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Esophageal Duplication Cyst: An unusual cause of Acute Chest Pain

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ABSTRACT
Chest pain is considered a known presentation on emergency department (ED) with variety of differential diagnosis ranging from life threatening to simple causes mostly cardiac, pulmonary, musculoskeletal, gastrointestinal disorders, somatization disorders and other rare causes. This wide differential become a challenge to the emergency physician sometimes to have clear diagnosis on the short ED stay. Although esophageal duplication cysts in adults are a rare entity and are mostly asymptomatic, acute presentation is possible and emergency physicians should understand the complications of this condition. In this report we describe an unusual cause of chest pain and shortness of breath in an adult that was related to this rare embryonic anomaly.
Key words: *Chest pain, Esophageal duplication cyst , Embryonic Anomaly,*

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INTRODUCTION

Chest pain is a common and challenging to emergency physician. Although mostly the diagnosis is not life threatening, emergency physicians must distinguish between those who need urgent intervention of a serious problem such as myocardial infarction and those with more benign entities. Clinical judgment using The basic clinical tools of history, physical examination are used to be the most important initial step on meeting this challenge, plus all varieties of diagnostic modalities have been developed to assist in diagnosis and risk stratification including electrocardiogram (ECG) , cardiac markers , risk scores, stress testing, and noninvasive imaging of the heart. Chest pain in emergency department (ED) that need evaluation was found to be around 8% to 10% of the 119 million ED visits annually [1].Esophageal duplication cysts are rare congenital anomalies that can be a cause of recurrent chest pain that can be missed easily as the definitive diagnosis can't be made in emergency department based on the usual diagnostic tools [2]. Diagnosis of an esophageal duplication cyst is usually made in infancy and childhood while investigating for respiratory distress and feeding difficulties [3]. In adults, esophageal duplication cysts are usually asymptomatic, and the diagnosis can be suspected most often from an incidental finding on a chest radiograph and confirmed by barium swallowing studies as needed [4]. Although the anomaly itself is not life-threatening, complications such as bleeding, infection, and mass effect are known to occur during the natural course of the disease [5]. These complications can be devastating, and the emergency physician should be able to recognize them for immediate action. Complete surgical excision is the standard treatment for symptomatic esophageal duplication cyst [6]. We describe the rare presentation of a symptomatic esophageal duplication cyst in an adult that was first thought to be a vascular pathology.

CASE REPORT

A 29-year-old woman, previously known to be healthy, presented with progressive chest pain and shortness of breath over the previous 2 months. The patient reported that her symptoms were aggravated when she was nursing or caring for her first child, who was 4 months old.

The patient presented herself to our emergency department because her symptoms worsened over the last 3 weeks conveniently timed with her starting to use oral contraceptive pills (OCP). Upon arrival to the hospital, the patient had stable vital signs except for mild tachycardia (104 beats/min), and no gross abnormality could be detected by chest or general physical exam. The patient's initial electrocardiogram (ECG), cardiac enzymes, were all unremarkable. A chest X-ray showed opacity in the right mediastinal area that was thought to be an enlarged pulmonary trunk or a vascular abnormality (Fig. 1). Thus, a computed tomography (CT) angiogram was requested mainly to exclude pulmonary embolism or any other vascular pathology.

The CT angiogram showed a well-defined hypodense lesion of fluid located in the subcarinal area and extending to the posterior mediastinum. The lesion, which measured approximately 5.6 × 5 cm at its greatest diameter, displaced the pulmonary veins anteriorly and was inseparable from the esophagus. No intraspinal extension was present and the adjacent vertebrae were grossly unremarkable. No evidence of pulmonary embolism was observed. The radiologist concluded that these findings were most likely consistent with esophageal duplication cyst (fig. 2).

After consultation, the thoracic surgeon elected to complete the patient's workup and management as an outpatient because the symptoms were attributed to mass effect and not to any other life-threatening complications. Two weeks later the patient was seen in the clinic were more investigation was done in the form of a barium swallow which confirmed the diagnosis based on the consultant radiologist and thoracic surgeon impressions where no other diagnosis can fit. The surgeon discussed operative excision of the tumor but the patient refused to do it and decided just to do regular follow up.

