

Original Article

Elastofibromatous change of the intestine: report of four lesions from three patients with review of the literature

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Abstract: Elastofibromatous change, also referred to as elastofibromatous polyp or elastofibroma, has been extremely rarely described in the gastrointestinal tract. This lesion is characterized histopathologically by an excessive accumulation of elastic fibers occasionally with a fibrous component involving the submucosa and/or muscularis mucosae of the gastrointestinal tract. Herein, we report four additional lesions of the intestine and review the clinicopathological features of this rare lesion. Three patients (76-, 72-, and 52-year-old males) were detected with polypoid lesions in the jejunum, transverse and sigmoid colons, and sigmoid colon, respectively. All four lesions showed fundamentally the same histopathological and immunohistochemical features. The polypoid lesions were covered by non-neoplastic epithelium, and degenerated and truncated elastic fibers occasionally with a fibrous component had accumulated in the submucosa and/or muscularis mucosae. The characteristic feature was the elastofibromatous change centered around collections of elastotic submucosal vessels. Desmin-positive degenerative ruptured smooth muscle fibers were scattered within the elastic fibers in the submucosa. Our analyses of the clinicopathological features of the previously reported 32 cases of elastofibromatous change of the gastrointestinal tract as well as the present cases demonstrated that this type of lesion is most commonly found in the colon or rectum (29 cases), males, and middle-aged to elderly persons. Although the pathogenesis remains unclear, the convincing hypothesis that this lesion represents elastic degeneration of submucosal vessels by previous persistent vascular injury has been proposed. The collections of degenerative elastotic vascular walls may have an important role in the development of this lesion.

Keywords: Elastofibromatous change, elastofibromatous polyp, elastofibroma, intestine

Introduction

Elastofibromatous change, also referred to as elastofibromatous polyp or elastofibroma, has been extremely rarely described in the gastrointestinal tract [1-9]. This change was first reported by Enjoji *et al.* in 1985, and since then, only a limited number of cases have been described in the literature [1-9]. This lesion is characterized histopathologically by an excessive accumulation of fine granular and/or fibrillar elastic fibers occasionally with a fibrous component involving the submucosa and/or muscularis mucosae of the gastrointestinal tract [8, 9]. Little attention has been paid to this lesion because of obvious benignity, and the cause and histogenesis remain unclear. Herein, we report four additional lesions of elastofibromatous change of the intestine (one

from the small intestine and three from the large intestine) and review the clinicopathological features of this rare lesion.

Case reports

Clinical presentations

Case 1: A 76-year-old Japanese male presented with persistent black stool at an outpatient clinic. A fecal occult blood test was positive, however, esophagogastrosopic and colonoscopic examinations failed to detect any tumorous lesions. Thus, a capsule endoscopic examination was performed, which revealed a small polypoid lesion in the jejunum. Subsequently, he underwent a small intestinal endoscopic examination, which demonstrated a polypoid lesion, measuring 4 x 3 mm in diameter, with a

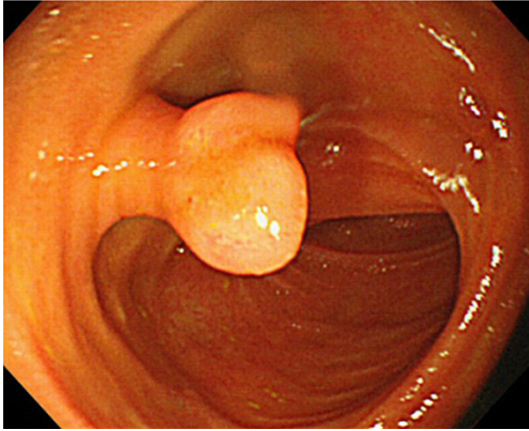


Figure 1. Endoscopic feature of Case 1. A small polypoid lesion with a central depression in the jejunum.

central depression in the jejunum (**Figure 1**). Endoscopic mucosal resection of the polyp was performed.

Case 2: A 70-year-old Japanese male was incidentally found to have three colon polyps by medical check-up. One was present in the transverse colon, measuring 3 mm in diameter, one was in the sigmoid colon, measuring 1.5 mm in diameter, and the remaining lesion was in the rectum, measuring 10 mm in diameter. Endoscopic mucosal resections were performed for all three polyps. The rectal polyp was a tubular adenoma.

