Benign Esophageal Tumors

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During endoscopy, various benign esophageal lesions are encountered in the esophagus. Most are asymptomatic and have no malignant potential. These various benign lesions can originate from different wall layers in the esophagus. According to its origin, esophageal tumors can be classified as epithelial and subepithelial tumors (SETs) (Table 5.1). Papilloma is the most common epithelial tumor in the esophagus and shows small, whitish-pink, wartlike exophytic projections on endoscopy. Although SETs originating from superficial side of the esophageal wall can present characteristic endoscopic findings such as yellowish, molar tooth-shaped appearance in granular cell tumor, the lesions originating from deep layer such as leiomyoma or gastrointestinal stromal tumor show similar morphology on endoscopy and even on endoscopic ultrasonography (EUS). Therefore, sometimes it is difficult to know the histological origin of the tumors endoscopically. In this chapter, endoscopic findings of various types of benign esophageal tumors will be discussed with examples.

Table 5.1 Classification of benign esophageal tumors

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

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5.1 Epithelial Tumors

5.1.1 Squamous Papilloma

Esophageal papilloma is a rare benign epithelial lesion. Lesions are characterized histologically by fingerlike fronds lined by an increased number of squamous cells. Although multiple lesions can be found, the majority 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 suspected, punch biopsy should be performed to differentiate this lesion from other similarly 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 large case causing dysphagia, endoscopic resection is not difficult.

5.1.2 Adenoma

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

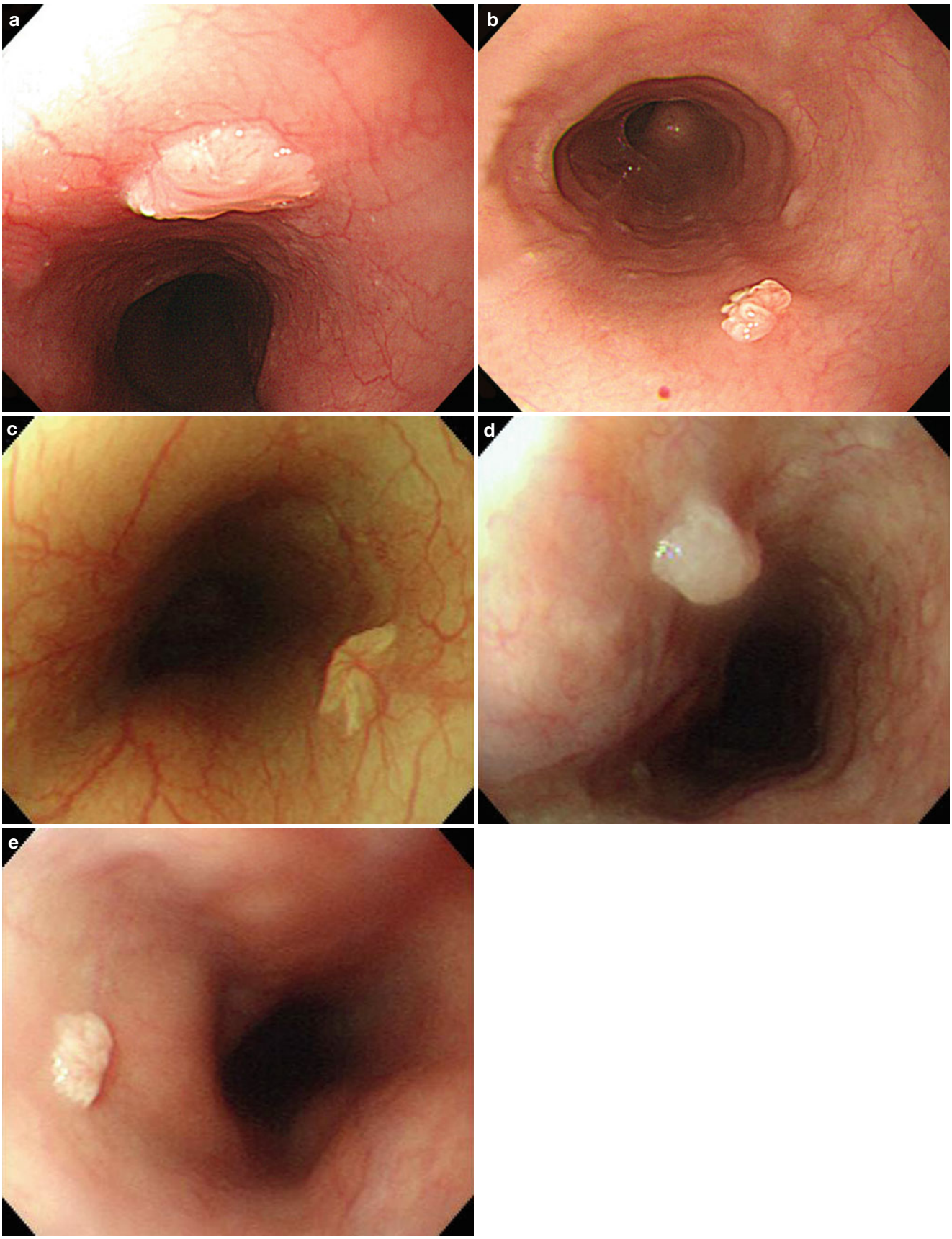

Fig. 5.1 Squamous papilloma of various shapes. (**a**) An approximately 8 mm, whitish, wartlike exophytic projection at the upper esophagus. (**b**) An approximately 5 mm, whitish-pink, exophytic projection with lobulated surface at the mid-esophagus. (**c**) An approximately 5 mm, whitish nodular lesion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

