

## A Case of Superficial Esophageal Cancer Complicated with Idiopathic Muscular Hypertrophy of the Esophagus

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We hereby reported a case of 60-year old man with superficial esophageal cancer complicated with idiopathic muscular hypertrophy of the esophagus. Endoscopic ultrasonography and CT showed the thickness of esophageal muscular layer, but the accurate diagnosis could not be entertained before operation. Idiopathic muscular hypertrophy of the esophagus is an entity rarely encountered, and most cases are diagnosed at postmortem examination. Only a few cases have been reported regarding its clinical symptoms and images. The etiology remains to be elucidated, and the pathologic features are characterized by significant thickness of inner circular muscular layer of esophagus without degeneration of plexus and ganglionic cells. This case report deals with superficial esophageal cancer complicated with idiopathic muscular hypertrophy of esophagus.

Literature review is also included.

**Key words :** idiopathic muscular hypertrophy, esophagus, cancer, achalasia

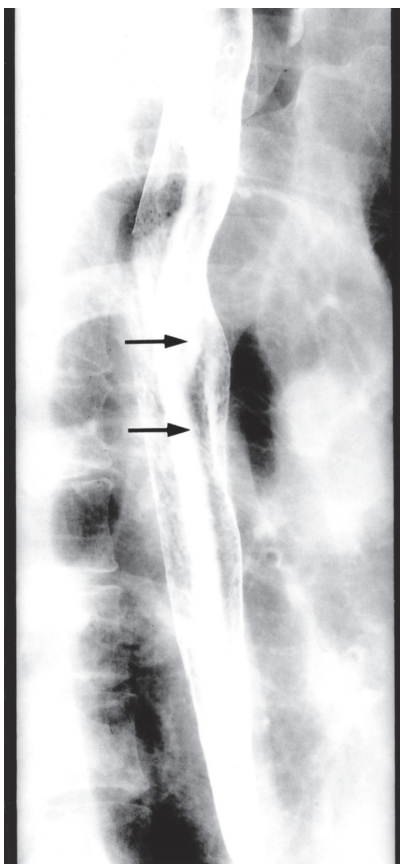
### INTRODUCTION

We report a case of superficial esophageal cancer which developed in esophagus with idiopathic muscular hypertrophy of the esophagus (IMHE), showing significant hypertrophy of muscular layer of entire length of esophagus from neck to abdomen. IMHE is a rare disease and most reported cases have been diagnosed at autopsy, and the description of its clinical variables have been sparse. The etiology of this disease is not clearly elucidated, and the pathologic features are characterized by a significant muscular hypertrophy of inner circular muscular layer without degeneration of plexus of lower esophagus and of ganglionic cells from our literature review. This case is the first case of esophageal cancer complicated with IMHE, as we could review.

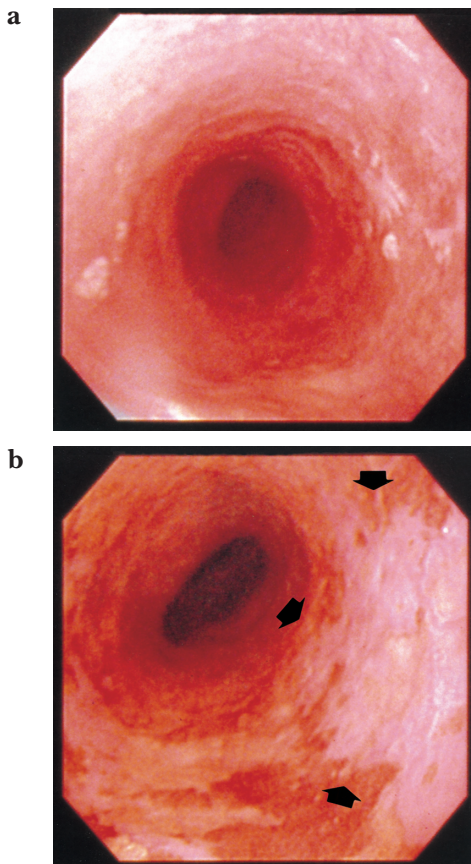
### CASE REPORT

A 60-year old man was pointed out to have abnormality on the esophageal mucosa by endoscopy at clinical examination in March 1996. The patient had been under medications for hypertension, otherwise no past history of particular diseases and no particular family history. He was daily consuming 3 quarter liter of alcohol and 60 cigarettes for 38 years. Laboratory data were unremarkable. The upper GI series at admission revealed a granular change of total esophageal mucosa and the stenotic change towards the distal part. But the passage of barium meal was unremarkable (Fig. 1), and the lesion depicting cancer was not clearly demonstrated.