Case 3: A 52-year-old Japanese male was incidentally found to have a small sessile polypoid lesion, measuring 0.5 x 1 mm in diameter, in the sigmoid colon by medical check-up. A biopsy of the polypoid lesion was performed.

Histopathological and immunohistochemical features

Case 1: Histopathological study revealed a polypoid lesion with central depression (**Figure 2A**). The surface of the polyp was covered by non-neoplastic small intestinal mucosa, and deposition of slightly eosinophilic to amphophilic fibrillar material was observed in the submucosa (**Figure 2A**). The arterial wall was degenerated and medial elastic fibers were also degenerated (**Figure 2B**). No deposition was observed in the muscularis mucosae (**Figure 2A**). Inflammatory cell infiltration was scant in both the muscularis mucosae and submucosa (**Figure 2A**). Elastica van Gieson staining clearly

demonstrated that these slightly eosinophilic to amphophilic materials were stained brownish to black, which indicated that they were degenerative elastic fibers (**Figure 2C, 2D**). These elastic fibers were randomly distributed and truncated (**Figure 2D**). The elastic fibers of the tunica media of the arterial wall in the submucosa were also degenerated (**Figure 2D**). No amyloid deposition was noted by amyloid staining.

Immunohistochemical studies were performed using an autostainer (Ventana) by the same method as previously reported [10-14]. The immunostaining for desmin revealed that degenerative ruptured smooth muscle fibers were scattered within the elastic fibers in the submucosa (**Figure 2E**).

Cases 2 and 3: The histopathological characteristics of the three polypoid lesions of the colon found in Cases 2 and 3 were fundamentally the same. The surface colon mucosa was slightly hyperplastic, however, no neoplastic change was noted (**Figures 3A and 4A**). Deposition of slightly eosinophilic to amphophilic fibrillar material was observed in the submucosa (**Figures 3A, 3B and 4A**). The arterial wall was degenerated and medial elastic fibers were also degenerated (**Figure 2B**). The fibrillar material was also deposited in the muscularis mucosae in the lesion of Case 3 (**Figure 4A**). Inflammatory cell infiltration was scant both in the muscularis mucosae and submucosa (**Figures 3A, 3B and 4A**). Elastica van Gieson staining clearly demonstrated that these slightly eosinophilic to amphophilic materials were stained brownish to black, which indicated that they were degenerative elastic fibers (**Figures 3C, 4B**). These elastic fibers were randomly distributed and truncated (**Figures 3C and 4B**). Mild fibrosclerosis was also observed in the submucosa, which was stained red by elastic van Gieson staining (**Figure 3C**). No amyloid deposition was noted by amyloid staining.

The immunostaining for desmin revealed that degenerative ruptured smooth muscle fibers were scattered within the elastic fibers in the submucosa (**Figures 3D and 4C**).

Accordingly, an ultimate diagnosis of elastofibromatous change of the intestine was made for all four lesions.