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 in diagnosing 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 covered with intact squamous mucosa in the esophageal wall (Figs. 5.2 and 5.3). Accurate origin, size, and nature of the tumors can be assessed with EUS [2]. Most typical findings in EUS are hypoechoic round mass originated from the second or fourth layer. Lesions originated from the second layer are usually small in size and can be easily removed by endoscopic mucosal resection technique. Cases less than 3 cm in size can be followed up without resections if there is no symptom.

5.2.2 Gastrointestinal Stromal Tumor

Gastrointestinal stromal tumor (GIST) is one of the most common GI mesenchymal tumors, which most likely originates from interstitial cell of Cajal (ICC), the majority of which are located in the myenteric plexus. Usually they are located in the stomach and proximal small bowel, although they can occur in any portion of the GI tract. However, esophageal GIST is very rare and there are only several case reports. On endoscopy and EUS, this tumor shows similar findings with leiomyoma from 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 from the tissue taken by resection or EUS-guided punch biopsy are necessary to diagnose and to exclude other SETs.

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 in immunohistochemical stain suggests that they originate from cells of neural origin. If present, dysphagia is the most common symptom, although most patients are asymptomatic. On endoscopy, typically they show yellowish-white, molar tooth-shaped, sessile polypoid appearance (Fig. 5.5). Usually they feel firm or rubbery when compressed with a forceps. Typical EUS finding is homogenous hypoechoic mass located within submucosal layer. It is not difficult to confirm this diagnosis with pinch 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.

5.2.4 Cystic Tumors

Cystic tumors are also rare in the esophagus. Endoscopy shows them as variously sized, easily compressible, soft masses (Fig. 5.6). On EUS, variously sized anechoic masses can be found in the arising layer [4]. There are several types of cystic lesions which can develop in the esophagus. First, cysts can arise from the lamina propria or submucosa as a result 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 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. Last, although very rare in esophagus, lymphangioma which 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 areas of the esophagus. Although not fully understood, elongation of nodular thickening of a redundant mucosal fold by repeated swallowing is thought as a mechanism. Histologically, they contain a mixture of fibrous, vascular, and adipose tissue and are covered by intact squamous epithelium. On endoscopy, these tumors also appear as protruding mass covered by intact mucosa in cervical esophagus. In the large symptomatic cases, 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 though as a possible mechanism. Inflammatory pseudopolyps and eosinophilic granulomas can be included in this category.

