Clinical Case Report

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Esophageal duplication cyst with hemivertebrae

A case report and literature review

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Abstract

Background: Esophageal duplication cysts (EDCs) are rare congenital anomalies that can be associated with symptomatic spinal abnormalities, but presentations due to EDC symptoms are rarely found in the presence of spinal abnormalities.

Case summary: A 6-month-old infant weighing approximately 5.0 kg presented with a 2-month pulmonary infection and more recent difficulty swallowing and nutritional intolerance that did not improve with medical treatment. Contrast-enhanced chest computed tomography showed a well-defined, mediastinal, homogeneous, low-density cystic mass of $11.9 \times 5.5 \times 5.1$ cm, compressing the liver and bending the trachea forward. Hemivertebrae were present (T4 and T3). Diagnostic laparoscopy was performed, but was converted to open surgery. After ensuring that the cyst was not within the abdominal cavity, thoracotomy was performed, and the cyst was completely resected. Pathophysiological examination revealed an EDC. The patient recovered well, without symptoms 6 months later.

Conclusions: Overall, noninvasive imaging and diagnostic procedures may not be sufficient to define the exact location of an EDC. Although hemivertebrae were present, they were asymptomatic and did not require treatment; only the EDC induced nonspecific symptoms that disappeared after surgery.

Abbreviations: CT = computed tomography, EDC = esophageal duplication cyst.

Keywords: esophageal duplication cyst, hemivertebrae, infant

1. Introduction

Congenital esophageal duplication cysts (EDCs) are rare congenital anomalies and can be associated with other congenital anomalies such as small intestinal duplication, esophageal atresia distal to the duplication, and tracheoesophageal fistulas.[1] They are often associated with spinal abnormalities, including scoliosis and fusion, which help achieve correct diagnosis.[1] Neuroenteric cysts have been reported to be associated with butterfly vertebra, spina bifida, and hemivertebrae.[2] The incidence of congenital esophageal cysts is estimated at 1:8200, with 2:1 male predominance.[3,4] Patients with esophageal cysts usually present with pulmonary infection, respiratory distress, difficulty swallowing, and nutritional intolerance due to compression and infection. Most of these cysts are benign, with asymptomatic anomalies that occur during foregut formation; neurological complications are usually the reasons for initial investigation. The preferred treatment is complete surgical resection, and the operation is not difficult. Physicians should be familiar with this disease. We herein report a rare case of a large esophageal duplication cyst with hemivertebrae with an initial presentation of esophageal symptoms.

2. Case report

A 6-month-old Uighur Chinese male infant presented with a history of pulmonary infection for at least 2 months. He had received treatments for pneumonia, using aerosolized second-generation antibiotics. Difficulty swallowing and nutritional intolerance appeared after the onset of pulmonary infection. The infant was born at 38 weeks (3200 g and Apgar score of 9), through caesarean section to a second-time pregnant mother. Antenatal and perinatal histories were uneventful. There was no family history of congenital diseases. On physical examination, the right lung had voiceless percussion, and wheezing could be heard in both lungs. The infant had nutritional intolerance and funnel chest. At admission, chest X-ray was performed, and the patient was diagnosed with bronchopneumonia. The electroencephalogram, limb electromyogram, cardiac ultrasound, blood count, and routine biochemical parameters were all normal.

Barium swallow examination did not show substantial luminal narrowing or trace pressure (Fig. 1A). The day after admission, 3-dimensional computed tomography (CT) reconstruction demonstrated a cystic mass located in the posterior mediastinum. This mass displayed well-defined margins and was fluid-filled, but free of air-fluid levels. The cyst was $11.9 \times 5.4 \times 5.1$ cm. The average

