Benign Esophageal Tumors

5

Kyung Sik Park

Contents

5.1 Epithelial Tumors

5.1.1 Squamous Papilloma

Esophageal papilloma is a rare benign epithelial tumor. On histological examination, the lesions are characterized histologically by fingerlike fronds lined by an increased number of squamous cells. Although multiple lesions can be found, most of these tumors are solitary. Various inflammatory conditions or human papilloma virus (HPV) seems to be associated with the pathogenesis of esophageal papilloma. Endoscopy reveals small, whitish-pink, wartlike exophytic projections (Fig. 5.1) [1]. If papilloma is suspected, punch biopsy should be performed to differentiate this lesion from other similar-looking lesions, such as early squamous cell carcinoma, and papillary leukoplakia. Since the malignant potential of this tumor is very low, regular follow-up is not recommended. In the case of a large tumor causing dysphagia, endoscopic resection is not difficult (Table 5.1).

5.1.2 Adenoma

Although esophageal adenomas without Barrett's esophagus have been reported, these lesions arise almost exclusively in the segments of Barrett's esophagus. This disease entity already has been discussed in the previous chapter.

K. S. Park
Department of Internal Medicine, Dongsan Medical Center, Keimyung University College of Medicine, Daegu, South Korea
e-mail: seenae99@dsmc.or.kr

H. J. Chun et al. (eds.), *Clinical Gastrointestinal Endoscopy*, https://doi.org/10.1007/978-981-10-4995-8_5

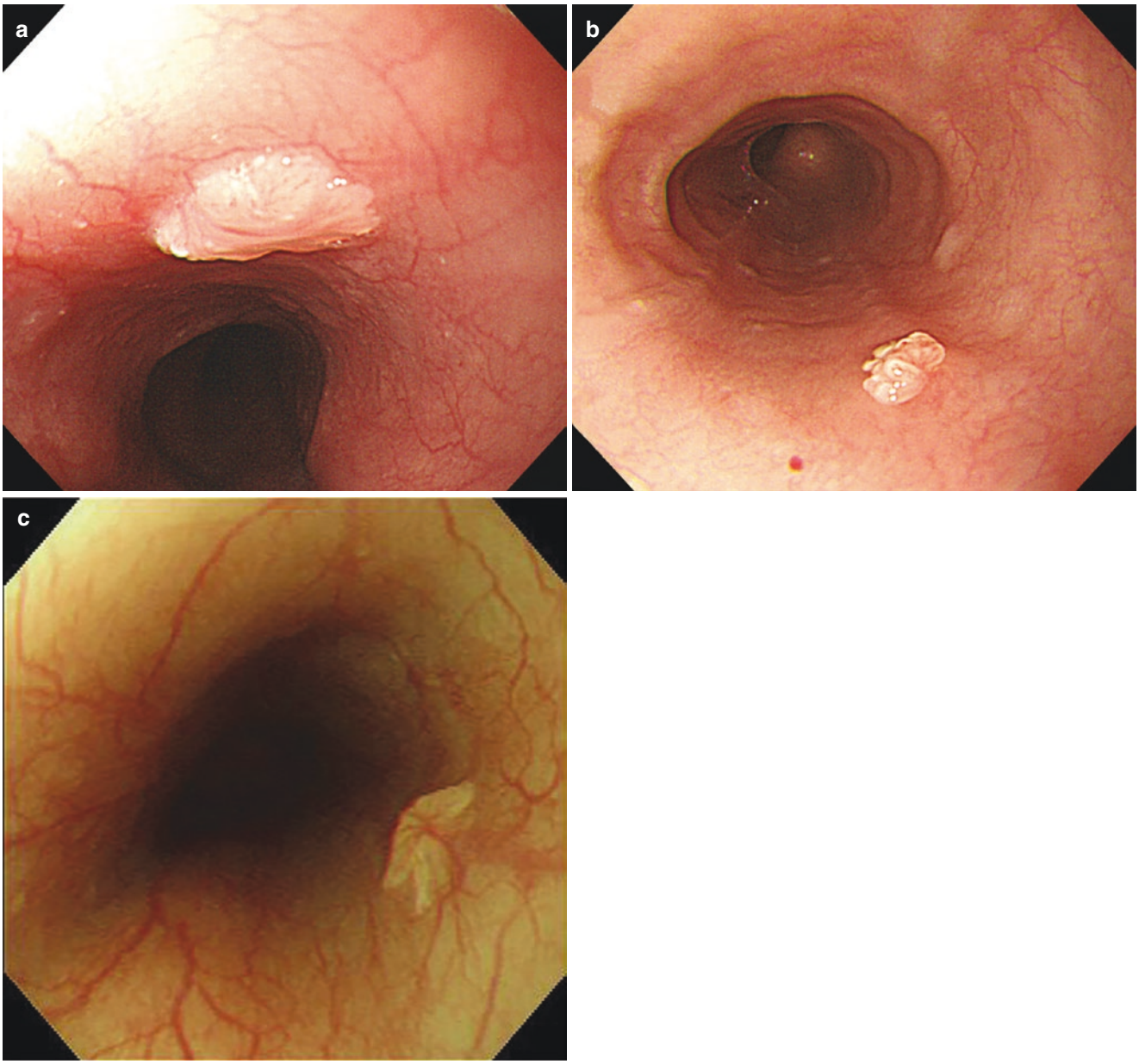

Fig. 5.1 Squamous papilloma of various shapes. (**a**) An approximately 8 mm, whitish, wartlike exophytic projection at the upper esophagus. (**b**) An approximately 5 mm, whitish-pink, exophytic projection with lobulated surface at the mid-esophagus. (**c**) An approximately 5 mm, whitish nodular lesion with atypical non-round shape at the upper esophagus. (**d**) An approximately 6 mm, whitish-pink, flat nodular lesion at the upper esophagus

