Case no. 11

72-year-old female

Previous medical history

For some years the patient had experienced dyspepsia, epigastric and retrosternal pyrosis (heartburn), gastro-esophageal reflux, and epigastralgia. Specific clinical examinations ordered for these disturbances diagnosed gallstones, giant sliding transhiatal hernia, and reflux-esophagitis. She thus underwent cholecystectomy, transhiatal hernia correction and Nissen-Rossetti fundoplication. We had performed the latter as an open procedure nearly five years before the observation described here.

Recent medical history

Approximately five years after the procedure cited above the patient began to experience dysphagia, for which she referred again to us.

Physical examination

The patient was in good general conditions without any overt signs of disease. The abdominal wall (continent) presented a well-consolidated xipho-umbilical post-laparotomic scar.

Diagnostic Workup

Blood tests revealed no pathologic conditions.

Radiologic examination detected dyskinetic and moderately ectasic features in the esophagus, the lower third of which showed a constricting neoformation of the lumen (Fig. 1 and 2).

Fig. 1 - Dyskinesia and dilation of the esophagus. Substenosis of the lower third of unknown origin.
Esophagoscopy confirmed the substenosis of the lower third of the esophagus due to extrinsic compression, without signs of mucosal disease (see video).

Computed tomography (CT) scan confirmed the presence of a paraesophageal mass 5-6 cm in diameter, which did not, however, impact on the nature of the lesion (fig. 3-4).
Magnetic resonance imaging (MRI) revealed what appeared to be the nonuniform structure of the neoformation, presumably cystic in nature.
The only possible diagnosis on the basis of findings was expansive paraesophageal neoformation of the posterior mediastinum. Video-assisted thoracoscopic resection was decided.

Surgical procedure (see video)

Patient is placed in lateral decubitus position with the left side down. Placement of 4 trocars, 3 of which are positioned on the posterior axillary line at intercostal space (IS) V, VI, and VII, and one on the mid axillary line at IS VI. The mediastinal pleura is opened and reveals at the lower 1/3 of the esophagus a protruding spherical formation approximately 5 cm in diameter that interrupts the muscular layer of the esophagus. Simultaneous esophagoscopy confirms that the videoscopic and endoscopic mass are one in the same: this latter, lined with normal mucosa, exerts pressure on the lumen of the esophagus, constricting it. The mass is easily compressible, upholding the hypothesis of its suspected cystic nature (see MRI). Transillumination and insufflation induced by the endoscope confirm the mass’s extrinsic origin and lack of involvement with the esophageal lumen. The neoformation is bluntly detached from the mucosal lining; on which circular muscle fibers nonetheless remain. At a certain point while resection proceeded, the mass ruptured and emitted a very dense, stringy, creamy white liquid. This confirmed the cystic nature of the formation and, given the contents’ features, the MRI findings of an inhomogeneous mass. The cyst was removed. Pneumatic check of esophageal wall integrity. Toilette of the thoracic cavity, tubular drainage, visual check of pulmonary expansion, and closing of the port sites.

Pathology

Macroscopic - The formation is made up almost exclusively of reddish-colored fibromuscular tissue, is approximately 5 cm in diameter, soft in texture, and has scattered hemorrhagic petechiae and a central cavity roughly 1.5 cm in diameter.

Microscopic - Histological examination of the material shows that it is fibromuscular tissue from the esophageal wall, virtually devoid of epithelial lining; scattered mucosa secreting glands are seen, as is a cystic cavitation covered mainly by columnar ciliated epithelium and, rarely, by pseudostratified cuboidal epithelium. Overall, these morphological characteristics are most likely indicative of an esophageal cyst due to retention.

Diagnosis: esophageal cyst

Postoperative course was regular. Postoperative radiological control (see video) showed normal esophagus function and morphology. The patient was released after a few days.

Follow up and outcome were uneventful.

Remarks

This case concerns a 72-year-old woman whom we had already seen and operated on for giant sliding transhiatal hernia, reflux-esophagitis and gallstones. These disorders, above all of the esophagus, had clearly warranted and received a full diagnostic work up at the time. These details underline the fact that when the patient first came under our observation there was certainly no evidence of the esophageal neoformation, which appeared with dysphagia only five years later.

The first question this case evokes regards the pathogenesis of the disorder: is it possible that a link exists between the previous diseases and/or the surgery performed five years earlier? Can the lesion’s histopathological features help us clarify the issue?

The lesion is situated in the submucosal layer of the lower third of the esophageal wall and emerges outwards, interrupting the muscle layers in which it is still wrapped. Indeed, the pathologist describes the lesion as a shell of muscular tissue presenting mucosa secreting glands and a cystic
formation covered by columnar ciliated and, in some areas pseudostratified cuboidal, epithelium. Even though the surgical and anatomic-pathological literature on the disease, especially in adults, is particularly meager (in view of its rarity), we can nonetheless dismiss some suspected pathogenic mechanisms, such as that of foregut (primitive) derivation: the histopathological features of the lesion, as well as the patient’s elderly age, rule out this hypothesis. Indeed, bronchogenic cysts contain isles of cartilaginous tissue, while enterogenic and duplication cysts present a well-organized 2-layer muscular structure; such characteristics were absent in our case. Furthermore, it is hard to believe that aberrant embryonic residues could manifest with such a latency at such an advanced age, even though some literature reports do document the event.

We are therefore left with the hypothesis of an acquired disorder. Indeed, the pathologist ventured a diagnosis of esophageal cyst due to retention. Cases like this have been described, albeit rarely. It would seem that such cysts may derive from the obstruction of submucosal glandular ducts (R.G. Lee - Diagnostic Surgical Pathology vol. 2° Raven Press 1994). If this were true, the affliction reported here could be considered secondary to the modifications of the esophageal mucosa induced by the previous reflux-esophagitis. The fundoplication would have resolved the esophagitis, but would not have had any effect on the more deeply underlying alterations of some glandular ducts. This, in any case, constitutes a yet-to-be-proven interpretation of an event that is rare in our experience and likewise in the literature.

From a clinical standpoint it can be said that the cyst was able to induce dysphagia, but was this a consequence of compression or of a break up of the muscular wall? The literature reports cases with infectious and hemorrhagic complications. As we have seen, the diagnosis was limited to the lesion’s identification, since it was unable to provide a clear-cut understanding of the neoformation’s constitution. Transesophageal US examination may have yielded some clues, but the procedure was not performed in the present case.

Video-assisted thoracoscopy proved to be an easy, safe and effective approach for the case’s resolution.