kg.

Gross examination of the resected specimen (Fig. 5) showed no apparent abnormalities in the stomach, including the cardiac orifice. An area of severe cicatricial stenosis approximately 4 cm in length (circumference : 27 mm) was observed from the lower thoracic esophagus to the abdominal esophagus. The lower part of the esophagus was distended in a spindle-shaped structure 10 cm in length, including the stricture, and had a maximal circumference of 75 mm, which was 48 mm greater than that of the area of the most severe stenosis. Moreover, the mucous membrane was serrated and had ruptured, with the muscle layer exposed over a wide range, and was accompanied by prominent hypertrophy of the lower part of the esophagus (most thickened part : 12 mm ; normal part : 4 mm).

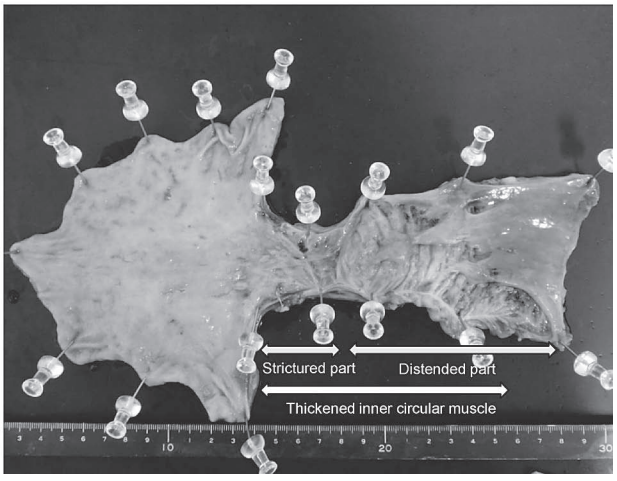

Fig. 5. Gross findings of the resected specimen
The stricture of the esophagus was most extensive on the oral side approximately 50 mm from the esophagogastric junction with oral distension producing a spindle-shaped structure with prominent thickening of the esophageal wall.

Histopathologic findings

In addition to the areas of stricture and expansion in the esophagus, fibrosis was diffusely observed within the mucosa, the submucosa, between bundles of muscle fibers, and in the adventitia. Fibrosis had also spread to the wall of the gastric cardia. Muscle hypertrophy was observed over a wide area, approximately 10 cm from the oral side of

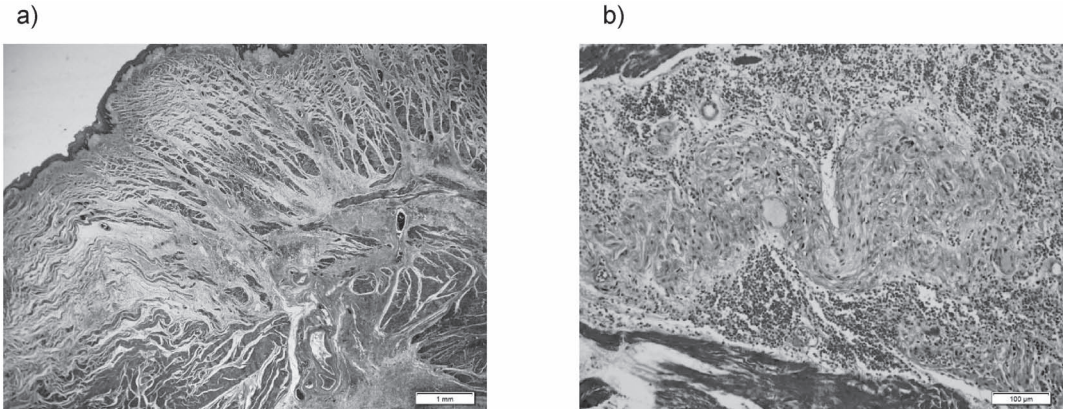

Fig. 6. Histopathologic findings
(a) Thickening of the muscles was prominently centered on the inner circular muscle layer of the esophagus. Masson staining (×20)
(b) Degeneration of Auerbach's plexus and ganglion cells was not observed. Masson staining (×400)

the stricture, including the distended part. The stratified squamous epithelium had thinned, and the normal structure of the wall of the esophagus had been disrupted. The outer longitudinal muscle layer was also thickened, but the thickening of the inner circular muscle layer was significant (Fig. 6a).