Elastofibromatous change of the intestine

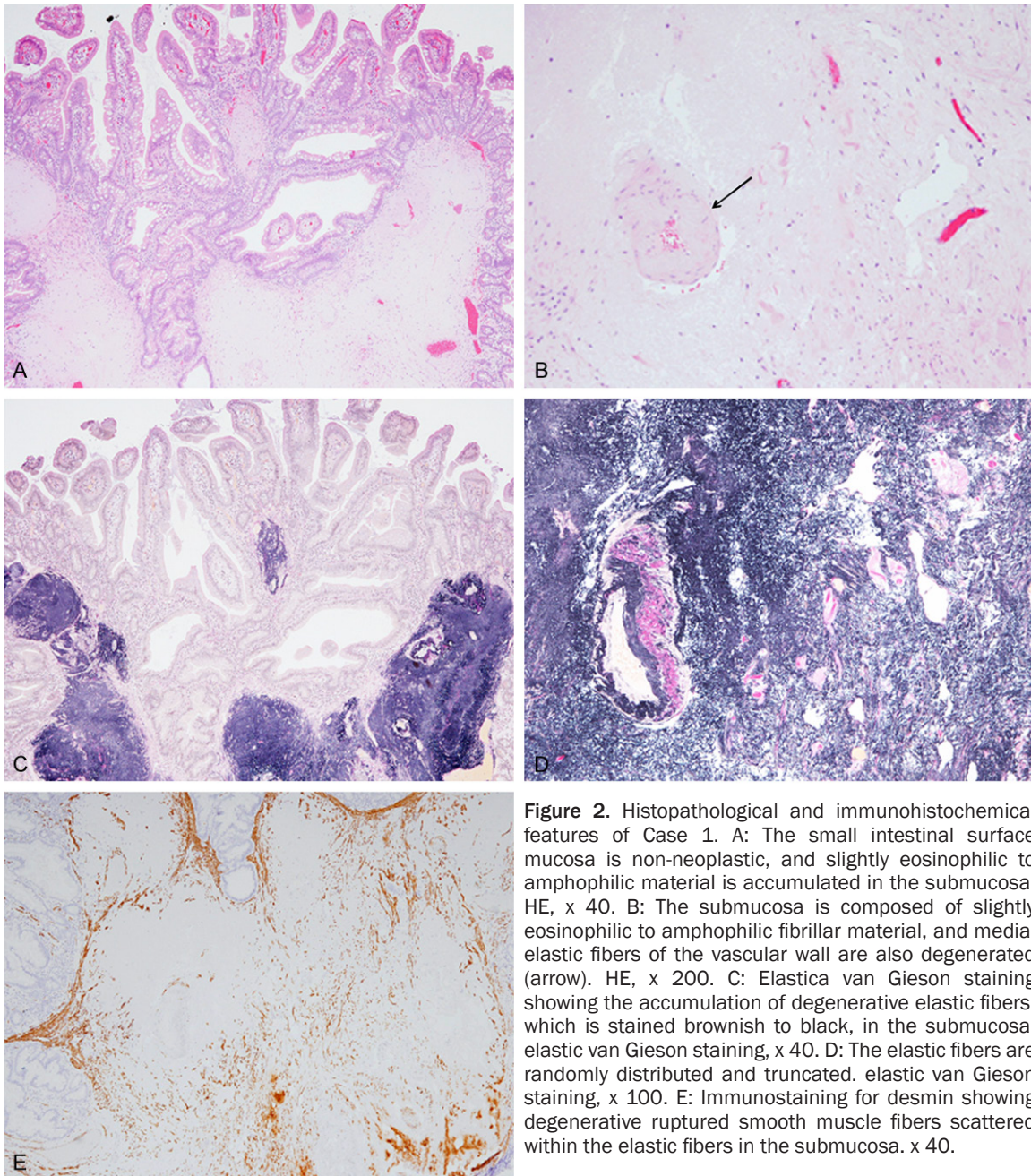


Figure 2. Histopathological and immunohistochemical features of Case 1. A: The small intestinal surface mucosa is non-neoplastic, and slightly eosinophilic to amphophilic material is accumulated in the submucosa. HE, x 40. B: The submucosa is composed of slightly eosinophilic to amphophilic fibrillar material, and medial elastic fibers of the vascular wall are also degenerated (arrow). HE, x 200. C: Elastica van Gieson staining showing the accumulation of degenerative elastic fibers, which is stained brownish to black, in the submucosa. elastic van Gieson staining, x 40. D: The elastic fibers are randomly distributed and truncated. elastic van Gieson staining, x 100. E: Immunostaining for desmin showing degenerative ruptured smooth muscle fibers scattered within the elastic fibers in the submucosa. x 40.

Discussion

In this report, we described four lesions of elastofibromatous change of the intestine from three patients. **Table 1** summarizes the clinicopathological features of the previously reported 32 cases of this type of lesion as well as the present ones [1-9]. The most common site is the colon or rectum (29 cases), and the sigmoid colon is most frequently affected (10 cases). The small intestine is a rare site of this lesion (4

cases), and this report is the first to demonstrate occurrence in the jejunum. This type of lesion shows a slight male predominance (male: female 20: 15) and mainly affects middle-aged to elderly persons (average age 57.34 years), although it can occur in young persons as well (a lesion occurring in a 24-year-old male has been reported [9]). Most of the cases had no clinical characteristics, however, two cases had elastofibromatous lesions in other organs [1, 5]. Enjoji *et al.* described the first document-