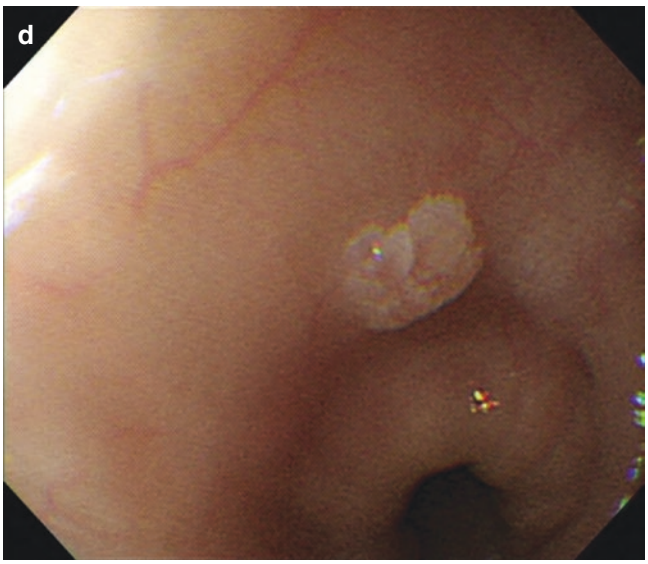

Fig. 5.1 (continued)

Table 5.1 Classification of benign esophageal tumors

Epithelial tumors
Adenoma
Nonepithelial tumors
Leiomyoma
Granular cell tumor
Cystic tumors
Bronchogenic cyst
Duplication cyst
Lymphangioma
Fibrovascular polyp
Inflammatory fibroid polyp
Lipoma
Hemangioma

5.2 Nonepithelial Tumors

Since subepithelial tumors are covered with intact squamous epithelium, it is difficult to know the histological origin of the tumors endoscopically. EUS is a standard diagnostic tool for the diagnosis of subepithelial tumors.

5.2.1 Leiomyoma

Leiomyoma is the most common esophageal benign tumor. It arises from the muscularis mucosa or muscularis propria. Most cases are found incidentally, because esophageal leiomyomas rarely cause symptoms when they are smaller than 5 cm in diameter. On endoscopy, they usually appear as variable-sized nonspecific protrusions in the esophageal wall covered with intact squamous mucosa (Figs. 5.2 and 5.3). Accurate origin, size, and nature of the tumors can be assessed with EUS [2]. The typical finding on EUS is a hypoechoic round mass originating from the second or fourth layer. Lesions originating from the second layer are usually small and can be easily removed by endoscopic mucosal resection technique. Cases with tumors less than 3 cm in size can be followed up without resections if there are no symptoms.