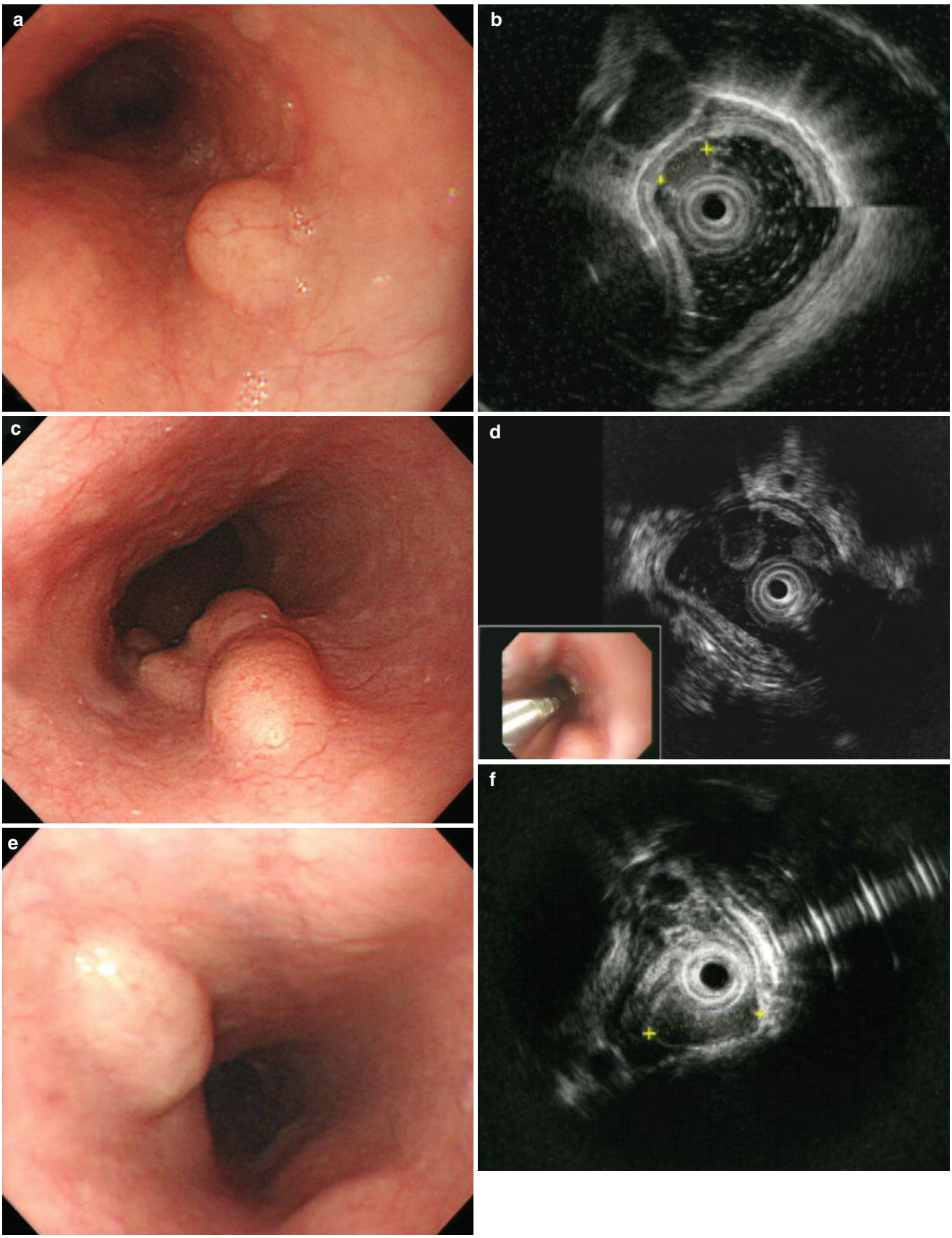

Fig. 5.2 Leiomyoma from muscularis mucosa. (**a**) Small protrusion covered with intact mucosa at mid-esophagus. (**b**) EUS shows 5 mm-sized homogenously hypoechoic mass continuous from the second layer (muscularis mucosa). (**c**) Lobulated protrusions covered with intact mucosa at mid-esophagus. (**d**) EUS shows 7 and 4 mm-sized homogenously hypoechoic lesions continuous from the second layer. (**e**) Protrusion covered with pale mucosa at upper esophagus. (**f**) EUS shows a 9.4 mm-sized homogenously hypoechoic mass continuous from the second layer

