

MINIREVIEWS

INCLUSION CYST OF ESOPHAGUS: CASE REPORT AND REVIEW OF THE LITERATURE

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ИНКЛЮЗИОННАЯ КИСТА ПИЩЕВОДА: КЛИНИЧЕСКИЙ СЛУЧАЙ И ОБЗОР ЛИТЕРАТУРЫ

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Cysts of esophagus are unusual findings and they are included in the group of the developmental cysts of mediastinum. Various types of cysts of esophagus are much less frequent than esophageal carcinoma and considerably less frequent than some other benign esophageal tumors. We present a rare case of inclusion cyst of the upper esophagus in a male patient, followed by a review of the literature regarding this subject.

Key Words: cyst, esophagus, inclusion.

Кисту пищевода, отнесенную к группе врожденной кисты средостения, выявляют крайне редко — значительно реже, чем прочие доброкачественные новообразования пищевода или карциномы. Мы сообщаем о редком случае инклюзионной кисты верхней части пищевода у пациента и приводим обзор данных литературы о данной патологии. **Ключевые слова:** киста, пищевод, инклюзия.

Cysts that involve the esophagus, account for 10 to 15% of all primary masses of the mediastinum and, although rare, are probably the second most common benign esophageal tumor, with leiomyoma being by far the most usual [1, 2].

Esophageal cysts are classified according to their embryological site of origin, into inclusion cysts (lined by squamous or columnar epithelium, sometimes ciliated), retention cysts also called mucocoeles (arising from cystic dilatation of submucosal glands), and developmental or duplication cysts (of esophageal, bronchial, or gastric origin) [2, 3]. We report a rare case of inclusion cyst of esophagus, located in the upper portion of the organ, with symptoms of dysphagia and drown sensation.

We describe a 41 year-old man, who was referred to a gastroenterologist with a history of dysphagia, regurgitation, drown sensation, repeated pharyngitis, pain in the sternal region and left hemithorax, for the last five months. The patient did not have any history of peptic ulcer, esophagitis, vomiting, anorexia, loss of weight or of systemic disease. The physical examination revealed a well-developed patient with no apparent distress. The rest of the physical examination was noncontributory. Esophago-gastro-duodenoscopy using a flexible esophagoscope (Olympus GIF-N300) demonstrated a polyp-like, smooth mass, covered by

normal overlying mucosa, in the proximal third of the esophagus (Fig. 1), near the upper sphincter of this organ, 1 cm of diameter. Excision of the mass was performed by a polypectomy snare loop. Multiple esophageal biopsies were taken.

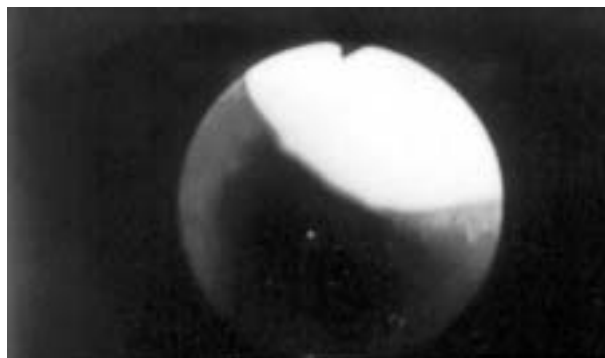


Fig. 1. Endoscopic finding of a polyp-like lesion in the upper third of esophagus

Two cysts were found in the cut surface of the mass measuring 0.4 and 0.5 cm in size respectively. Microscopically, the cysts were lined by cubo-columnar ciliated epithelium, with no evidence of a muscular layer (Fig. 2). Six esophageal biopsies showed a little mucosal hyperplasia, with scattered chronic inflammatory cell infiltrate, and no evidence of malignancy. In the mucosa of stomach and duodenum no pathological alterations were found.

Mass lesions within the mediastinum can be divided into two broad categories; congenital/acquired cyst-

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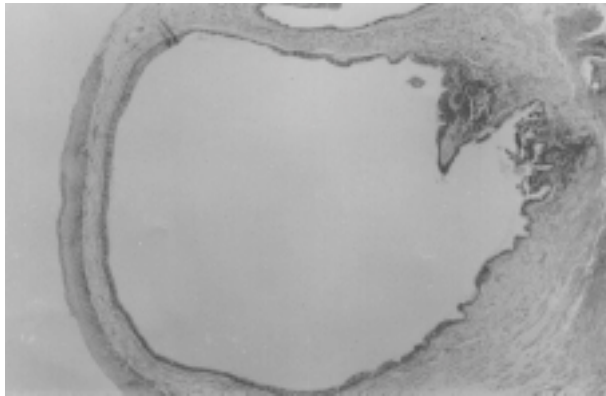


Fig. 2. Inclusion cyst of esophagus lined by columnar ciliated epithelium with no evidence of muscular layer, without containing cartilage. The overlying mucosa appears normal (H–E, original magnification x 100)

tic lesions and neoplastic tumors. Cystic lesions occur throughout the mediastinum and the majority of them are of bronchopulmonary and esophageal origin [3].