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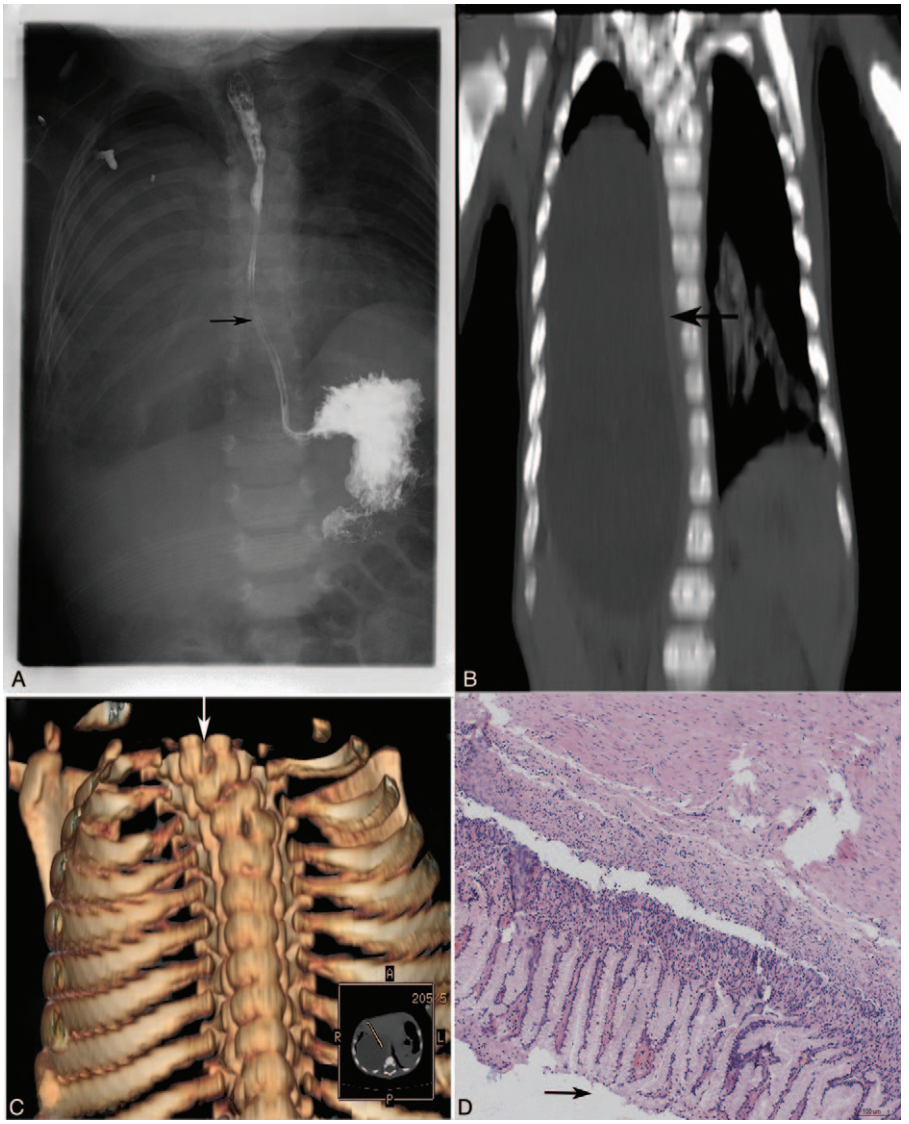

Figure 1. Imaging findings of a case with esophageal duplication cyst with asymptomatic hemivertebrae. In this 6-month-old male infant, barium swallow examination was inconclusive (A). Computed tomography scan showed an esophageal duplication cyst (arrow), whose coronal is depicted. The cyst was 11.9 × 5.4 × 5.1 cm, with an average wall thickness of 0.5 cm, compressing the trachea, right main bronchus, right inferior lobe (anteriorly), and liver (B). Three-dimensional computed tomography reconstruction showed hemivertebrae located in the upper thoracic spine (T4 and T3) (C). Histopathological examination after hematoxylin and eosin staining showed gastrointestinal-type mucosa (magnification, ×40) (D).

wall thickness was 0.5 cm and increased from the extra-pleural space or enterocelia, isolated within the esophagus. The cyst compressed the fore lying trachea, right main bronchus, right inferior lobe (anteriorly), and liver (Fig. 1B). The hemivertebrae were located in the upper thoracic spine (T4 and T3) (Fig. 1C).