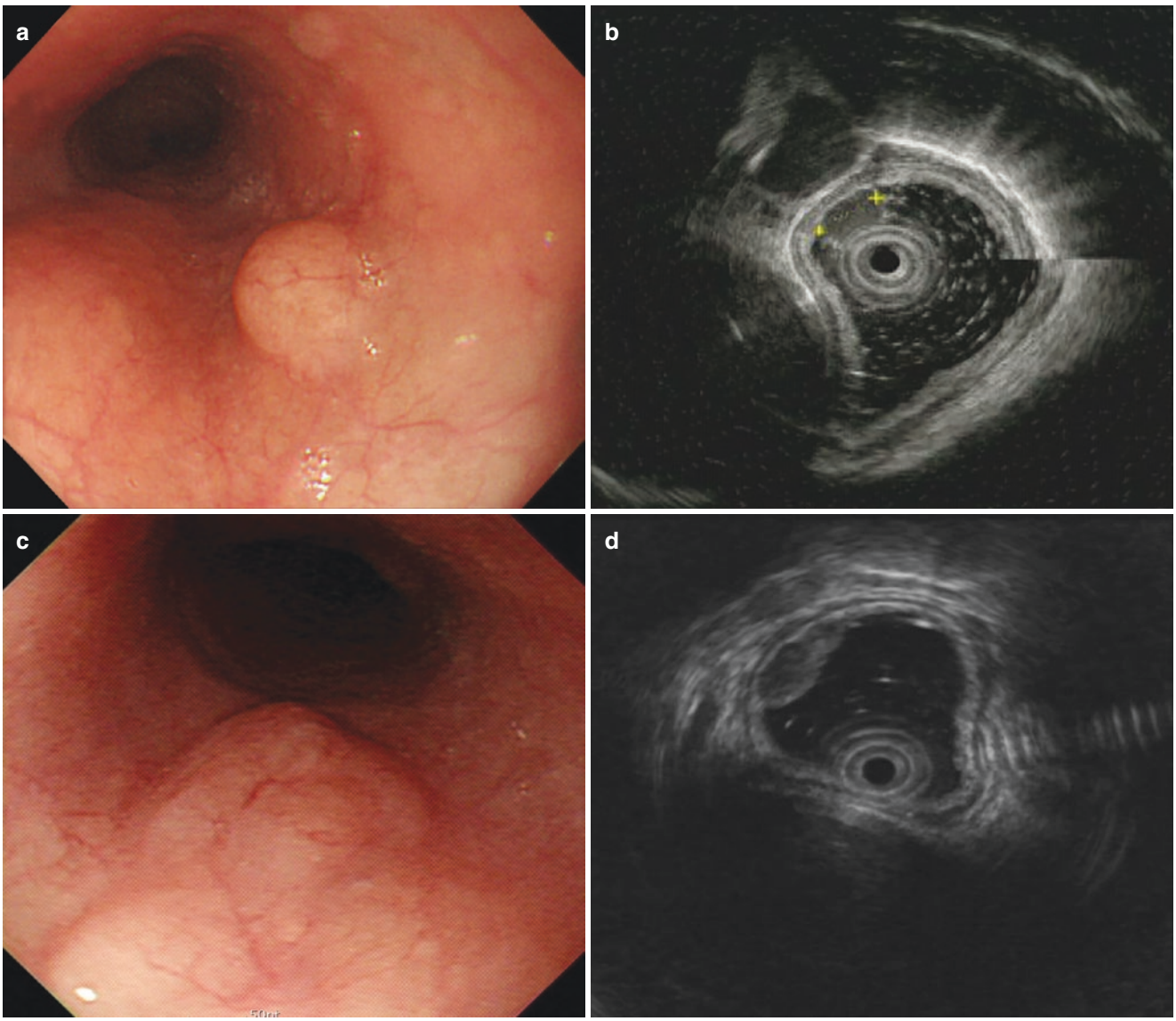

Fig. 5.2 Leiomyoma from muscularis mucosa. (**a**) Small protrusion covered with intact mucosa at mid-esophagus. (**b**) EUS shows 5 mm-sized homogenously hypoechoic mass continuous from the second layer (muscularis mucosa). (**c**) Protrusion covered with intact mucosa at mid-esophagus. (**d**) EUS shows 9.8 mm-sized homogenously hypoechoic mass continuous from the second layer

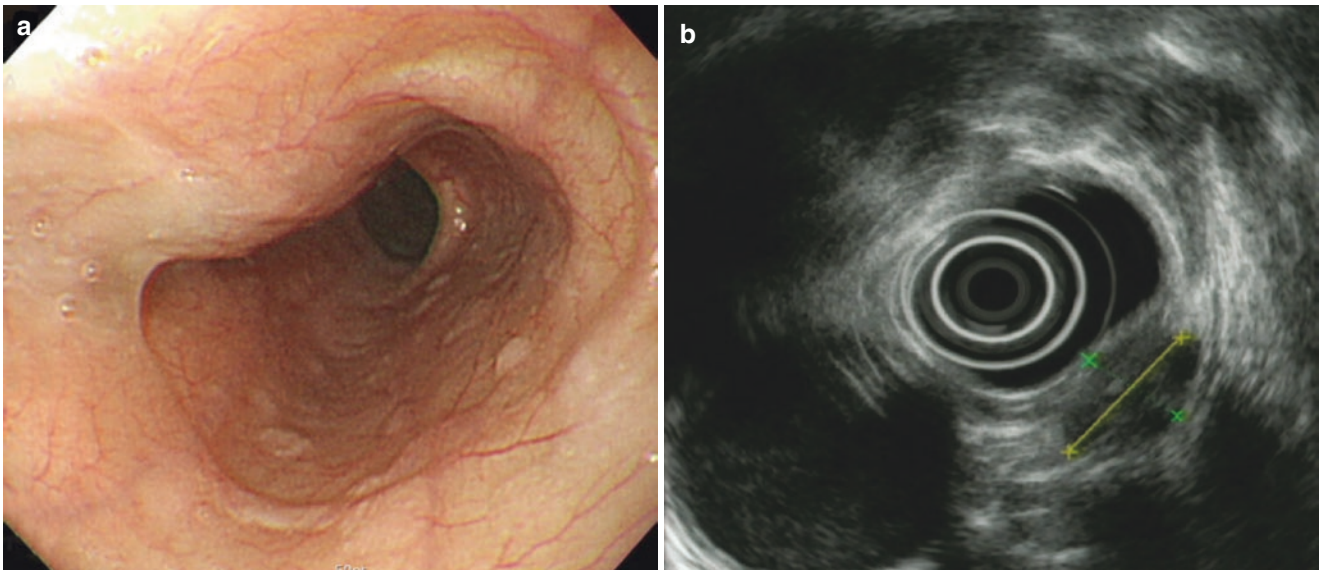

Fig. 5.3 Leiomyoma from muscularis propria. (**a**) Blunt protrusion covered with intact mucosa at mid-esophagus. (**b**) EUS shows a 16 mm-sized homogenously hypoechoic mass continuous from the fourth layer (muscularis propria)

5.2.2 Gastrointestinal Stromal Tumor

Gastrointestinal stromal tumor (GIST) is one of the most common GI mesenchymal tumors, which most likely originates from the interstitial cells of Cajal (ICCs), the majority of which are located in the myenteric plexus. They are usually located in the stomach and proximal small bowel, although they can possibly occur in any portion of the GI tract. However, esophageal GIST is very rare and there are only a few case reports. On endoscopy and EUS, findings are similar to those of leiomyoma from the muscularis propria; a protrusion covered with intact mucosa is the main finding. Therefore, it usually cannot be discriminated from leiomyoma endoscopically (Fig. 5.4) [3]. Immunohistochemical stains for CD117, DOG-1, S100 protein, smooth muscle actin, and desmin in the tissue obtained through resection or EUS-guided punch biopsy are necessary to diagnose and to exclude other SETs.