Endoscopy showed rough mucosa of entire esophagus, and also a shallow depression with tiny white granular changes starting at 30 cm from incisor. Iodine staining revealed an un-



**Fig. 1** Esophagogram showed a granular change of mucosa and stenotic change towards the distal part. The passage of barium meal was unremarkable.



**Fig. 2a, b** Endoscopic examination showed shallow depression with tiny granular change by conventional observation.(2a) Iodine staining revealed an unstained lesion.(2b)

stained lesion occupying half of the lumen at 30 to 33 cm from incisor, for which diagnosis of 0-IIc was made. Other tiny unstained lesions were also found scattered in the anal portion and stenotic change was observed towards the lower esophagus (Fig. 2a, b).

Endoscopic ultrasonography showed hypertrophy of smooth muscular layer in the entire esophagus, as shown also by chest CT (Fig. 3, Fig. 4).

Preoperative diagnosis of esophageal carcinoma was type 0-IIc, with the depth of m3. And the cause of esophageal muscular hypertrophy could not be clarified. Subtotal esophagectomy was carried out by thoracotomy for esophageal cancer and reconstruction was made by gastric tube.

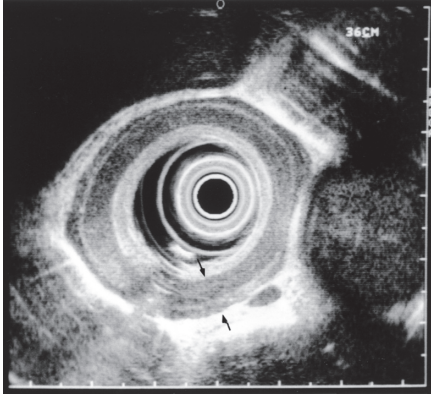
At operation, esophagus was extremely thick, which was verified at the oral edge after

intrathoracic dissection of esophagus. Insertion of the head of EEA to cervical esophagus for automatic suture was considered to be risky due to the thick esophagus, and manual layer to layer anastomosis between the gastric tube and cervical esophagus was performed. Postoperative course was uneventful and oral feeding was possible at the 14<sup>th</sup> postoperative day, taking the standard course, and no disturbance of alimentary passage by IMHE was encountered. And the patient was discharged at the 29<sup>th</sup> postoperative day.

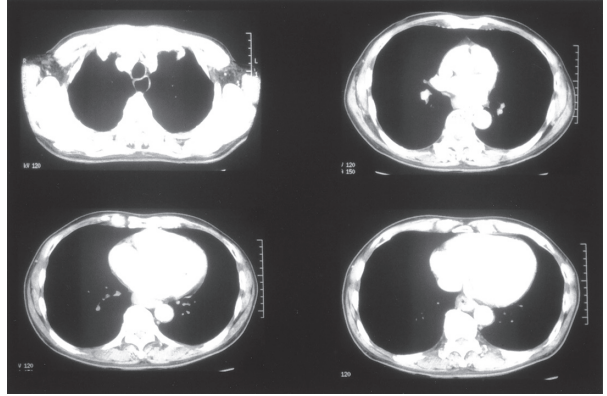
#### **PATHOLOGIC FINDINGS**

##### **Macroscopic findings**

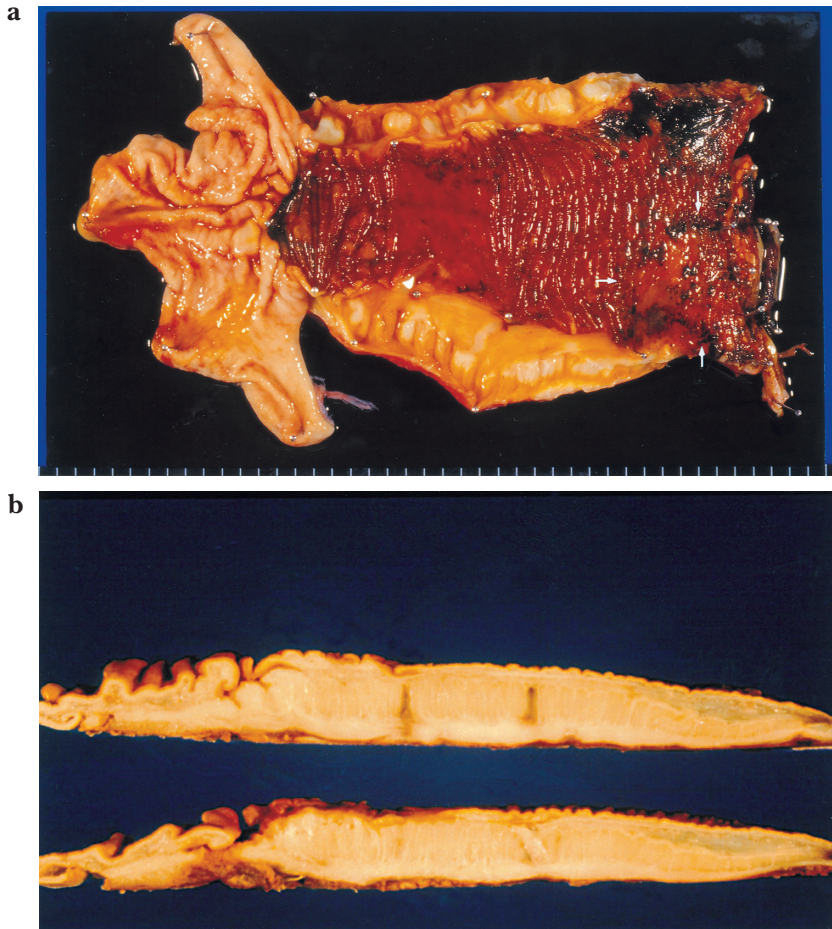
Removed esophagus was significantly hypertrophic in the inner circular muscular layer, and longitudinal sections along the esophagus showed the gradual decrease of hypertrophy