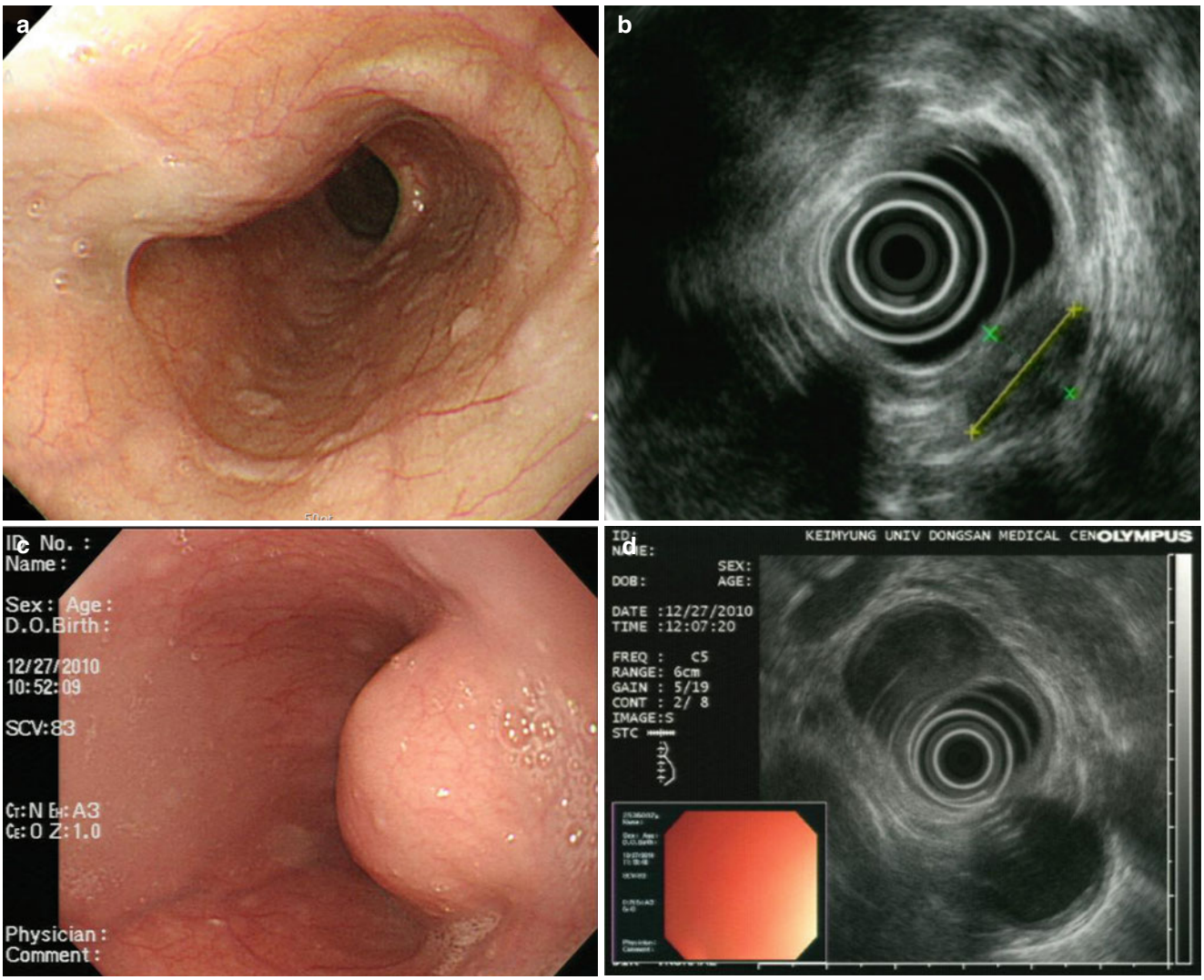

Fig. 5.3 Leiomyoma from muscularis propria. (**a**) Blunt protrusion covered with intact mucosa at mid-esophagus. (**b**) EUS shows a 16 mm-sized homogenously hypoechoic mass continuous from the fourth layer (muscularis propria). (**c**) Blunt protrusion covered with intact mucosa at lower esophagus. (**d**) EUS shows a maximally 18 mm-sized homogenously hypoechoic mass continuous from the fourth layer

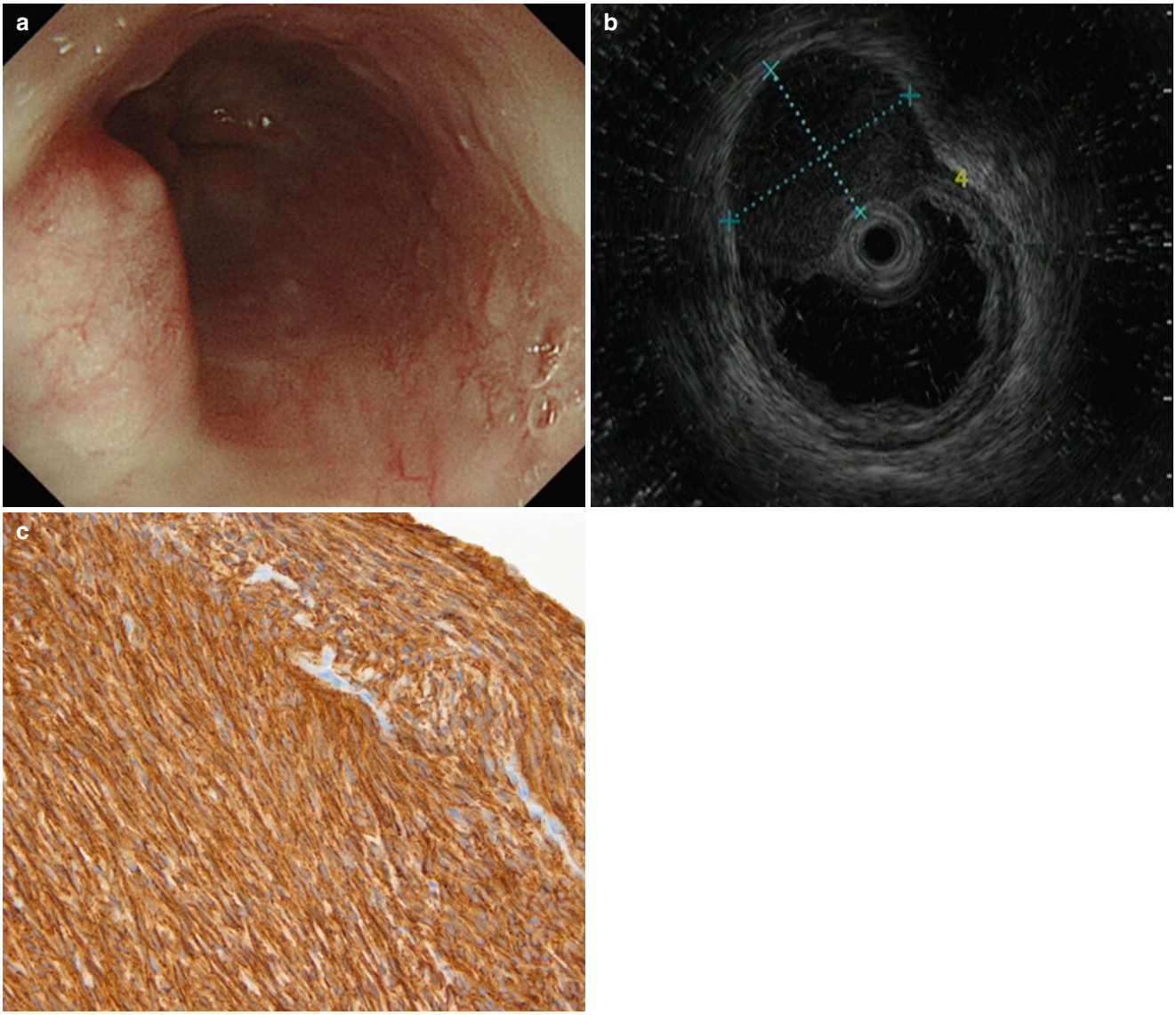

Fig. 5.4 GIST in the esophagus. (**a**) Blunt protrusion covered with intact mucosa at the distal esophagus. (**b**) EUS shows maximally 15 mm-sized homogenously hypoechoic mass continuous from the fourth layer (muscularis propria). (**c**) Immunohistochemical stains for CD117 are positive