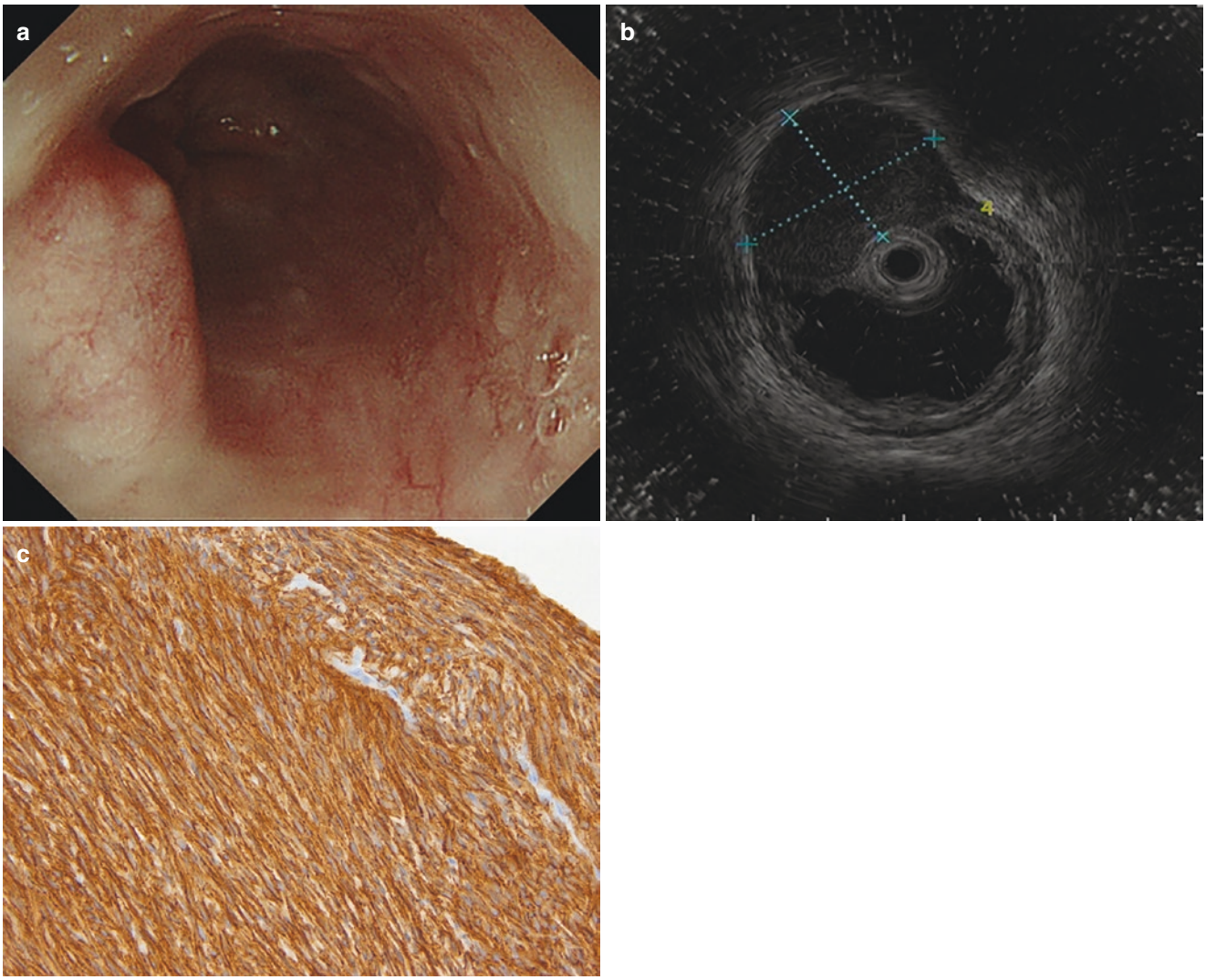

Fig. 5.4 GIST in the esophagus. (**a**) Blunt protrusion covered with intact mucosa at the distal esophagus. (**b**) EUS shows maximally 15 mm-sized homogenously hypoechoic mass continuous from the fourth layer (muscularis propria). (**c**) Immunohistochemical stains for CD117 are positive

5.2.3 Granular Cell Tumor

Granular cell tumor is a rare esophageal tumor. The esophagus is the most common site for this tumor in the GI tract. Strong positivity for S100 protein on immunohistochemical staining suggests that the tumors originated from cells of neural origin. If present, dysphagia is the most common symptom, although most patients are asymptomatic. On endoscopy, they typically show a yellowish-white, molar tooth-shaped, sessile polypoid appearance (Fig. 5.5). Usually, they feel firm or rubbery when compressed with a forceps. The typical EUS finding is a homogenous hypoechoic mass located within the submucosal layer. It is not difficult to confirm this diagnosis with punch biopsy because this tumor is located close to the epithelial layer. Although rare, this tumor has malignant potential. Therefore, removal with excisional biopsy or EMR is recommended, if possible.

