Esophageal duplication cyst recurrence: case report and literature review

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Abstract: Esophageal duplication cysts are rare congenital malformations, although most cases are diagnosed in childhood, in adult patients the disease is often symptomatic and requires invasive treatment. This kind of cyst can be found during endoscopy or Chest-X-Ray, but elective investigations consist in chest computed tomography (CT) scan or magnetic resonance imaging (MRI), if necessary, together with gastroscopy performed with endoscopic ultrasound (EUS). The therapeutic alternatives currently available are the surgical or endoscopic approach. In the past surgery was performed in thoracotomy, but several authors have reported cases treated with a minimally invasive approach with excellent short-term and long-term results. We present a case report of a 59-year-old Caucasian woman, affected by esophageal duplication cyst, and treated in 2003 with surgical fenestration in right triportal video-assisted thoracoscopic surgery (VATS) with complete regression of symptoms and resumption of normal nutrition. The cyst recurred in 2019 and the patient underwent further surgery with removal of the cyst in the right uniportal VATS. Post-operative course was regular, the symptoms disappeared, and no complications occurred. During follow-up, chest CT scan showed a complete pathological resolution without cystic recurrence. Currently, many authors agree that the minimally invasive surgical approach is the treatment of choice for esophageal duplication cysts. Our case report shows that VATS is efficient and safe even in treatment of surgical recurrences.

Keywords: Esophageal duplication cyst; video-assisted thoracic surgery (VATS); mediastinal cyst; case report

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Introduction

Esophageal duplication cysts are rare congenital malformations (1), represent approximately 10–15% of all foregut duplication cysts and are typically located in the right postero-inferior mediastinum. Intestinal duplication cysts included enteric duplication cysts, bronchogenic and neuroenteric cysts. It has been observed that at the end of the third week of embryo formation, the dorsal portion of the embryonic intestine elongates to form the esophagus and the ventral portion turn into the respiratory tract. In this phase, errors in the development of the foregut can occurs such as the appearance of bronchogenic cysts and esophageal duplications. Due to this embryological relationship, many experts define these lesions as foregut cysts or foregut duplication errors, without identifying the exact etiology of the mediastinal cysts (2). Epidemiological studies report an incidence of one case in 8,200 live births (3), about 70–75% of these cysts are diagnosed in children, the remaining 25–30% are diagnosed in adults. This kind of cyst can be asymptomatic and found incidentally during endoscopy or radiological investigations with Chest-X-Ray, computed tomography (CT) scan or magnetic resonance imaging (MRI) (4,5). The Majority of these cysts are diagnosed in childhood but when present in adults they have more likely symptomatic presentation due to the compression of nearby structures (6). The most common symptoms in these cases are dysphagia, chest pain

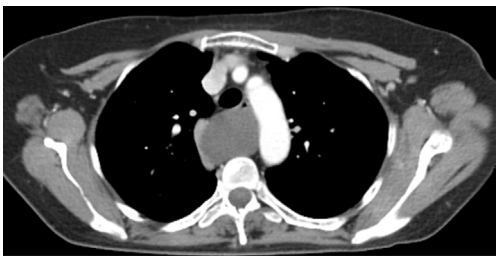

Figure 1 Chest CT axial section shows the presence of a cystic neoformation of the posterior mediastinum which deflects and compresses the esophageal lumen to the left and the Azygos vein to the right. CT, computed tomography.

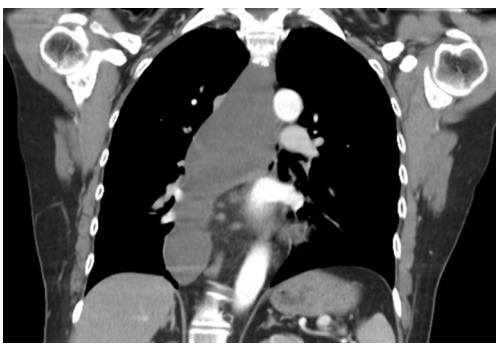

Figure 2 Chest CT coronal section shows the greater extension of the cystic lesion in the posterior mediastinum, from the diaphragm to the neck. CT, computed tomography.

and respiratory distress such as wheezing and coughing up to aspiration pneumonia. Duplication cysts can be also complicated by intracystic bleeding, perforation and infection, especially those with esophageal communication. Squamous metaplasia may also be present, but instances of malignancy are rare. The diagnosis is histological but, numerous investigations, can help to develop a differential diagnosis. Imaging studies such as chest radiographs and barium swallow studies can reveal a mediastinal mass with esophageal compression along with tracheal displacement and narrowing. Radiological examinations with chest CT scan and/or MRI allow to accurately assess dimensions, the cleavage plan of the mass with neighbouring organs and to differentiate the cystic nature from other possible tumours of the posterior mediastinum (4,7). Esophagus-gastric endoscopy along with endoscopic ultrasound (EUS), helps the differential diagnosis with the solid masses of the mediastinum, identifies the presence of any communications between the lumen of the cyst and the esophageal lumen and allows in selected cases the execution of biopsies and the possibility of endoscopic treatment with puncture and emptying of the cyst through the esophageal wall (5,8). Medical treatment of esophageal duplication cysts is not conclusive and does not improve symptoms; these cysts should be invasive treated even if asymptomatic, as they tend to enlarge over time and can cause serious complications such as obstruction, rupture, bleeding, infection or aspiration resulting in pneumonia (9,10). Invasive treatment consists in surgery with fenestration or complete excision of the cyst; however benign characteristics and the slow evolution push many people to prefer a less invasive approach with endoscopic treatment. We expose the successful case of video-assisted thoracoscopic surgery (VATS) treatment of recurrent esophageal cyst. We present the following article in accordance with the CARE reporting checklist (available at https://dx.doi.org/10.21037/aoe-2020-26).