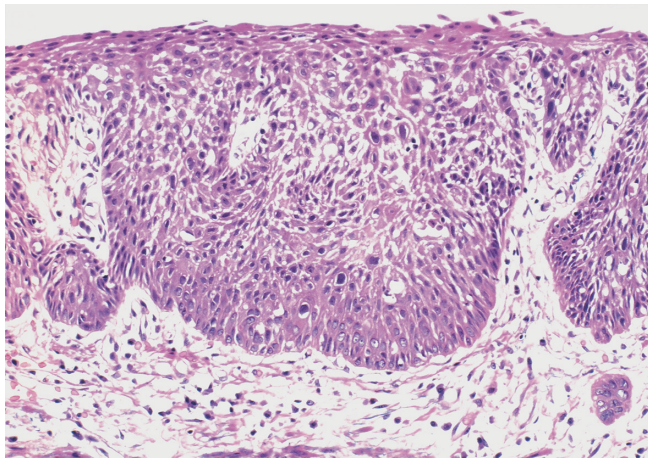
**Fig. 3** Endoscopic ultrasonography revealed hypertrophy of muscular layer.



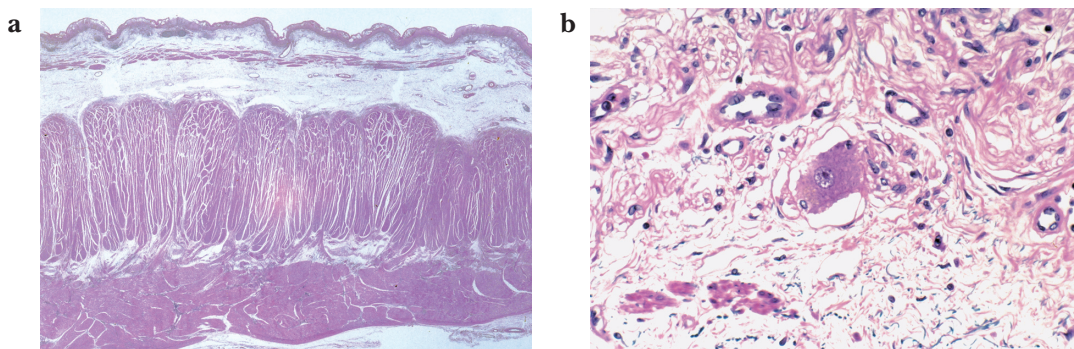
**Fig. 4** Chest CT scan revealed thickness of entire esophagus.



**Fig. 5a, b** Removed esophagus was significantly hypertrophic in the inner circular muscular layer (5a). Longitudinal section along the esophagus showed the gradual increase in hypertrophy towards oral direction (5b).



**Fig. 6** Microscopic findings of well differentiated squamous cell carcinoma, of m2. (Hematoxylin and Eosin  $\times$  200)



**Fig. 7a, b** Longitudinal section of esophagus shows hypertrophy of muscularis. (Hematoxylin and Eosin  $\times$  4) (7a)  
Microscopically, no degeneration of the plexus myentericus is noted. (Hematoxylin and Eosin  $\times$  300) (7b)

toward oral direction. The esophageal cancer was recognized as tiny granular change on shallow depressed area, which was recognized more clearly as unstained area by iodine staining. The thickness of the esophageal wall was mostly attributed to the hypertrophy of inner circular muscular layer, 15mm in maximum thickness which was three times thicker than the standard (Fig. 5a, b).

#### **Microscopic findings**

The esophageal cancer was well differentiated squamous cell carcinoma, depth of m2, without vascular or lymphatic invasions (Fig. 6). The hypertrophic areas of inner circular mus-

cular layer showed diffuse hypertrophy of individual smooth muscle cells. Pathology revealed no degeneration of muscular layer and plexus, ganglionic cells, different from esophageal achalasia, and diagnosis was made as IMHE (Fig. 7a, b).