The structure of the wall of the mucous membrane and submucosa of the upper intrathoracic esophagus was preserved, but the muscular layer of the mucosa was thickened.

Neither inflammatory cell permeation nor degeneration was observed in the stomach or esophagus. Regarding Auerbach's plexus, distribution density was obtained, ganglion cells were present as normal, and achalasia was ruled out by histologic findings (Fig. 6b).

On the basis of the medical history, clinical course, and surgical and pathological findings, IMH of the esophagus was diagnosed accompanying other causes in the past such as reflux of the acidic digestive fluids.

Discussion

Esophageal IMH is a rare condition characterized by hypertrophy of the muscles of the lower part of the esophagus ; no consensus has yet been reached on its pathogenesis[3-18]. Previously reported cases and the present case of IMH of the digestive tract are shown in Table 1. Of the 62 cases, 24 cases were asymptomatic and were incidentally diagnosed at autopsy. Fifty-eight cases arose in the esophagus, with 8 of these being in neonates or children 15 years or younger[19-22]. Moreover, muscle hypertrophy in the pylorus was observed in 8 cases. Of the 49 cases for which sex was described, 49 (82%) occurred in males.

Various causes of esophageal IMH have been proposed, but the etiology remains unknown. Although inflammation may trigger IMH, such changes may be secondary to the stricture due to muscle hypertrophy[3-18]. On the other hand, Guthrie et al. reported on an 11-year-old girl who had multiple IMH lesions in the esophagus, pylorus, duodenum, and jejunum which suggested the possibility of congenital muscle hypertrophy[19]. Although symptoms in our patient had been present for many years, a detailed investigation was not performed until she was an adult. However, because of the episodes of vomiting in the neonatal period and infancy, congenital muscle hypertrophy cannot be ruled out.

In most cases of esophageal IMH, endoscopic examination shows narrowing of the lumen of the lower part of the esophagus and fusiform or cylindrical orad distension of the esophagus. Because similar anatomical changes were observed in the present case, they might be characteristic of esophageal IMH. On the other hand, the esophageal mucous membrane in IMH has not been investigated in detail. The scars and fibrosis, resembling the veins of a leaf, observed in the esophageal mucous membrane of the present case were similar to those in corrosive esophagitis, but there have been no such reports to date. Accordingly, it is possible that the mucosal damage in the present case was caused by accompanying conditions, such as the reflux of

Table 1. 62 cases of idiopathic muscular hypertrophy.