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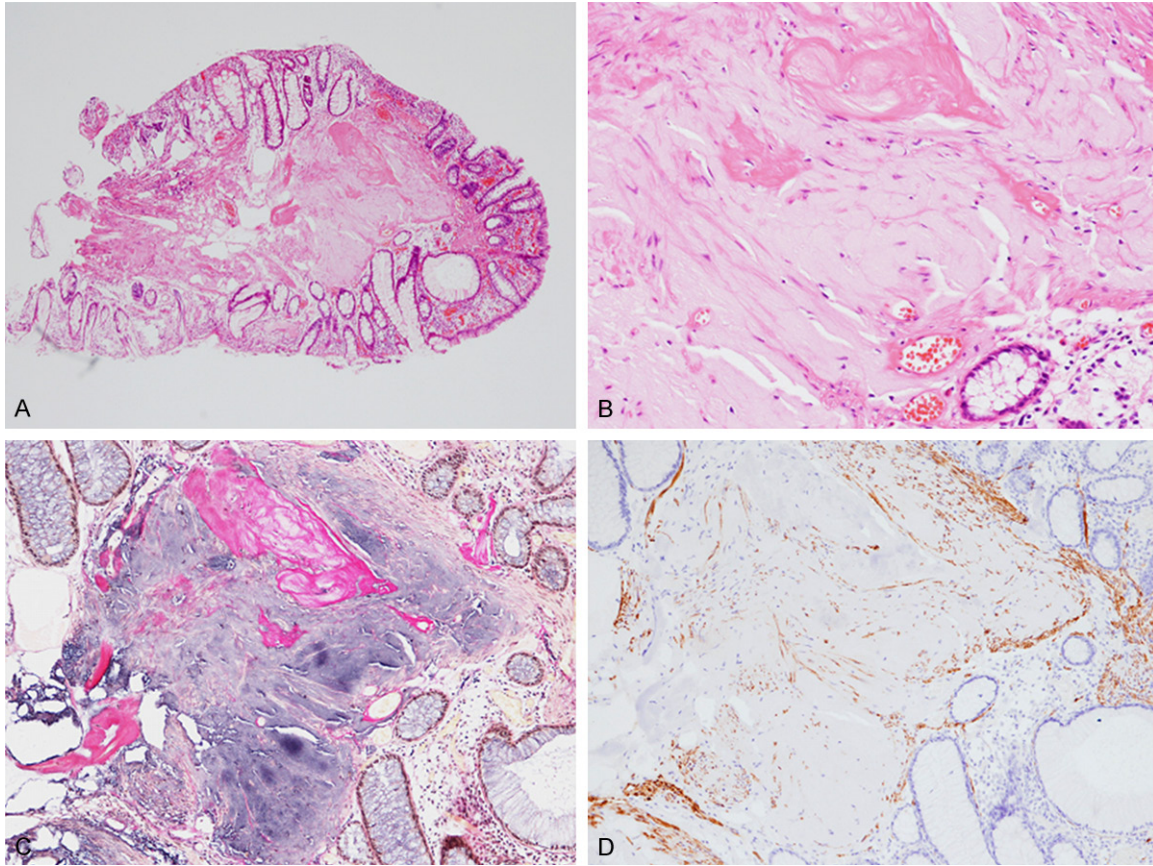


Figure 3. Histopathological features of the transverse colon lesion of Case 2. A: The surface mucosa is slightly hyperplastic, and slightly eosinophilic to amphophilic material is accumulated in the submucosa. HE, x 40. B: The submucosa is composed of slightly eosinophilic to amphophilic fibrillar material, and fibrosis is also noted. HE, x 100. C: Elastica van Gieson staining showing the accumulation of degenerative elastic fibers accompanied by fibrosis in the submucosa, x 40. D: Immunostaining for desmin showing degenerative ruptured smooth muscle fibers scattered within the elastic fibers in the submucosa. x 40.

ed case of elastofibromatous change of the stomach in a patient with elastofibroma dorsi of the bilateral scapular regions [1], and Schiffman reported an autopsy case of elastofibromatous change in both the small intestine and bronchus [5]. Moreover, most of the cases had a single lesion, however, the present Case 2 had two lesions in the transverse and sigmoid colons, and Hobbs *et al.* reported a case of multifocal lesions in the colon [8].