Congenital esophageal cysts are difficult to categorise because of their complex embryogenesis. They are usually classified as duplication, bronchogenic, gastric, inclusion cysts in the congenital group, as retention (mucocele), single or multiple (esophagitis cystica) cysts in the acquired group or neuro–enteric cysts. Bronchogenic cysts are more common than the gastrointestinal or esophageal duplication cysts [4].

In order to establish the incidence of esophageal cysts, we reviewed the largest series of autopsies, among those by Platcha, Attah, Scheffer and Moersh, a total of 49,196 autopsy cases, with only six being demonstrated as esophageal cysts. In other words, an incidence of one in 8.200 was found [1]. In our autopsy series of 820 cases, in a three year period (1998 to 2001), no case of esophageal mass was found.

Inclusion cysts of esophagus, as in our case, are intramurally located having epithelium of respiratory or squamous type. They are not covered by two muscle layers, nor do they contain cartilage, therefore, they can not be called as duplication or bronchogenic cysts. Due to uncertainty of etiology, the term inclusion cyst is used. The distribution, sex predominance, and symptoms are similar to those of bronchogenic or duplication cysts. We found a total of 32 cases of inclusion cysts described in the literature, including of course, surgical specimens, in both infants and adults. Among them, 66% were located in the lower third of esophagus, 24% in the middle and 10% in the upper third. Their size varied greatly, with the smallest measuring 0,5 cm and the largest 20 cm. All cysts found in the upper third presented with severe respiratory symptoms before the age of two years. In contrast, for cases involving the middle and lower third of esophagus, the average age of diagnosis was 34 years old, with dysphagia, retrosternal pain and cough being the main presenting symptoms. Cardiac arrhythmias due to heart compression have also been reported. Twenty five percent of these cysts were asymptomatic at the time of diagnosis. Inclusion esophageal cysts are never associated with vertebral abnormalities and massive mediastinal bleeding has been reported only once [5–8].

On the other hand, duplication of esophagus is the second most common duplication of the gastrointestinal tract. The cyst appears the following morphological features: it is found within the esophageal wall, it is covered by two muscle layers and it contains squamous or columnar epithelium, ciliated or not. Esophageal duplication cysts may be associated with duplications elsewhere in the alimentary tract, but never with vertebral abnormalities as is found with posterior mediastinal duplications [9].

Bronchogenic cysts of the esophagus are extremely rare in comparison with the relatively common bronchogenic cysts of the lung. Thus, a cyst is classified as bronchogenic when it is located intramurally and contains cartilage. These cysts have never been associated with vertebral abnormalities [10].

Acquired cysts may be single or multiple. When multiple, the term *esophagitis cystica* has been used. The normal esophagus contains glands in the mucosa and submucosa; most of these glands are found in the upper third of esophagus. For unknown reasons, these glands may develop into cystic growths [3].

A round regular mass in the *posterior mediastinum* on a routine chest radiograph should always suggest the diagnosis of paraesophageal cyst. Modern techniques of diagnostic imaging like endoscopic ultrasonography are very accurate in visualizing these lesions and in differentiating cysts from solid mucosal esophageal masses. In addition, the examination can establish the exact location of the mass in relation to the esophageal wall and *mediastinum* [11–113].

In general, the usual clinical course of an esophageal cyst is benign and many patients remain symptom free. Surgical resection is the treatment of choice, even when the lesion is asymptomatic. Likewise, enucleation and polypectomy by esophagoscope, represents a useful nonsurgical alternative treatment for cysts accesible through the endoscope [4, 14]. Although radiological, ultrasonographic and endoscopic studies may demonstrate the presence of a benign intramural tumor, definite diagnosis can not be established until pathological examination of the specimen is made [1, 4, 9]. An untreated cyst may enlarge and may be complicated with intracystic haemorrhage, perforation, or infection [6, 15]. Malignization of the cyst has also been reported, so removal should always be considered when a mediastinal esophageal cyst is suspected. McGregor et al [1] reported the first case of squamous cell carcinoma arising from an esophageal intramural inclusion squamous epithelial cyst in a 65 year old black man. Tapia et al [9] reported a case of squamous cell carcinoma arising in the partial duplication of the distal esophagus. Santacroce et al [16] described the unusual association of an early adenocarcinoma on short tongues of Barrett's esophagus with an underlining esophageal cyst.

In conclusion, the cysts of esophagus represent a rare finding and unmenacing growths, which rarely cause severe symptoms. Although their preoperative diagnosis is fairly difficult, once the cyst is removed, the patient is usually asymptomatic.

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