#### **DISCUSSION**

IMHE was first described by Baillie [1] in 1799, and later reviewed by Woods [2] of 8 cases by adding one of his case in 1932. Then Sloper [3] reviewed 32 cases in 1954 including reported 25 cases between 1799 and 1948 and his 7 cases. This review included only one alive case, and the rest was made diagnosis at

**Table 1** Thickness of esophageal wall and muscularis propria twenty cases of resected esophageal specimen.

	Esophageal wall (mm)	muscularis propria inner circular (mm)	muscularis propria outer longitudinal (mm)
oral side	3.7 + 1.1	1.1 + 0.4	1.0 + 0.3
middle portion	3.6 + 0.7	1.0 + 0.2	0.8 + 0.2
anal side	3.6 + 0.8	1.1 + 0.2	1.0 + 0.2

(n = 20)

autopsy. Some IMHE patients were detected after causing spontaneous esophageal perforation, one of them found at autopsy reported by Stephen [4], and 2 cases out of 5 found also at autopsy reported by Wayne [5]. Thus, most cases are diagnosed at postmortem examination, and the reports describing symptoms and images have been very sparse. So we hereby reported one case and reviewed the clinicopathological feature of IMHE of 26 reported cases depicting clinical signs and their images [6-10].

The age ranged from 2 to 68 years (median age: 55), including 21 males and 6 females. The principle symptoms are dysphagia and poststernal pain, caused by esophageal stenosis and diffuse spasm. The patients included 25 symptomatic and 2 asymptomatic patients (one case causing spontaneous esophageal perforation and the present case).

The features of esophagography, endoscopy and chest CT were reviewed from the previous reports. The esophagography shows spasm and contraction or stenosis of esophagus according to 25 cases with description. The endoscopy shows also stenosis and mild distension of esophagus according to 25 described cases. The first description of chest CT on IMHE was by Agostini *et al.* [11] advocating diffuse thickening of esophageal wall and large nodules, but only esophageal thickening was found in the present case and three other reported cases.

The pathological features of IMHE include significant thickness of inner circular muscular layer without abnormal degeneration of muscle fiber. The present case showed also significant thick inner circular and outer longitudinal muscular layer of 7.5 mm and 3.5 mm, respectively, in thickness, while the standard total esophageal wall is thinner than

4 mm, containing 1 mm of inner circular and 1 mm of outer longitudinal muscular layer at the oral edge, middle portion, anal side of fixed esophagus from 20 cases of esophageal cancer. This comparison indicates the significant hypertrophy of muscular layer in IMHE (Table 1). The absence of degeneration in Auerbach plexus and ganglionic cells is the difference from esophageal achalasia. The lymphocytes migration around ganglionic cells and eosinophils and lymphocytes migration to intermuscular septa are also the features of IMHE [12].

The cause of IMHE is not yet clearly known. Two cases of 2 year and 7 year-old patients may suggest genetic factor. The abnormality of neural function is one possible etiology when the spasm and contraction of esophagus are taken into consideration although pathology shows no abnormality in plexus and ganglionic cells.

As a treatment, Thomas [15] reports the efficacy of 'long esophagomyotomy through the left side of chest' in 14 cases of 'giant muscular hypertrophy' among 112 cases of motor dysfunction undergoing Heller's myotomy. The treatment of choice to IMHE cases with severe dysphagia seems to be myotomy. The indications of esophagectomy for IMHE have been already reported to include the following occasions, 1) the malignant lesion cannot be ruled out. 2) myotomy was tried but not effective. 3) accompanying lesion exists on the esophagus besides IMHE.

To our knowledge, 22 cases who underwent surgical operation included 7 cases of esophagectomy in fact, indicated in 2 cases that could not be ruled out malignant lesion on morphological diagnosis, and in 3 cases where Heller's myotomy was not effective, and in one case of

spontaneous esophageal perforation. Our case was indicated esophagectomy for esophageal cancer and co-existence of IMHE was found incidentally.

Achalasia is an idiopathic esophageal motility disorder, known as a risk of squamous cell dysplasia and cancer. It is characterized by a non-relaxing lower esophageal sphincter and lack of peristalsis, causing food stasis, which makes esophageal epithelium inflamed and acanthotic. The present case is out of its category, as shown by no passage disturbance to the stomach and no evidence of acanthosis on esophageal mucosa. The only risks considered for esophageal cancer in this patient are heavy consumption of alcohol and cigarettes and old age and male sex. Complication of malignant tumors of other organs with IMHE have been rarely reported, but we emphasize this is the first report as esophageal cancer complicated with IMHE. The relation of esophageal cancer and IMHE is not known, but we consider the present case as an incidental coexistence of these two disorders.

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