Case presentation

A case of a 59-year-old Caucasian woman, with the appearance in 2002 of progressive dysphagia, first with solids and then with liquids, associated with significant weight loss. The patient underwent EGDS and chest CT scan with contrast, finding an esophageal duplication cysts without communication with the lumen of the esophagus. In September 2003 she underwent surgery to empty and fenestrate the cyst in right triportal VATS with complete regression of symptoms and resumption of normal nutrition. Following the reappearance of dysphagia, the patient was subjected on February 2019 to a new EGDS examination and chest CT scan with contrast which documented recurrence of the cyst as shown in *Figure 1*, with significantly increased volume compared to the previous one in 2003 (cranio-caudal extension from the root of the neck to the diaphragm for over 24 cm as shown in *Figures 2* and *3*, with a diameter of about 7.5×4 cm). Chest MRI with contrast (gadolinium) confirm the cystic nature of the lesion and the presence of clear plane of cleavage with the vertebral column but not with esophageal wall as shown in *Figures 4* and *5*. The patient underwent surgery on May 2019 with exeresis of the cyst in right uniportal VATS (intraoperative finding shown in the *Figure 6*). The duplication cyst was blunt dissected, incised, emptied and excised (as shown in the *Video 1*). It was not possible to safety remove a small part of cystic wall that was fused with esophageal wall to the upper-third and

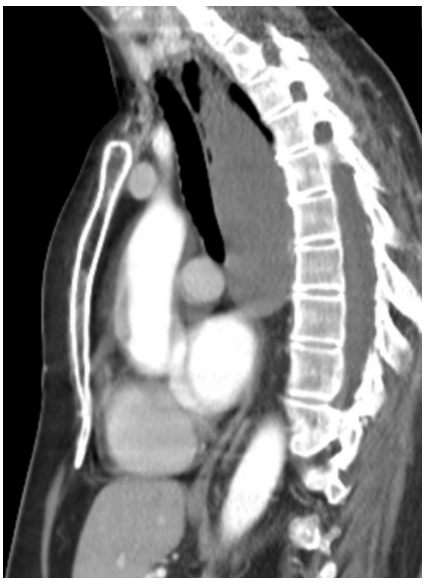

Figure 3 Chest CT sagittal section shows esophageal deviation anteriorly and to the left with cystic lesion along the spine. CT, computed tomography.

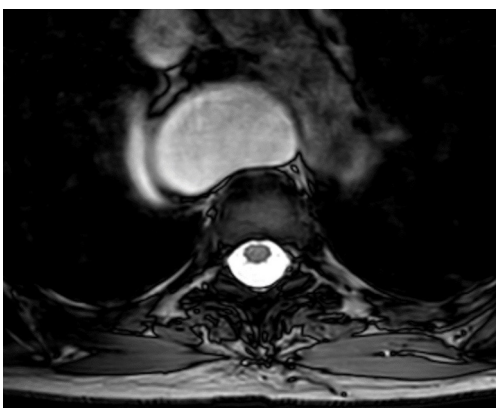

Figure 4 MRI confirms the CT findings and the cystic nature of the lesion with mucoid content. MRI, magnetic resonance imaging.

middle level, site of the previous fenestration. The integrity of the esophageal wall was confirmed by intraoperative EGDS. Histological examination confirmed the esophageal duplication cyst (pathological finding shown in *Figure* 7). Patient had a regular post-operative course, with pain easily controlled with oral therapy. Feeding with a common diet was early resumed in the first post-operative day with no episodes of dysphagia or regurgitation. Chest tube was removed on the second post-operative day and the patient was discharged. During the follow-up, no chest pain or

Figure 5 MRI sagittal section shows a clear plane of cleavage with the vertebral column but not with esophageal wall. MRI, magnetic resonance imaging.