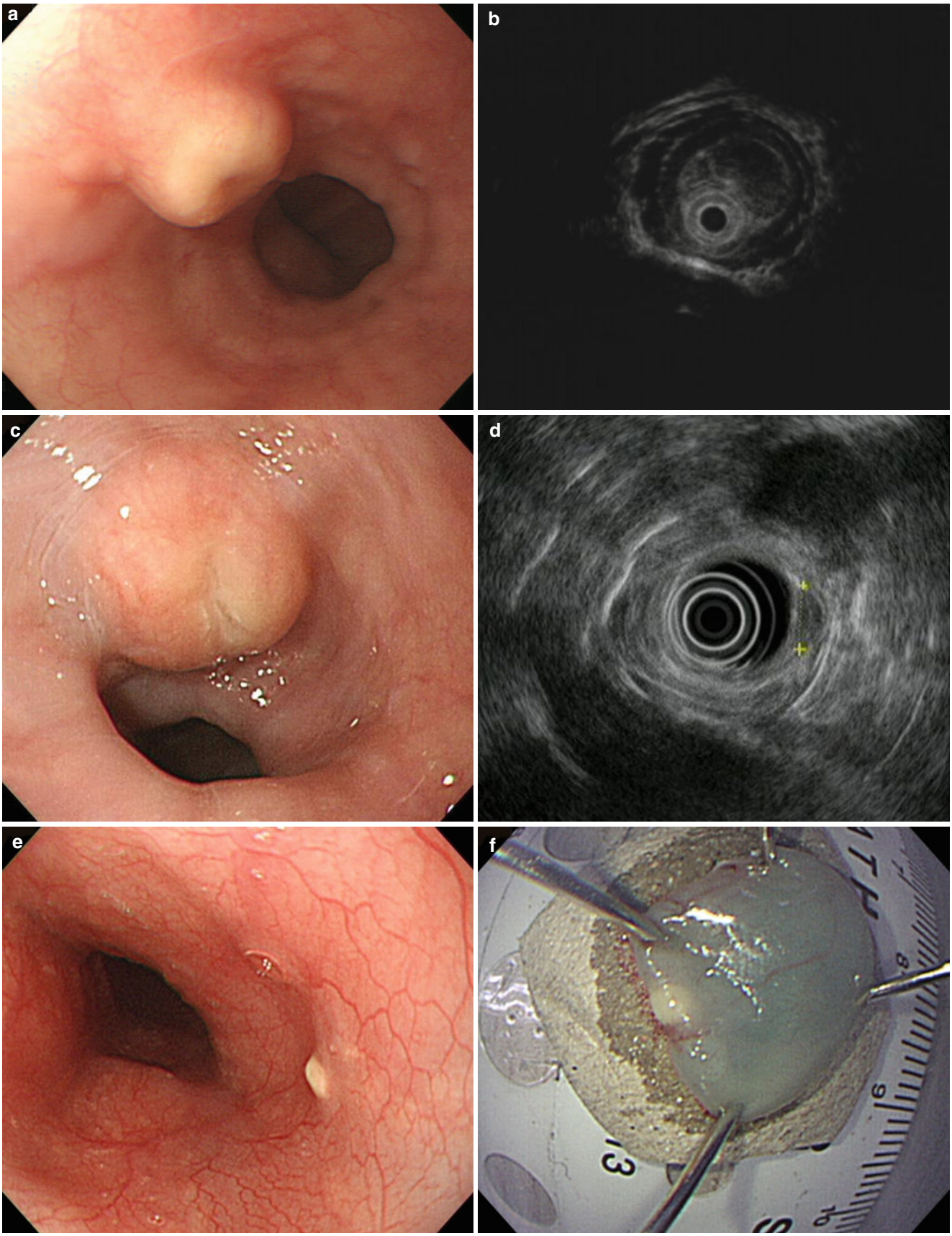

Fig. 5.5 Granular cell tumor. (**a**) Yellowish-white, molar tooth-shaped protrusion covered with intact mucosa at the mid-esophagus. (**b**) EUS shows an 8 mm-sized homogenously hypoechoic mass located within the submucosal layer. (**c**) Another yellowish-white, molar tooth-shaped protrusion covered with slightly inflamed mucosa at the mid-esophagus. (**d**) EUS shows a 12 mm-sized homogenously hypoechoic mass located within submucosal layer. (**e**) An approximately 3 mm-sized, confirmed granular cell tumor at upper esophagus with punch biopsy. (**f**) Endoscopic mucosal resection