Because the cyst was oversized and compressed both the right main bronchus and liver, we performed a diagnostic laparoscopy 7 days upon admission, but this remained inconclusive regarding the exact localization and extent of the cyst. Laparoscopy was converted to open surgery, and the cyst was confirmed to protrude in the abdominal cavity from the thoracic cavity. Then, antero-lateral thoracotomy was performed. The cyst was dissected from the adjacent tissues and completely excised. We withdrew about 150 mL of a clear, jelly-like fluid from the cyst. Closed tube drainage was performed for 3 days, and discontinued when chest X-ray showed improvement. The patient was discharged 6 days later when chest X-ray showed improvement of bronchopneumonia, without change in the condition of the spine. The infant recovered uneventfully. Histopathological examination after hematoxylin and eosin staining of the cyst showed gastrointestinal-type mucosa with well-developed muscularis propria and serosa (Fig. 1D).

Since neurological examinations were normal, no treatment for hemivertebrae was undertaken at that time. The infant showed normal growth and development at 6-month follow-up. No X-ray was performed at that time.

Informed consent was obtained from the patient's parents.

3. Discussion

Esophageal duplication cysts are rare, benign mediastinal masses that occur in the 6th week of embryonic life.[5] They can be associated with symptomatic spinal abnormalities, but presentations due to esophageal duplication cyst symptoms are rarely found in the presence of spinal abnormalities. We reported the

Table 1

Demographic data, clinical and image findings, surgical interventions, and outcomes in selected cases with esophageal duplication cysts.

Case	Sex	Age	Country of study	Clinical symptoms	Imaging findings	Final diagnosis	Surgical intervention	Outcome	Reference
1	Female	54 y	Czech Republic	Dysphagia and a painful swallowing	10 × 4 × 4 cm infected cyst with a dense content corresponding to the pus localized in the lower posterior mediastinum	Inflamed mediastinal esophageal duplication cyst	Partial laparoscopic resection	Favorable	Zdenek et al[15]
2	Female	21 y	Japan	Repeated chest pain	Meditational mass at caudal side of tracheal carina (maximum diameter, about 3 cm).	Mediastinal abscess	Thoracoscopy	Favorable	Takemura et al[19]
3	Female	12 y	The United States	Shortness of breath	2 × 1.5 cm cystic lesion	Esophageal duplication cyst	Robotic-assisted thoracoscopic resection	Favorable	Obasi et al[20]
4	Male	15 y	The United States	History of spina-bifida, hydrocephalus, VP shunt, and back pain	2 × 2 cm retrocaval right-sided thoracic mass	Esophageal duplication cyst	Robotic-assisted thoracoscopic resection	Favorable	Obasi et al[20]
5	Male	35 y	India	Gradually progressive dysphagia	2.6 × 1.6 cm heterogeneous echotextured lesion with anechoic component in the lower esophagus	Esophageal duplication cyst	Surgical excision	Favorable	Chaudhary et al[17]
6	Male	3 y	China	Intermittent fever of acute onset and dry cough	0 × 5.4 × 5.8 cm oval-shaped, cyst-like tumor located in the extrapleural space	Double esophageal duplication cysts	En bloc resection	Favorable	Zhang et al[18]
7	Male	6 mo	Switzerland	Slowly fed with a bottle; otherwise no symptoms	2.1 × 1.5 × 2.6 cm cystic mass in the left posteroinferior mediastinum	Intrathoracic esophageal cystic duplication	Transhiatal laparoscopy	Favorable	Mengu Ma et al[11]
8	Female	Neonate	Poland	Congenital thoracic cystic mass	Cystic mass measuring 35 mm in proximity to right lung base	Esophageal duplication cyst	Thoracoscopic cyst excision	Favorable	Cuch et al[12]
9	Male	24 y	Oman	No apparent symptom	3.7 × 2.3 × 1.5 cm well-defined cystic lesion	True intramural esophageal duplication cyst	Video-assisted thoracoscopic surgery (VATS) excision	Favorable	Al-Riyami and Al-Sawafi[14]
10	Female	18 y	India	Fever, cough, and breathlessness	Tubular cystic lesion measuring 9.1 × 3.1 × 2.9 cm in the left paratracheal region	Thoracic esophageal duplication cyst	Antibiotics	Conservative management	Tomar et al[6]
11	Male	30 y	India	Gradually progressive dysphagia	3.5 × 2.3 × 3 cm well-defined homogenous hypodense cystic lesion	Esophageal duplication cyst	Complete surgical excision of the cyst	Favorable	Sonthalia et al[13]
12	Male	16 mo	United Arab Emirates	Diaphragmatic hernia	2.5 × 1.3 cm cyst in distal esophagus	Distal esophageal duplication cyst with gastro-esophageal reflux disease	Left thoracotomy, excision of the duplication cyst and thoracic fundoplication	Favorable	Jan et al[16]
13	Male	6 mo	China	Esophageal symptoms	11.9 × 5.5 × 5.1 cm cystic mass	Esophageal duplication cyst	Thoracotomy	Favorable	This study

