Complicated Esophageal Duplication Cyst in Adult: A Case Report and Review of the Literature

Bassem Nasr, Fethi Derbel, Mohamed Ben Mabrouk, Jaafar Mazhoud, Waad Farhat, Malek Barka, Ben Ltaifa Afef, Mohamed Azzaza, Ben Abdelkader Atef, Youssef Chaker, Ali Ben Ali

INTRODUCTION

Esophageal duplication cysts (EDCs) encountered within the gastrointestinal duplication cysts (GDCs) are rare embryonic malformations. The incidence of GDCs, which are mostly diagnosed in early childhood, is 1:100,000, with only about 160 cases described in adults. The frequent localizations are the ileum (50%) and esophagus (25%)[1].

Up to 80% of esophageal duplication cysts are diagnosed in childhood: respiratory distress or nutritional difficulty due to mass effect. The majority of patients develop symptoms in the adolescence period, whereas symptomatic cysts are rarely encountered in adulthood (<7%) and the diagnosis is made most often from an incidental finding on the chest radiograph[2,5].

In adults, although these cysts are usually asymptomatic, they can cause dysphagia or back pain or be complicated by intracystic hemorrhage, rupture, pulmonary or esophageal hemorrhage, and infection[2,3]. Malignant transformation has also been reported[2,4,6]. Complete surgical excision is the standard treatment[7,8]. Most cysts are resected due to active symptoms. However, cysts that are incidentally found and asymptomatic are surgically resected to prevent potential complications from untreated duplication cysts[9-11].

Treatment of symptomatic esophageal cysts can be either surgical or endoscopic. Surgical resection of the cyst is usually carried out in childhood or in adults, in cases of lesions impossible to treat endoscopically. Surgical treatment is currently moving from thoracotomy to less-invasive procedures, such as video-assisted thoracoscopic surgery and to endoscopic treatments which, however, still remain challenging interventions[7,10,12].

We report a case of squamous cell carcinoma arising within an esophageal duplication cyst in adult with acute clinical presentation (perforation into thorax), treated with surgical resection.
CASE REPORT

A 30 years old man, without significant past medical history, presented a 7 day history of right sided pleuritic chest pain and fever, he had a vague abdominal pain.

On examination, he was febrile (38.5°C), polypnoeic, and the examination of the chest showed signs of consolidation at the right base.

The white cell count was 21000/ml and the rate of CRP was at 240 mg/L.

A thoracic CT showed an air containing lesion with fluid level associated with pleural effusion (Figure 1) suggesting complicated hydatid cyst of the lung associated with a hiatal hernia. A barium swallow showed 2 posteriors fluid levels which communicate with esophagus (Figure 2) suggesting complicated hiatal hernia containing stomach.

So the patient underwent a surgery. First time, the abdomen was open via medium laparotomy: the stomach and the eso-gastric junction was normal, we didn't found a hiatal hernia. Second time, the chest was open via a right posterolateral thoracotomy and entered through the sixth intercostal space.

A cystic esophageal duplication complicated by perforation and bronchiolar fistulae associated with a pyothorax were found (Figure 3 A, B, C and D).

The duplication was resected, the esophagus and the fistulae were sutured and the pyothorax was drained (Figure 4).

Histological examination of the resected piece showed a squamous cell carcinoma invading the different layers of the esophageal wall (Figure 5).

It’s a squamous cell carcinoma of a cystic communicating duplicity of the thoracic esophagus complicated by perforation.

The immediate postoperative period was favorable. The patient was extubated on day 2 postoperatively. Five days after, the patient developed a respiratory distress, so, he was intubated. A chest CT scan showed large pleural effusion abundance. The patient was reoperated. A pyothorax was found and evacuated, and the esophageal sutures were sealed. The patient developed a septic shock and died on day 10 postoperatively.

DISCUSSION

Duplications of the alimentary tract are infrequent anomalies that can affect any portion of the gastrointestinal tract[9].

Oesophageal duplications are the second most common form of enteric duplication after the ileum ones. They are tubular or cystic structures of variable size, which arise from the posterior mediastinum and lie in a retropleural location[17,19].

In our case, the duplication of the oesophagus was cystic.
A review of 96 patients with 101 alimentary tract duplications reported by Holcomb et al(27) showed that twenty-one duplications were confined to the oesophagus and all of these duplications were cystic. Seven of these EDCs contained ectopic gastric tissue.

For an esophageal cyst to be classified as duplication, it must meet the following criteria: (1) The cyst must be within or attached to the esophageal wall, (2) it must be covered by 2 muscle layers, (3) and the lining must be squamous, columnar, cuboidal, pseudostratified, or ciliated epithelium(7).

Most of the cysts were reported to arise in the lower oesophagus, with only 23% occurring in the upper third(9). In this case, the EDC was present in the lower third of the oesophagus.

Clinical presentation varies, with the most common presenting symptoms being dysphagia, epigastric discomfort, and retrosternal pain(11,13,18).

Karman I et al(19) reported a rare case of EDC revealed by back pain. Respiratory symptoms (chronic cough, recurrent respiratory infections) or tracheal compression are more common with upper oesophageal cysts and manifest in early childhood(6,10,13,16).

Obasi PC et al(20) presented 2 pediatric cases with new onset shortness of breath and chest pain. Acute onset is due to complication by intracystic hemorrhage, perforation, and infection, especially in those with oesophageal communication(21).

In our case, the patient was completely asymptomatic until the onset of a right sided pleuritic chest pain, fever and a vague abdominal pain. A similar case to ours was reported by Grewal(19). Surgical exploration found a cystic esophageal duplication complicated by perforation and bronchial fistulae associated with a pyothorax.

Malignant transformation is an extremely rare event within oesophageal cysts. The most common histology is an adenocarcinoma, though squamous cell carcinoma has also been reported to arise within foregut cysts(9). In our case, Histological examination of the resected piece showed a squamous cell carcinoma invading the different layers of the esophageal wall.

Diagnosis can be established incidentally from routine chest radiographs or with a barium esophagram showing external compression of the oesophagus. However, computed tomography has the advantage over conventional diagnostic procedures because it demonstrates the cystic nature of the mass and its relationship to adjacent structures in a noninvasive manner. It permits simultaneous visualization of the surgical field, (2) greater range of motion with tremor filtration and motion scaling(23).

Obasi et al(21) presented 2 pediatric patients who underwent successful surgical resection of esophageal duplication cysts via RATS using the da Vinci surgical system. They had no operative complications but did have excellent postoperative outcomes including decreased pain and early patient discharge.

**CONFLICT OF INTERESTS**

There are no conflicts of interest with regard to the present study.

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**Peer reviewers:** Lars Grenacher, Professor, Department of Diagnostic and Interventional Radiology, University of Heidelberg, Im Neuenheimer Feld 110, 69120 Heidelberg, Germany.