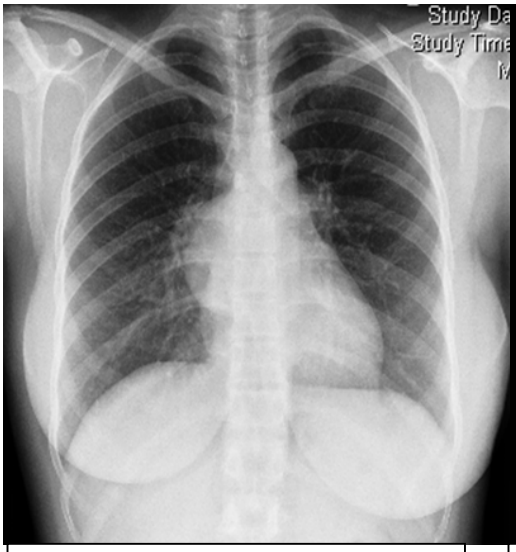

Figure 1. A PA chest radiograph showing a rounded well-defined opacity in the posterior mediastinum.

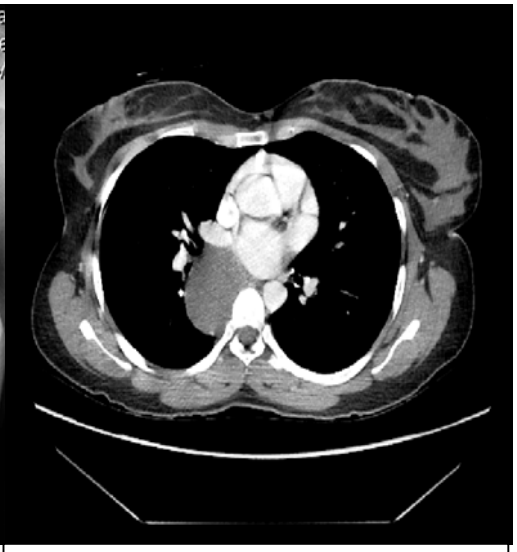

Figure 2.An axial image of the contrast-enhanced CT showing a fluid attenuating cystic structure in the retrocardiac region.

DISCUSSION

Congenital cysts are malformations developing from the endoderm and mesoderm of the digestive and respiratory system in the early weeks of gestation. Unilocular or multiloculardysontogenic cysts are most commonly located in the thoracic area adjacent to the trachea and bronchus. The development of an esophageal duplication cyst in the esophageal wall is extremely rare and has been estimated to occur in 1 out of 8000 live births [7].They frequently cause symptoms mostly due to pressure on adjacent structures. The location of an esophageal duplication cyst determines the effect of any symptoms that may occur: those located in the lower one-third of the esophagus cause most difficulty swallowing, while those located in the upper and middle one-third of the esophagus cause respiratory symptoms and central chest pain, respectively. Chest pain has been reported in 10 percent of patients with those cysts. And more commonly dysphagia ,epigasteric pain in 70 and 20 percent respectively.[8]80 percent are diagnosed before the age of 2 [8].

Infection, bleeding, and mass effect are the most common complications that warrant medical care. In the present case, we concluded that the patient's symptoms were related mainly to the mass effect. We believe that nursing her infant caused the patient to lean forward with greater frequency; thus, her

symptoms became more frequent and prominent. Acute rupture, rapid enlargement, and malignant transformation are other rare complications of esophageal duplication cysts that are reported in the literature [9].

Esophageal duplication cysts are evaluated through chest radiographs, barium esophograms, CT scans, and magnetic resonance imaging. These exams provide information only about the size, location, consistency, and extension of the mass. Definite diagnosis was previously made only with histopathology after complete resection; however, with the evolution of endoscopial ultrasonography, accurate preoperative diagnosis is now possible [10].

Complete excision, the treatment of choice, can be performed through thoracotomy, laparotomy, or minimally invasive procedures including laparoscopy, thoracoscopy, transluminal (transesophageal) endoscopic approaches, or laparoscopic robotic-assisted transhiatalesophagectomy, as reported recently [11]. We conclude that esophageal cysts are hidden rare causes of chest pain and shortness of breath that can be easily missed during initial presentation.

CONCLUSION

Esophageal duplication cysts are rare congenital anomalies that can be symptomatic and mimic other serious chest and cardiac pathologies and also carry risks of rare but serious complications.We reported this case to reinforce on having high index of suspicion by first line physicians who encounter similar scenarios.

DISCLOSURE

The authors declare that they have no competing interest in this work.

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