case of a 6-month-old infant who presented with respiratory tract infection symptoms, which were later demonstrated to be caused by a large esophageal duplication cyst accompanied by asymptomatic hemivertebrae. This case suggests that noninvasive imaging and diagnostic procedures may not be sufficient to identify the exact localization of esophageal duplication cysts. Although hemivertebrae were present, they were asymptomatic and did not require treatment; only the esophageal duplication cyst induced nonspecific symptoms that disappeared after surgery. A previously published case was also incidentally diagnosed with esophageal duplication cyst after being investigated for respiratory tract symptoms, but this case had no spinal deformity.[6]

Esophageal duplication cysts are rare (incidence of 1:8200).[7] They can be associated with spinal abnormalities, but the patients usually present with symptoms of spinal abnormalities and are incidentally diagnosed with esophageal duplication.[1,2,4,8] In the present study, the esophageal duplication cyst was very large, which led to an original misdiagnosis of common bile duct cysts or pancreatic pseudocysts, and laparoscopic surgical exploration was then performed. Fortunately, the finally diagnosed esophageal duplication cyst only required minimal surgery and could be performed during the same operating time.

Only 10% of esophageal cysts interact with the lumen of the esophagus.[9] There have been very few cases in which the cyst is connected to the lower half of the esophagus, and large cysts of this type usually compress the lower half of the esophagus.[10] In the present case, barium swallow examination was inconclusive, which led to an incorrect diagnosis. The cyst was completely and successfully resected from the adjacent tissues by open surgery. A previous case was reported to have been operated by laparoscopy,[11] but laparoscopy was inconclusive in the present case. The exact diagnosis could be made only after pathological examination. Such cysts can lead to complications such as hemorrhage, infection, and malignancies, and are found in both genders, virtually all ethnicities, and a wide range of ages, as shown in Table 1.[6,11–20] In the current case, the cyst was very large, and might have gone unnoticed because it was initially asymptomatic until it reached a large volume that obstructed the respiratory airways. In addition, the child might have not been followed up closely after birth by an experienced pediatrician. This stresses the importance of regular postnatal examinations. Complete surgical excision is typically curative because recurrence is rare.[6,21] The present case recovered well.

Because the neurological functions were normal and the patient was asymptomatic, no spinal surgery was undertaken. The infant had normal growth and development at 6-month follow-up.

In conclusion, esophageal duplication cysts are relatively rare and may have no specific symptoms. A combination of multiple

medical imaging techniques, such as esophagography, CT scan, and barium swallow examination can assist in diagnosing esophageal duplication cysts and defining their relationships with the neighboring anatomy. Curative surgery is not difficult and is needed to provide strong clinical evidence for diagnosis. Complete surgical excision is typically curative because recurrence is rare.

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