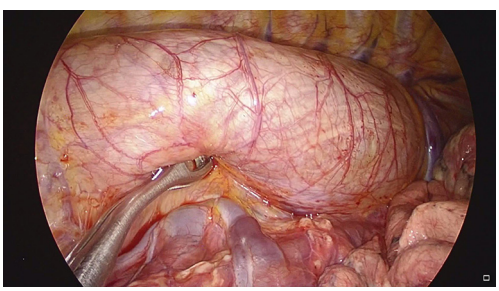

Figure 6 The surgical finding with VATS approach confirms presence of posterior mediastinal tumour, of cystic nature, growing from the esophageal wall. VATS, video-assisted thoracoscopic surgery.

dysphagia relapse was observed. Control with six-month chest CT scan showed complete pathological resolution without cystic recurrence. All procedures performed were in accordance with the ethical standards of the institutional research committee and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

Discussion

Esophageal duplication cysts represent a rare and benign form of mediastinal tumor. Cyst degeneration in neoplasm

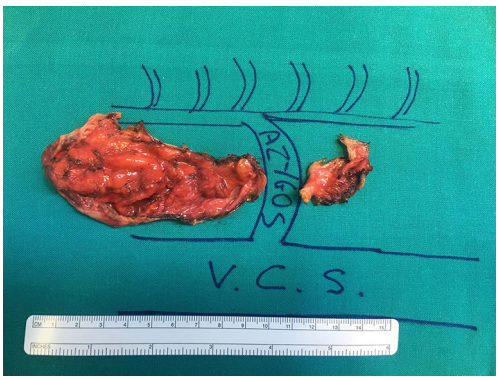

Figure 7 Surgical specimen of the esophageal duplication cyst.

has been described by some authors but it's extremely rare (11,12). Instead, complications of cysts as obstruction, bleeding, infection, or rupture are common and well described (9,10).

Symptomatic patients needed an invasive treatment of the cyst as there are no effective medical therapies. In asymptomatic patients, there are currently no clear guidelines on the treatment to be performed, but most authors agree with the indication for invasive treatment to avoid possible serious complications due to the natural pathological history. Due to the benign nature of this kind of cyst, different invasive treatments have been proposed and used. The review of literature show as many studies reports clinicals outcomes in treatments of inveterate and untreated esophageal duplication cysts. Our intent is to critically evaluate the approach to the treatment of esophageal duplication cysts by paying attention to the problem of recurrence which in many cases makes future minimally invasive approaches difficult if not impossible. The different approaches reported for treatment of this pathology consist in puncture and endoscopic emptying of the cyst, other endoscopic treatments, fenestration or resection of the cyst with thoracoscopic or robotic approach, up to traditional thoracotomy surgery. The most common endoscopic treatments consist of needle aspiration under esophageal ultrasound guide. This technique is often not recommended due to the risk of infection, the high rate of recurrence and the possible appearance of adhesions or leaks with the esophageal wall, which can complicate the pathology and make further surgical resection difficult. Some authors describe endoscopic submucosal tunnel dissection as an option to excise an esophageal bronchogenic cyst (13,14). This kind of cysts are rare, but the technique reported suggests the use of the endoscopic approach to treat esophageal duplication cysts in an effective and minimally invasive way. Other authors reported the results of endoscopic fenestration of esophageal duplication cyst performed by using EUS guidance (15,16). However, there are no studies in the literature showing long-term outcomes and complications after these procedures. For these reasons, many authors suggest that surgery is the best approach to treat esophageal duplication cyst even if asymptomatic (17-19). Surgical treatments consist of fenestration or complete removal of the cyst, usually performed with the thoracotomy technique, however, several cases of cysts removed effectively with minimally invasive technique have been reported in the literature, performed with VATS or robotic-assisted thoracoscopic surgery (RATS). After surgical fenestration or excision, whether it is performed with traditional thoracotomy or with minimally invasive technique, patients' outcomes are good both in the short term and in the long term. Recurrence is rare but has been reported (20). Complications related to esophageal cyst resection include tracheal and esophageal injuries, pseudodiverticulum development, and nerve injury or paralysis. There are many studies on surgical efficacy in children, however, few studies have investigated indications treatment and postoperative outcomes in adult population. In a case series of twenty-seven patients affected by esophageal duplication cyst and treated by surgical resection, Cioffi *et al.* showed as minimally invasive surgery guaranteed the same outcomes, a better cosmetic result, less pain, early patient mobilization, lower risk of infectious complications and early discharge (10). In a case series of nine patients, Hazelrigg *et al.* confirmed similar results (21). Other case reports have shown good postoperative results in patients treated with removal of esophageal duplication cysts with both VATS and RATS techniques (17,19,20,22). Currently, most authors treat recurrences of the cyst with a traditional thoracotomy surgery due to the presence of adhesions, possibility of leak, and local altered anatomy in site of previous treatment, with difficulty or impossibility to recognize a clear limit between esophageal wall and cystic wall. Our case report demonstrates how thoracoscopic treatment can be valid and safe even in cases of recurrences after previous invasive treatments.

Conclusions

This kind of benign pathology of the thoracic esophagus represents a surgical challenge. Patients, at the same time requiring safe, effective and minimally invasive treatments,

have currently shifted attention to techniques performed with an endoscopic approach. In the past, previous chest surgery or failure of minimally invasive approach required traditional thoracotomy surgery. This case shows that minimally invasive surgery, if performed by expert operators, can be used safely even in case of recurrent disease after previous surgery. In the current context, esophageal cystic resection performed with minimally invasive surgery such as VATS technique, represents the suggest therapeutic option for the lower risk of recurrence, best esthetical results, less post-operative pain and the possibility to treat patients underwent previous surgery.

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Footnote

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