No.	Author (Year)	Age (yrs)	Sex	Location	Symptoms	Remarks	Treatment
1	Reher (1885)	60	M	Esophagus	+	Rectal carcinoma	−
2	Pitt (1888)	68	F	Esophagus	N.D	Brain hemorrhage	−
3	Rolleston (1899)	59	M	Esophagus	−	Fracture of ribs	−
4	Elliesen (1903)	39	M	Esophagus	−	Tuberculosis, meningitis	−
5	Ehlers (1907)	56	M	Esophagus, Pylorus	−	Pneumonia	−
6	Mollison (1919)	62	N.D	Esophagus	N.D	−	−
7	Pritchart (1920)	0	M	Esophagus, Pylorus, Ileo-cecal	N.D	Pneumonia	−
8	Rake (1926)	67	M	Esophagus	−	Pneumonia	−
9	Von Brucke (1928)	67	M	Esophagus	+	Pneumonia	−
10		63	M	Esophagus	−	Hemiplegia	−
11	Goedel (1929)	39	M	Esophagus	−	−	−
12	Wood (1932)	69	M	Esophagus	−	Colitis	−
13	Rossle (1935)	67	M	Pylorus, Jejunum	N.D	−	−
14		27	F	Antrum, Ileum	+	Tuberculosis	−
15		N.D	M	Esophagus, Pylorus	N.D	−	−
16		57	M	Esophagus	N.D	Hernia	−
17	Helmke (1939)	41	M	Esophagus	−	Myoma	−
18		57	M	Esophagus	−	Myoma	−
19		64	M	Esophagus	−	Myoma	−
20		59	M	Esophagus, Pylorus	−	Myoma	−
21	Guthrie (1945)	11	F	Esophagus, Pylorus Duodenum, Jejunum	−	Nephritis	−
22	Schroeder (1949)	74	M	Stomach	+	Myocardial infarct	−
23	Sloper (1954)	53	M	Esophagus	−	Lung carcinoma	−
24		31	F	Esophagus	−	Thymus carcinoma	−
25		58	M	Esophagus	−	Sarcoma(pelvis, mediastinal)	−
26		60	M	Esophagus, Pylorus	−	Glioblastoma	−
27		41	M	Esophagus	+	Tuberculosis	Vagotomy
28		63	M	Esophagus	−	Rectal carcinoma	−
29		47	M	Esophagus	−	Cecal carcinoma	−
30	Marston (1959)	51	M	Esophagus	+	−	Myotomy
31	Spencer (1961)	3	M	Esophagus, Pylorus Duodenum, Jejunum	+	Microcephaly	−
32	Uhrich (1965)	0	M	Esophagus, Stomach, Ileum	+	Cerebral damage	Gastrectomy
33	Cocker (1966)	0	N.D	Esophagus, Pylorus	N.D	Cardiomegaly	−
34	Wall (1967)	29	F	Esophagus	+	Leiomyoma of esophagus	Subtotal esophagectomy
35 ～ 48	Ferguson (1969) 14 cases	Average 52	M : 11 F : 3	Esophagus : 14	+ : 14	−	Myotomy
49	Peison (1971)	64	M	Esophagus	+	Amiotrophic lateral sclerosis	−
50 ～ 55	Demian (1978) 6 cases	Average 64	M : 6	Esophagus : 6	− : 5 + : 1	−	−
56	Zeller(1979)	68	F	Esophagus	+	−	−
57	Yamagiwa(1988)	0	M	Jejunum, Ileum	+	Sclerocornea, Cryptochidism	Small intestine resection
58	Legius (1990)	7	M	Esophagus	+	Alport syndrome	Esophagectomy
59	Grabowski(1996)	4	F	Esophagus	+	−	Esophagectomy
60		15	M	Esophagus	+	−	Balloon dilatation (Repeated)
61	Shimada (2003)	60	M	Esophagus	−	Esophageal carcinoma	Subtotal esophagectomy
62	Yuda (2013)	44	F	Esophagus	+	−	Transhiatal esophagectomy

M : Male F : Female N.D : not described.

acidic digestive fluids. Shimada et al. in 2003 reported a case of esophageal IMH in which superficial esophageal cancer was observed as a complication, and endoscopic ultrasonography was used to confirm thickening of the inner muscle layer[23]. Perhaps if endoscopic ultrasonography had been performed in the present case, IMH, which is characterized by thickening of the inner muscle, might have been diagnosed before surgery.

In 1988, Agostini et al. reported computed tomographic findings of esophageal IMH[24]. The characteristics findings were diffuse thickening of the esophageal wall, the presence of nodules, and distension of the esophagus on the oral side but did not, however, allow IMH to be differentiated from other diseases, such as esophageal cancer and achalasia.

In the present case, the histopathologic finding most characteristic of esophageal IMH was the prominent thickening of the inner circular muscle layer, which extended approximately 10 cm from the oral side of the stricture, including the distended portion. Degeneration of Auerbach's plexus or ganglion cells is not observed in cases of IMH, including the present case, but is a pathological finding of achalasia.

Regarding treatment, Ferguson et al. reported on outcomes of Heller myotomy in 112 cases of esophageal motor disorders, including 14 cases of IMH. Symptomatic improvement was observed in 12 of 13 cases that could be followed up ; however, in 1 case dysphagia did not improve after surgery, and the muscles of the upper part of the esophagus were also resected[25]. Moreover, Shimada et al. performed subtotal resection of the esophagus as a treatment for superficial esophageal cancer as a complication of esophageal IMH[22]. Although a definitive diagnosis was not obtained before surgery in the present case, surgical resection was indicated because the dysphagia and vomiting were becoming more frequent and were accompanied by a severe stricture through which the endoscope fiber could not be passed. Regarding the indications for surgery in patients with esophageal IMH, previous reports suggest : a) Heller myotomy is indicated if the muscle hypertrophy is localized, b) esophagectomy is indicated for superimposed cancer or a suspicion thereof, and c) transhiatal esophagectomy is indicated if muscle hypertrophy extends from the abdominal esophagus to the thoracic esophagus in combination with severe stricture.

Conclusion

We have reported a case of esophageal IMH accompanied prominent by cicatricial stenosis and mucosal damage in which surgical resection was successfully performed.

Authors have no conflict of interest.

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