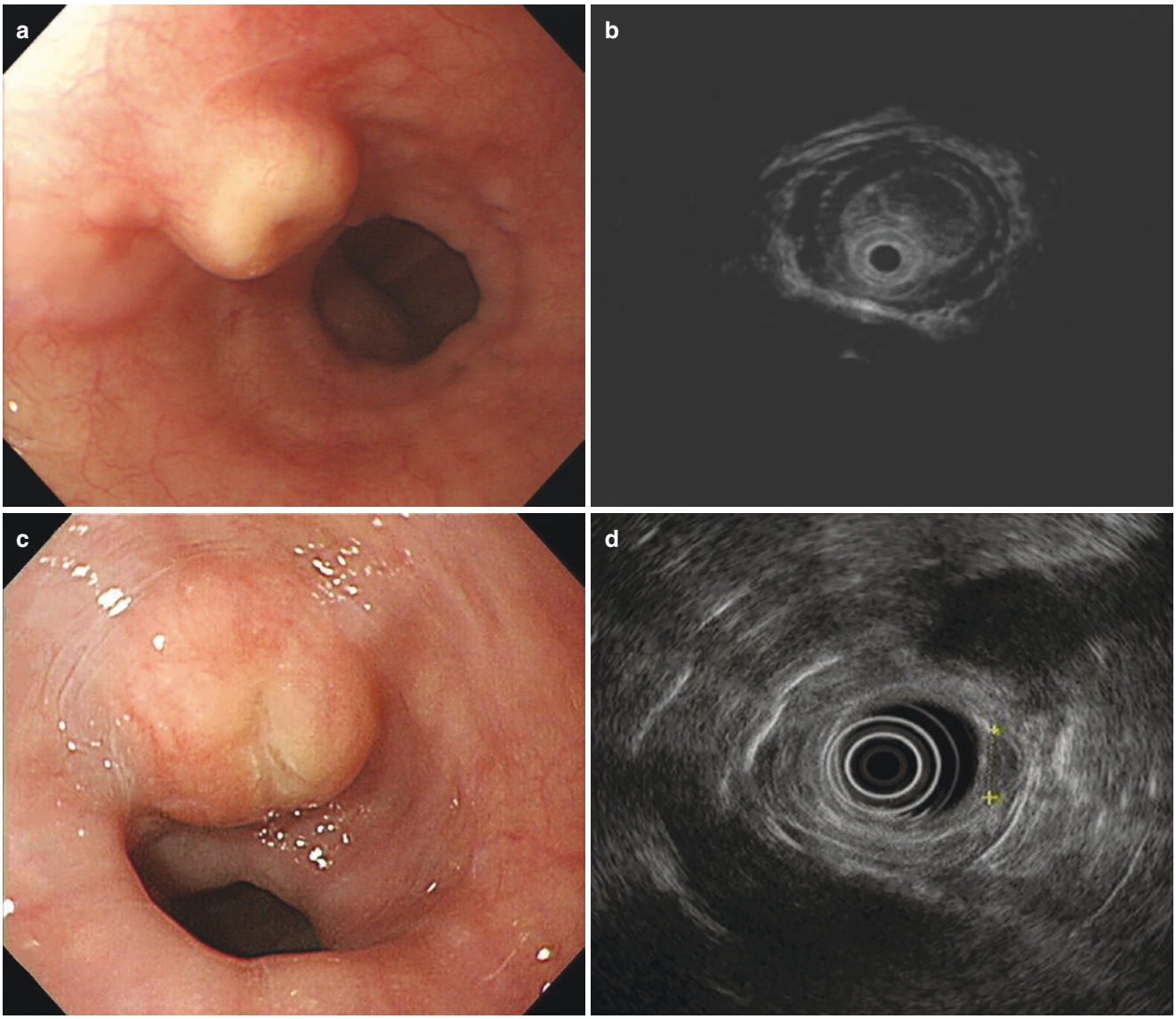

Fig. 5.5 Granular cell tumor. (**a**) Yellowish-white, molar tooth-shaped protrusion covered with intact mucosa at the mid-esophagus. (**b**) EUS shows an 8 mm-sized homogenously hypoechoic mass within the submucosal layer. (**c**) Yellowish-white, molar tooth-shaped protrusion covered with slightly inflamed mucosa at the mid-esophagus. (**d**) EUS shows a 12 mm-sized homogenously hypoechoic mass within submucosal layer. (**e**) Yellowish-white, molar tooth-shaped protrusion covered with slightly inflamed mucosa at the mid-esophagus. (**f**) EUS shows a 10 mm-sized homogenously hypoechoic mass within submucosal layer

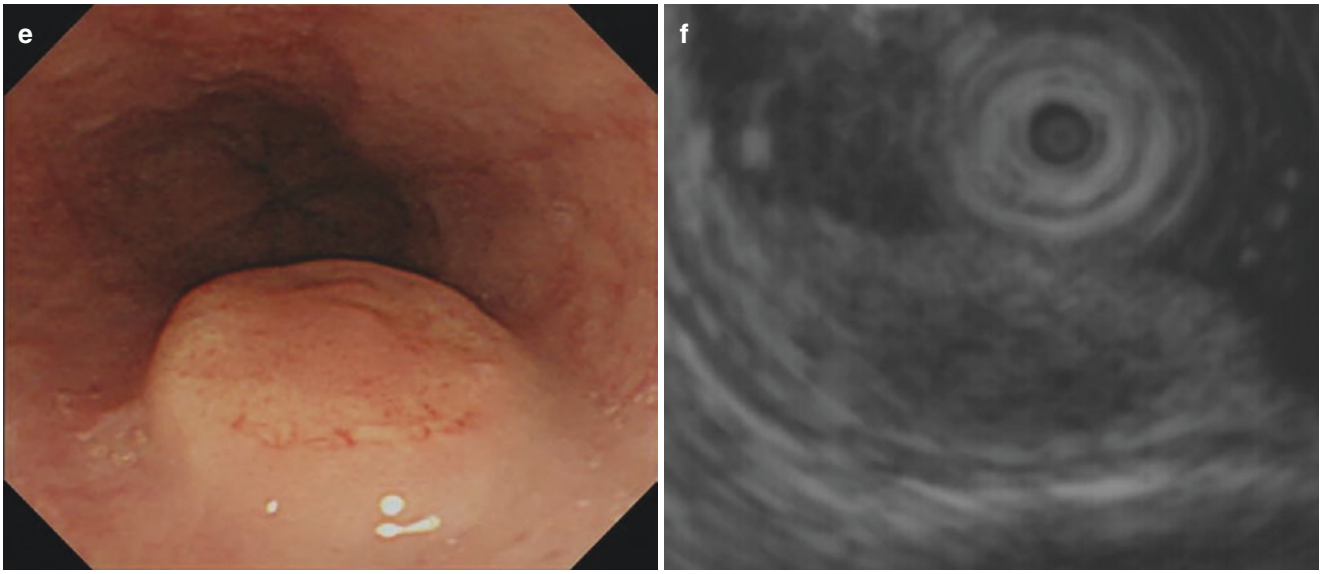

Fig. 5.5 (continued)

5.2.4 Cystic Tumors

Cystic tumors are also rare in the esophagus. Endoscopy reveals variously sized, easily compressible, and soft masses (Fig. 5.6). On EUS, variously sized anechoic masses can be found in the layer of origin [4]. Several types of cystic lesions can develop in the esophagus. First, cysts can arise from the lamina propria or submucosa because of various inflammatory responses as discussed previously. Second, many esophageal cysts arise from mediastinal structures such as in bronchogenic cysts. In this type, the cystic wall is lined by an epithelial layer. Third, duplication cysts as a kind of congenital anomaly may arise in the esophagus during the early embryonic development. In this type, the lesions are covered by two muscle layers. Finally, although very rare in the esophagus, lymphangioma that results from malformations of sequestered lymphatic tissue also shows a cystic nature.

5.2.5 Fibrovascular Polyp

Fibrovascular polyps are nonneoplastic intraluminal masses, which usually develop in the cervical part of the esophagus. Although not fully understood, the mechanism is believed to be the elongation of nodular thickening of a redundant mucosal fold by repeated swallowing. Histologically, they contain a mixture of fibrous, vascular, and adipose tissues and are covered by intact squamous epithelium. On endoscopy, these tumors also appear as protruding masses covered by intact mucosa in the cervical esophagus. In cases of large symptomatic masses, endoscopic or surgical resection may be considered. EUS should be performed before endoscopic resection to rule out the presence of a large vessel.