The histopathological characteristics of elastofibromatous change of the gastrointestinal tract are as follows: i) truncated, globular elastic fibers, which are composed of finely granular and/or fibrillar, pale, eosinophilic to gray-tangled amphophilic materials, occasionally with a fibrous component (elastofibromatous change) accumulate in the submucosa and/or muscularis mucosae, ii) this change can be also

observed in the subserosa or serosa in a minority of cases, iii) this change occasionally appears centered around collections of elastotic submucosal vessels, iv) overlying mucosa is composed of non-neoplastic epithelium, and v) no amyloid deposition is noted [1-9]. Although the present four lesions were all endoscopic mucosal resection or biopsy specimens, and the changes in the subserosa or serosa were not available, the histopathological features of the present cases were typical features according to those mentioned above. The main differential diagnostic consideration is amyloidosis [3]. However, differentiating from amyloidosis can be easily done using amyloid staining.

The pathogenesis of elastofibromatous change of the gastrointestinal tract remains unclear. However, it has been proposed that this change is reactive and/or degenerative origin associ-

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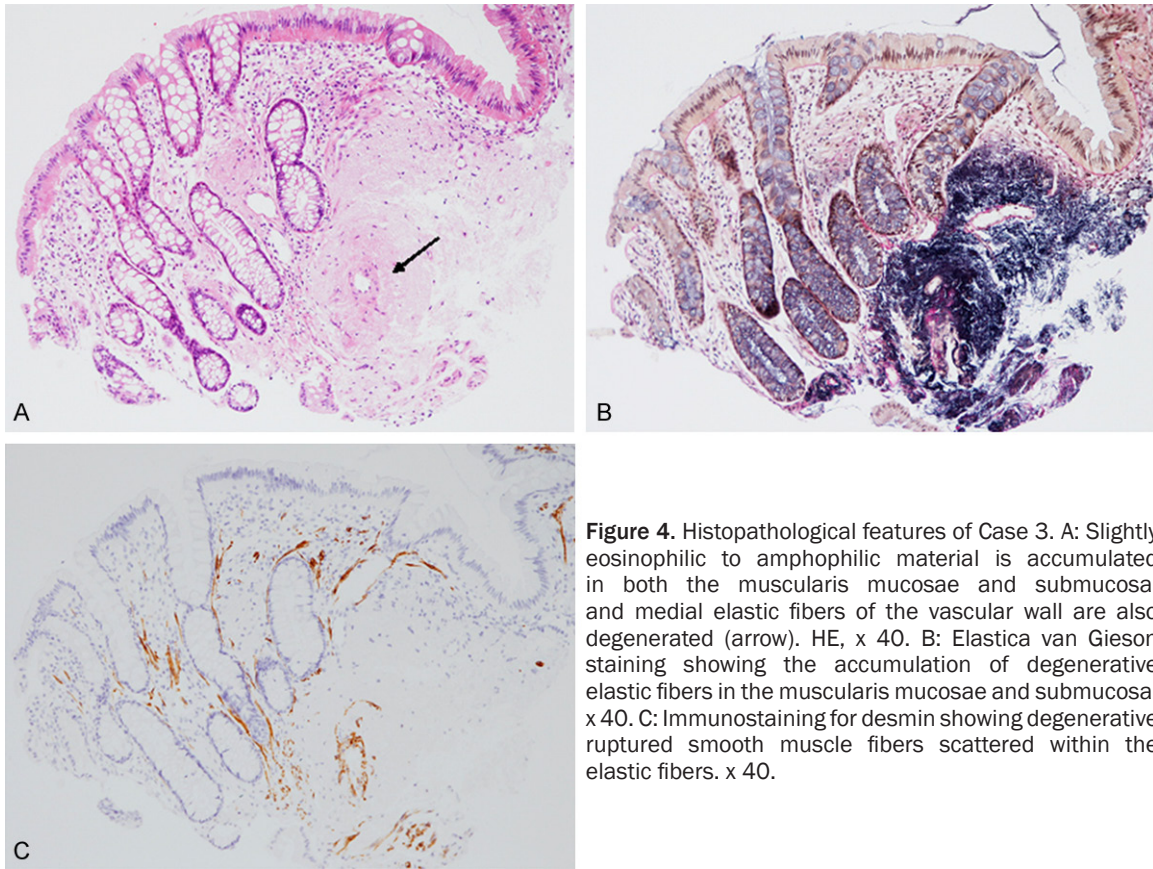


Figure 4. Histopathological features of Case 3. A: Slightly eosinophilic to amphophilic material is accumulated in both the muscularis mucosae and submucosa, and medial elastic fibers of the vascular wall are also degenerated (arrow). HE, x 40. B: Elastica van Gieson staining showing the accumulation of degenerative elastic fibers in the muscularis mucosae and submucosa, x 40. C: Immunostaining for desmin showing degenerative ruptured smooth muscle fibers scattered within the elastic fibers. x 40.