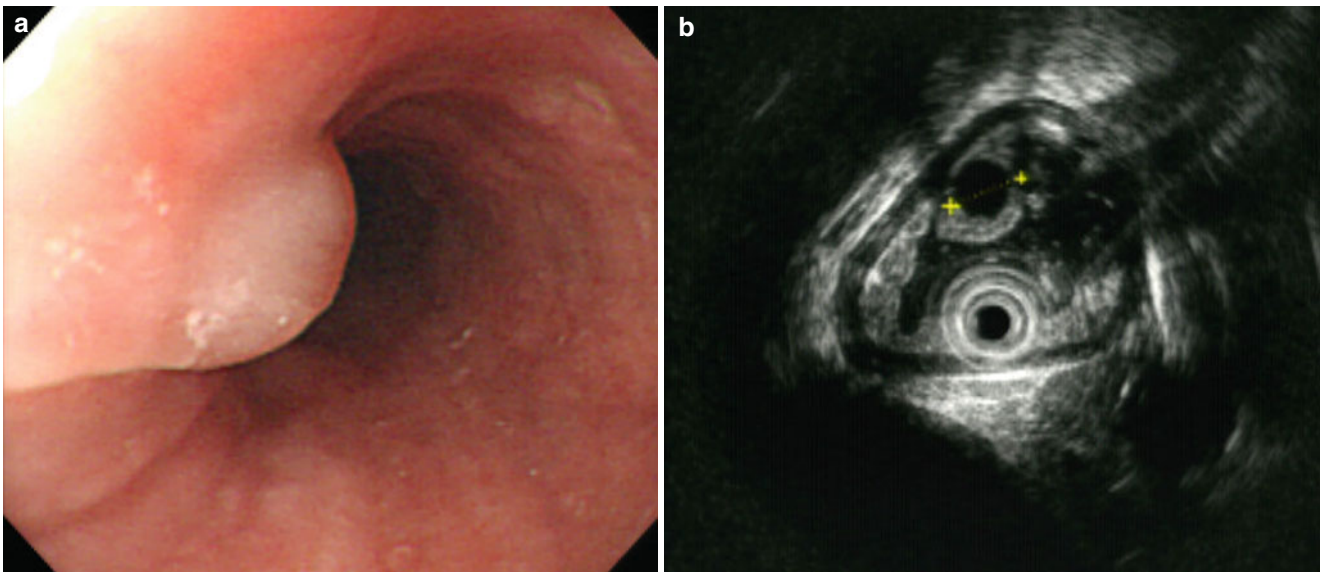

Fig. 5.6 Submucosal cyst. (**a**) Small soft mass covered with intact, transparent-looking mucosa. (**b**) EUS shows a 4.7 mm-sized anechoic lesion within the submucosal layer

5.3 Lipoma

In contrast to the stomach or small intestine, lipoma is very rare in the esophagus. On endoscopy, it shows pale or yellowish-colored soft mass (Fig. 5.7). Homogenously hyperechoic mass located in submucosal layer on EUS strongly suggests lipoma.

5.3.1 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 nodular, soft, and bluish-red protrusions on endoscopy (Fig. 5.8) [5]. When pressed with biopsy forceps, they show pale discoloration. Simple phlebectasia in submucosal layer is a nontumorous condition occasionally misdiagnosed as hemangioma because both lesions can show bluish protrusions covered with intact squamous mucosa on endoscopy (Fig. 5.9).

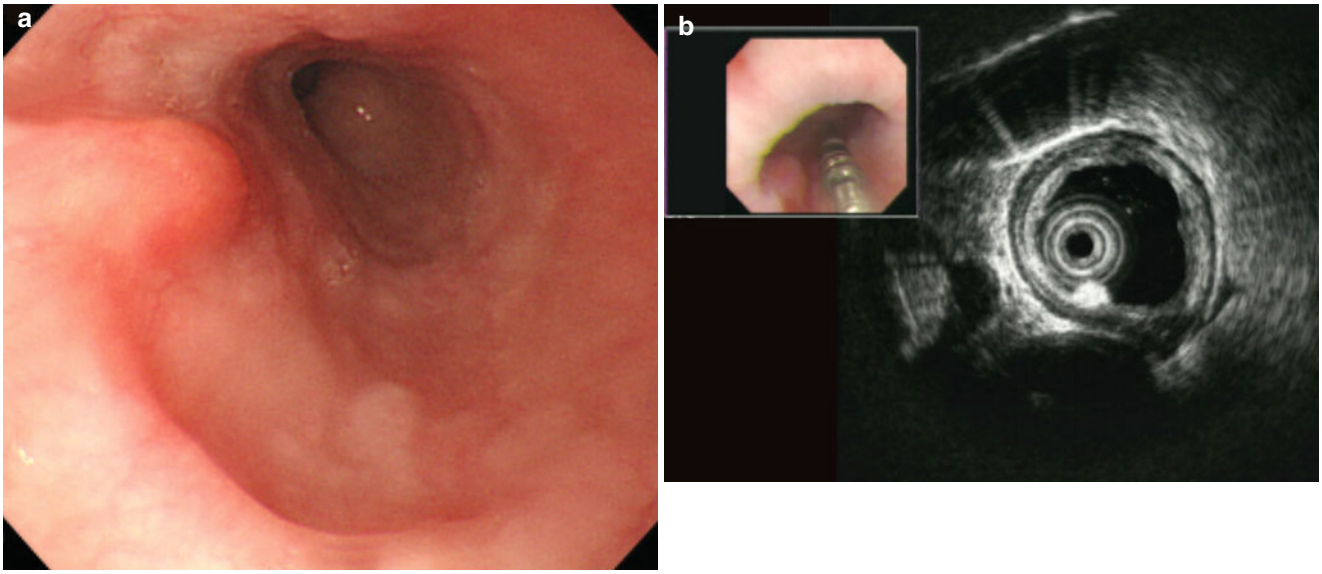

Fig. 5.7 Lipoma. (**a**) Small yellowish-colored soft mass at the mid-esophagus. (**b**) EUS shows a 4 mm-sized hyperechoic lesion within the submucosal layer