5.2.6 Inflammatory Fibroid Polyp

Inflammatory fibroid polyp denotes a variety of lesions composed of reactive blood vessels, fibroblasts, and various inflammatory cells. This tumor is also rare in the esophagus. Inflammatory response to acid reflux is considered a possible mechanism. Inflammatory pseudopolyps and eosinophilic granulomas can be included in this category.

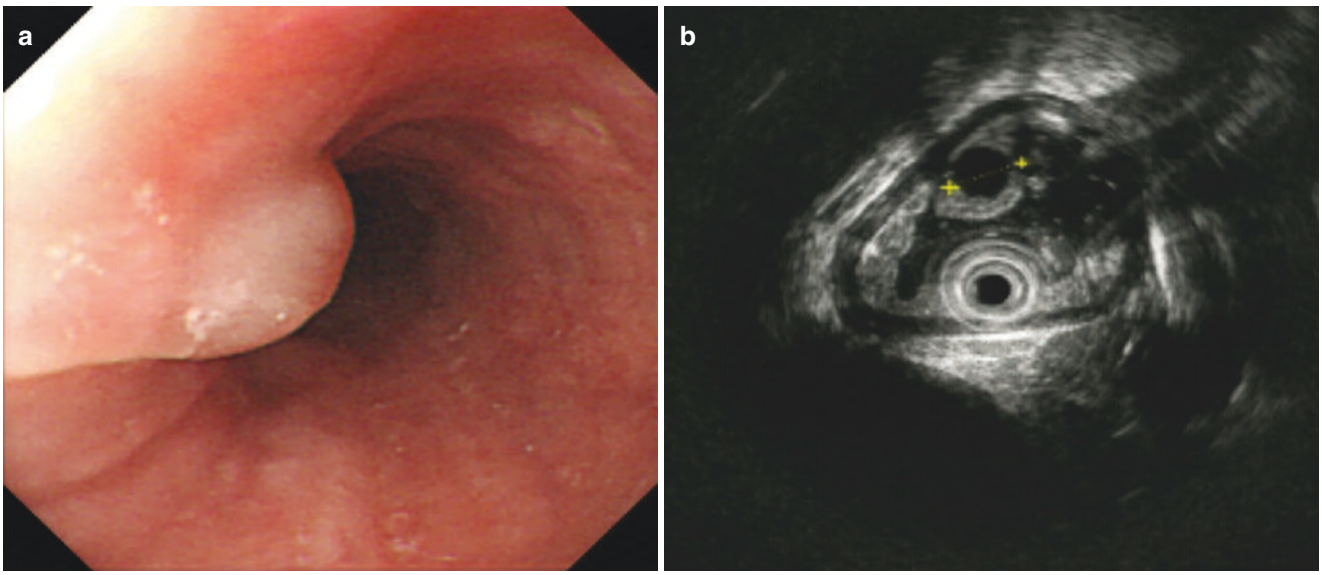

Fig. 5.6 Submucosal cyst. (**a**) Small soft mass covered with intact, transparent-looking mucosa. (**b**) EUS shows a 4.7 mm-sized anechoic lesion within the submucosal layer

5.2.7 Lipoma

In contrast to the stomach or small intestine, the esophagus is rarely affected by lipoma. On endoscopy, it is observed as pale or yellowish-colored soft mass (Fig. 5.7). A homogenously hyperechoic mass located in the submucosal layer on EUS strongly suggests lipoma.

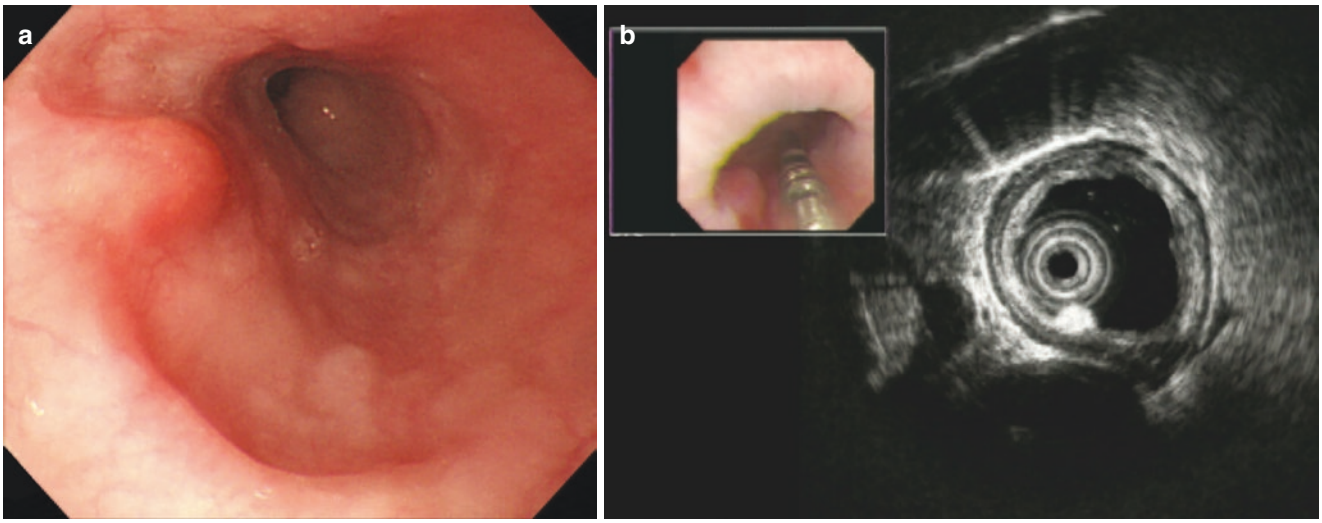

Fig. 5.7 Lipoma. (**a**) Small yellowish-colored soft mass at the mid-esophagus. (**b**) EUS shows a 4 mm-sized hyperechoic lesion within the submucosal layer

5.2.8 Hemangioma

Hemangiomas are also very rare benign tumors in the esophagus and are usually found incidentally. They are lined with vascular endothelium. If present, they appear as nodular, soft, and bluish-red protrusions on endoscopy (Fig. 5.8) [5]. When pressed with biopsy forceps, they show pale discoloration. Simple phlebectasia in the submucosal layer is a nontumorous condition occasionally misdiagnosed as hemangioma because both lesions can present as bluish protrusions covered with intact squamous mucosa on endoscopy (Fig. 5.9).