Table 1. Clinicopathological features of elastofibromatous change in the gastrointestinal tract

Case No.	Age/Gender	Location	Clinical characteristics	Reference
1	69/Female	Stomach	Elastofibroma dorsi of the bilateral scapular regions	[1]
2	49/Male	Transverse colon	None	[2]
3	58/Female	Rectum	History of multiple myeloma, and endoscopically rectum was normal-appearing	[3]
4	55/Male	Colon	None	[4]
5	34/Male	Descending colon	None	[4]
6	30/Female	Sigmoid colon	None	[4]
7	32/Male	Rectum	None	[4]
8	58/Female	Descending colon	None	[4]
9	35/Female	Sigmoid colon	None	[4]
10	46/Male	Sigmoid colon	Psoriasis	[4]
11	88/Female	Small intestine	Deposition was also observed in the bronchus.	[5]
12	69/Female	Sigmoid colon	None	[6]
13	72/Male	Sigmoid colon	None	[7]
14	63/Female	Colon	Multifocal lesions	[8]
15	53/Male	Colon	None	[8]
16	61/Female	Sigmoid colon	None	[8]
17	77/Male	Ileum	Small bowel obstruction	[8]
18	77/Male	Stomach	Refractory gastroesophageal reflux, post-gastrectomy	[8]
19	51/Female	Sigmoid colon	None	[8]
20	71/Female	Descending colon	Diverticulitis	[8]
21	83/Male	Transverse colon	None	[8]
22	55/Male	Stomach	Markedly thickened gastric wall, post-gastrectomy	[8]
23	61/Female	Cecum	None	[8]
24	47/Male	Colon	None	[8]

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25	50/Female	Transverse colon	None	[8]
26	47/Male	Descending colon	None	[8]
27	45/Male	Colon	None	[9]
28	60/Female	Sigmoid colon	None	[9]
29	55/Female	Rectum	Diverticulitis	[9]
30	70/Male	Colon	None	[9]
31	24/Male	Ileum	None	[9]
32	64/Male	Colon	Colon cancer	[9]
Case 1	76/Male	Jejunum	Black stool	
Case 2	70/Male	Transverse colon	None	
		Sigmoid colon		
Case 3	52/Male	Sigmoid colon	None	

ated with tissue irritation [4, 8, 9]. Most cases of this lesion, including the present cases, have degenerative elastotic fibers of the vessels in the submucosa. Therefore, the possibility that elastofibromatous change of the gastrointestinal tract represents elastic degeneration of superficial submucosal vessels, resulting in progressive elastic degeneration, obliteration of vascular lumens, and subsequent fibrosis, has been raised [8, 9]. This hypothesis is convincing because the presence of elastofibromatous change centered around collections of submucosal elastotic vessels occurs in most of the cases. Thus, Märkl *et al.* proposed the term "gastrointestinal angioelastosis" for this type of lesion [9]. Moreover, this report clearly demonstrated that desmin-positive degenerative ruptured smooth muscle fibers were scattered within the elastic fibers in the submucosa. This observation might be related to the degenerative change of the vascular wall in the submucosa.

Elastofibroma is defined as a benign, ill-defined proliferation of elastofibromatous tissue characterized by an excessive number of abnormal elastic fibers [15]. This tumor has an unknown etiology, however it is thought to be induced by a response to repeated trauma or friction between the lower scapula and the thoracic wall [15]. In contrast, recent studies have shown that elastofibroma shows a clonal neoplastic change in nature [16, 17]. Two cases of gastrointestinal elastofibromatous change associated with elastofibromatous lesions outside the gastrointestinal tract (one associated with elastofibroma dorsi of the scapular regions [1] and the other associated with elastofibromatous change in the bronchus [5]) have been documented. Accordingly, a minority of elastofibromatous change of the gastrointestinal tract is

associated with lesions in other organ, however, most cases are restricted to the gastrointestinal tract, and this type of lesion is thought to occur as a result of previous persistent vascular injury.

Disclosure of conflict of interest

None.

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