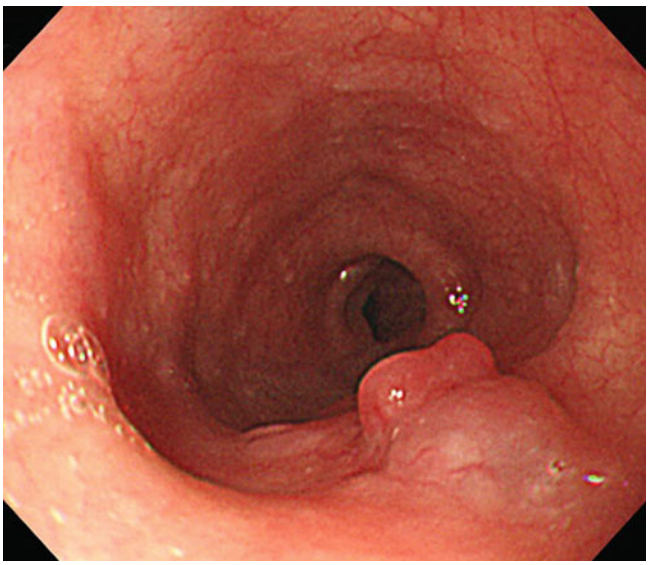

Fig. 5.8 Capillary hemangioma. An approximately 1 × 1.5 cm-sized, soft, and bluish-red protrusion is noted at the mid-esophagus

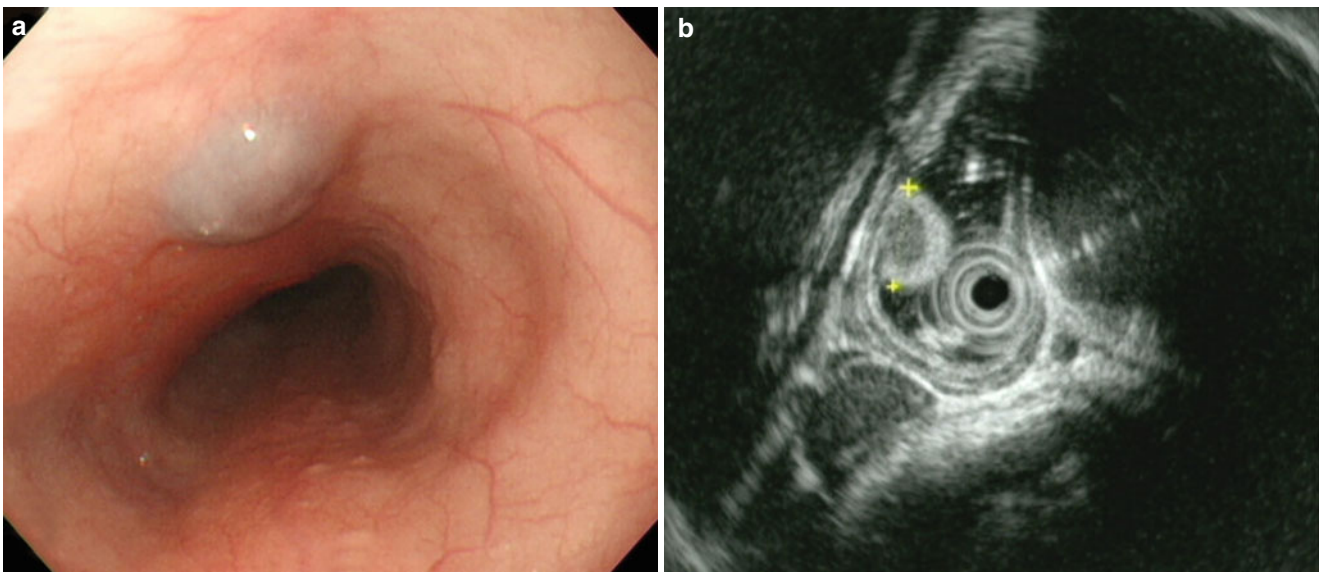

Fig. 5.9 Phlebectasia. (**a**) Upper GI endoscopy shows bluish protrusion less than 1 cm in diameter. (**b**) EUS shows a 6 mm-sized hyperechoic mass within the third layer and also shows internal hypoechoic focus

Interesting Case

A 45-year-old male visited a health promotion center for an upper GI endoscopy screening. He had no specific GI symptoms. He was a nonsmoker and social drinker. During the procedure, multiple yellowish granular spots were observed at the mid to distal esophagus (Fig. 5.10). Pathologic review of the biopsy specimen taken from the largest lesion revealed several lobules of the cells with sebaceous differentiation (Fig. 5.11). Therefore, he was diagnosed with heterotopic sebaceous glands in the esophagus. Since this extremely rare condition has shown no malignant potential, no therapeutic procedure was performed. During the follow-up procedure after 2 years, similar findings were observed (Fig. 5.12).