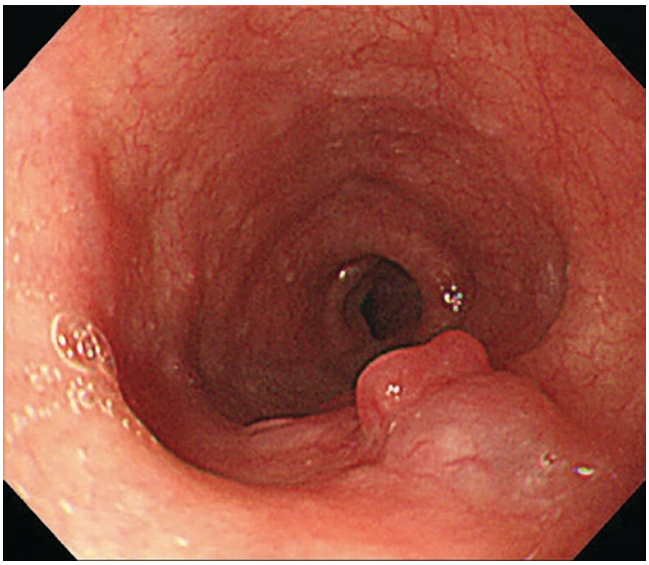

Fig. 5.8 Capillary hemangioma. An approximately 1 × 1.5 cm-sized, soft, and bluish-red protrusion is noted at the mid-esophagus

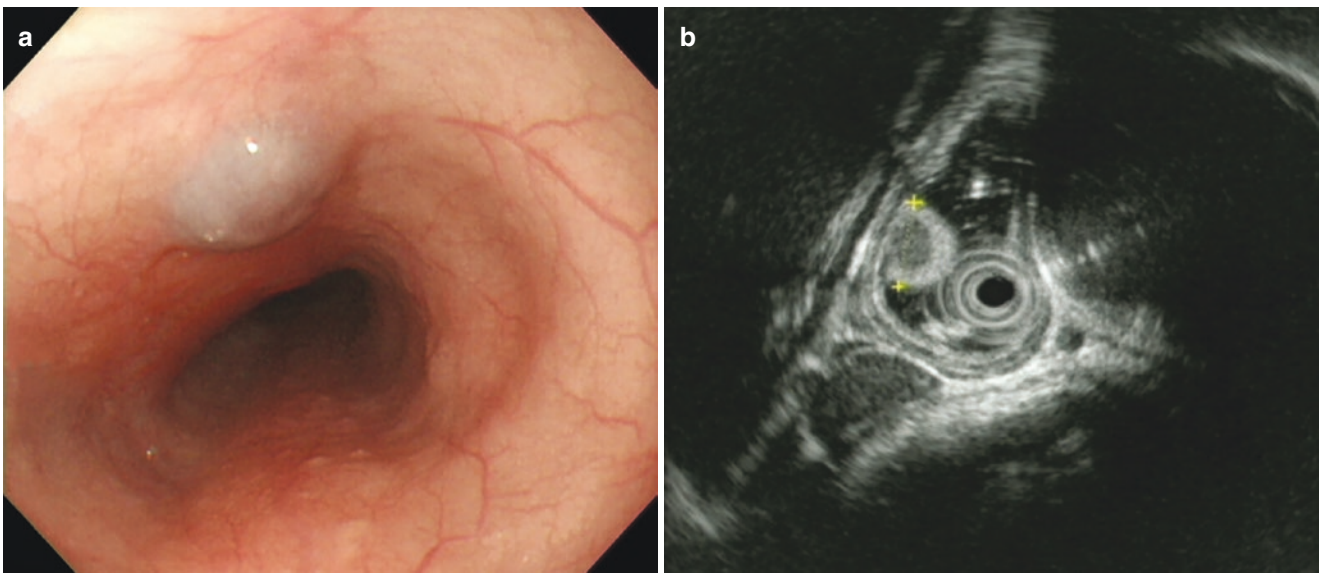

Fig. 5.9 Phlebectasia. (**a**) Upper GI endoscopy shows bluish protrusion less than 1 cm in diameter. (**b**) EUS shows a 6 mm-sized hyperechoic mass within the third layer and also shows internal hypoechoic focus

Quiz

A 45-year-old man visited a health promotion center for an upper GI endoscopy screening. He had no specific GI symptoms. He was a nonsmoker and social drinker. Upper GI endoscopy and microscopic findings are presented in Figs. 5.10 and 5.11, respectively.

Question. What is your diagnosis?

Pathologic review of the biopsy specimen taken from the largest lesion revealed several lobules of the cells with sebaceous differentiation. This finding is consistent with the diagnosis of heterotopic sebaceous glands in the esophagus. Since this extremely rare condition had shown no malignant potential, no therapeutic procedure was performed. Follow-up UGI endoscopy after 2 years is showing similar findings as those of the previous examination (Fig. 5.12).

Answer. Heterotopic sebaceous glands in the esophagus.

Acknowledgement Special thanks to Prof. Jun Haeng Lee for providing wonderful photos.