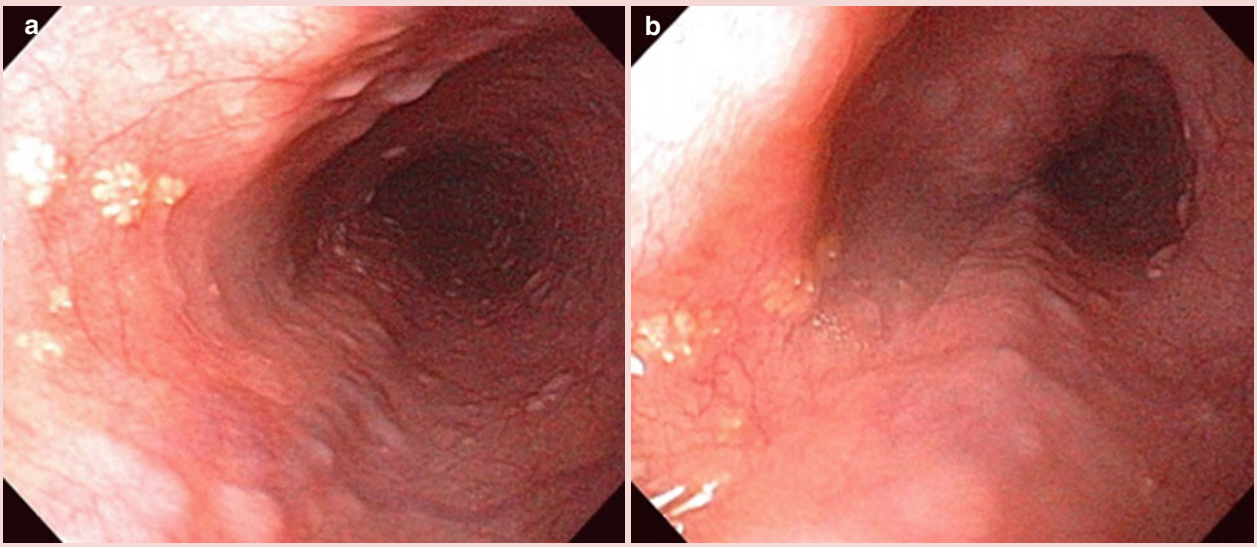

Fig. 5.10 Endoscopic finding. Multiple yellowish granular spots are observed at mid to distal esophagus. (**a**) Upper esophagus, (**b**) mid-esophagus

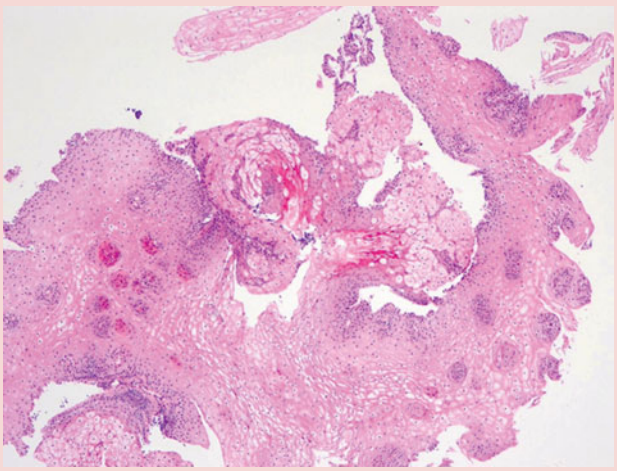

Fig. 5.11 Pathologic finding. Several lobules of the cells show sebaceous differentiation consistent with heterotopic sebaceous gland

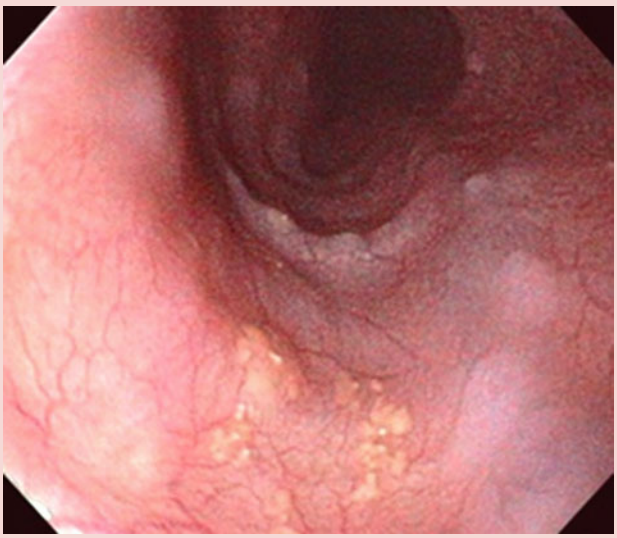

Fig. 5.12 Endoscopic finding after 2 years. Multiple yellowish granular spots are still observed

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