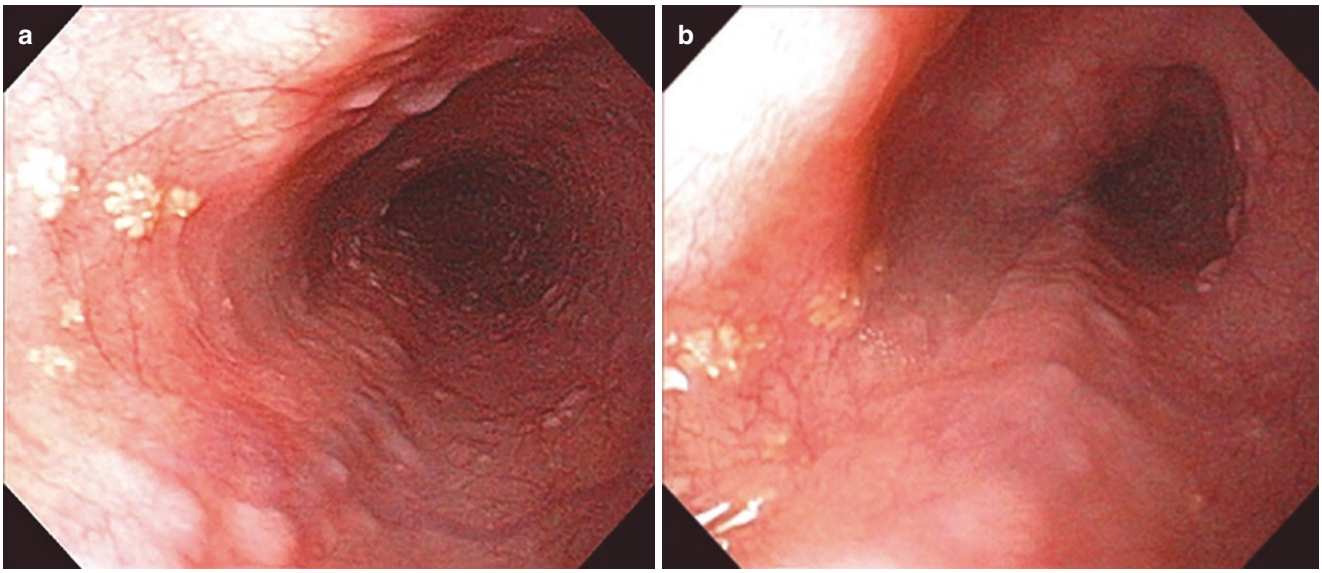

Fig. 5.10 Endoscopic findings. Multiple yellowish granular spots are observed at mid to distal esophagus. (**a**) Upper esophagus, (**b**) mid-esophagus

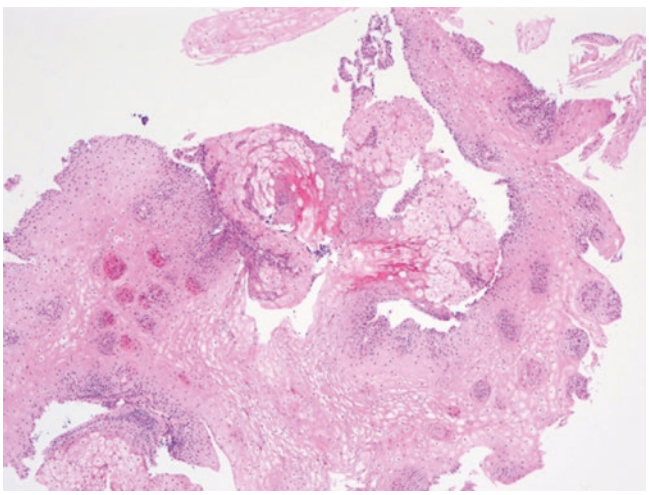

Fig. 5.11 Pathologic findings. Several lobules of the cells show sebaceous differentiation consistent with heterotopic sebaceous glands

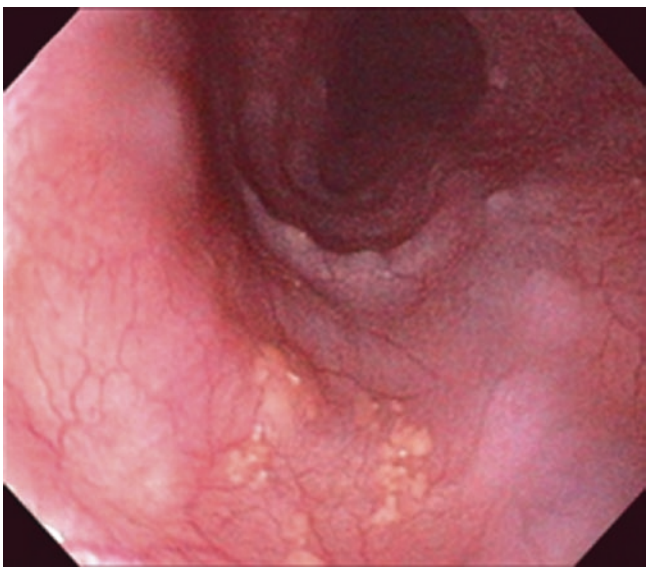

Fig. 5.12 Endoscopic findings after 2 years. Multiple yellowish granular spots are still observed

References

1. Korean Society of Gastrointestinal Endoscopy. Atlas of gastrointestinal endoscopy. 1st ed. Seoul: Daehan Medical Book; 2011.
2. Rice TW. Benign esophageal tumors: esophagoscopy and endoscopic esophageal ultrasound. Semin Thorac Cardiovasc Surg. 2003;15:20–6.
3. Classen M, Tytgat GNJ, Lightdale CJ. Gastroenterological endoscopy. 2nd ed. London: Thieme; 2010.
4. Bernat M, Strutynska-Karpinska M, Lewandowski A, et al. Benign esophageal tumors. Wiad Lek. 1993;46:24–7.
5. Wiechowski S, Filipiak K, Walecka A, et al. Benign intramural esophageal tumors. Pol Tyg Lek. 1979;34:1871–2.