Case Report

Foregut Duplication Cyst: An Unusual Presentation During Childhood

Ahmad Hammoud, Mohammad Hourani, Mouniat Akoum, Mariam Rajab

Department of Pediatrics, Makassed General Hospital, Beirut, Lebanon

Abstract

Congenital duplications can occur anywhere in the GIT, one third of all duplications are foregut duplications (esophagus, stomach, first and second part of duodenum). Respiratory symptoms are the most common symptoms in foregut duplications, most cases present with respiratory distress which may be present from birth, or symptoms may be insidious with cough, wheeze, or recurrent respiratory infections. We are presenting a 2-year-old boy presenting with cough and fever. Radiological investigation showed left mediastinal mass that was removed by excisional biopsy and revealed an esophageal cyst. Cough with or without fever could be rare presentations for esophageal cyst.

Keywords: Foregut duplication cyst, Congenital malformation, Esophageal duplication

Address for correspondence: Dr. Ahmad Hammoud, Department of Pediatrics, Makassed General Hospital, Riad El-Solh Street, P.O. 6301, Beirut 11072210, Lebanon. E-mail: ahah_85@hotmail.com

Introduction

Congenital duplications can occur anywhere in the GIT, one third of all duplications are foregut duplications (esophagus, stomach, first and second part of duodenum).[1] These duplications may be proximal or distal, usually proximal or mediastinal occur early in embryonic life.[2]

Foregut duplications show predominance in girls especially if there is broncho-pulmonary involvement.[3] Diagnosis is usually made early in life, with a mean age of diagnosis of 18 months, but diagnosis has been made as early as at birth, and as late as the end of the first decade, asymptomatic cases may escape detection until adulthood.[4]

Respiratory symptoms are the most common in foregut duplications, due to its position. Most cases present with respiratory distress, which may be present from birth, sometimes the symptoms may be of sudden onset with cough, wheeze, or recurrent respiratory infections. In rare cases the cyst may perforate into the bronchial tree, and the patient may present with respiratory distress and hemoptysis.[5]

Extension of the cyst into the infradiaphragmatic area may cause gastrointestinal symptoms; and extension into the neural canal can cause signs of spinal cord compression.[6]

Gastric mucosa is the predominant mucosa of foregut duplication cysts.[7] More than one epithelial cell type may line the duplication cyst. Adenocarcinoma has been reported as malignant degeneration in an intrathoracic duplication cyst of foregut origin.[8]

We are presenting a case of esophageal duplication cyst in a 2-year old, presenting with cough and fever.

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Case Report

The patient is a 2-year-old boy, product of cesarean section, to a G2P1A1 mother. The boy was well vaccinated except for PCV and ROTA vaccine, otherwise up to date. He was previously healthy, until he presented with cough of 10 days and fever of 3 days duration.

The cough was dry and mainly at bed time and was not relieved by cough syrup, with no change in character since 10 days and no cyanotic attacks. The fever was moderate with no chills and responding well to antipyretics. There was also slightly decreased in appetite and activity, loose stool, no vomiting, and no respiratory changes with activity.

His past medical history showed acute tonsillitis 3 month prior to presentation with no history of previous hospitalization and no history of surgical intervention. Family history showed a history of lung cancer with the boy's grandmother.

Upon presentation, the boy was well looking, not in distress, and the complete physical examination was normal for a child in his age. Chest radiography was done and showed left posterior mediastinal mass.

Upper gastrointestinal series (Barium Swallow) showed normal esophagus, stomach and duodenum, with a rounded left basal mediastinal mass but not affecting the esophagus or compressing it [Figure 1].

CT scan of chest and mediastinum was then done and showed a 3.5 × 3 × 2 cm^3 well-defined, encapsulated, cyst-like lesion of the posterior mediastinum in a left para-aortic/para-vertebral location. The lesion is in close relation to the aorta and bronchus to the left lower lobe. There is an enhancing capsule with enhancing content with evidence of peri-lesional thickening of pleura.

Next step was an MRI of the dorsal spine, which showed a left-sided posterior mediastinal mass extending along T6 through T8 vertebrae. It is relatively well defined, encasing the aorta, abutting the vertebrae, costovertebral and costotransverse joints, and is associated with lung infiltrates over lied by a thin layer of pleural fluid [Figure 2].

In order to establish a diagnosis an excisional biopsy, so surgery was done and pathology result showed regular non-keratinizing stratified squamous epithelium, focally blending with the respiratory type peuodostratified columnar epithelium. The cyst contained around 50 ml of white turbid fluid. Smears, cytospin and cell block showed superficial mature keratinocytes with no evidence of atypia.

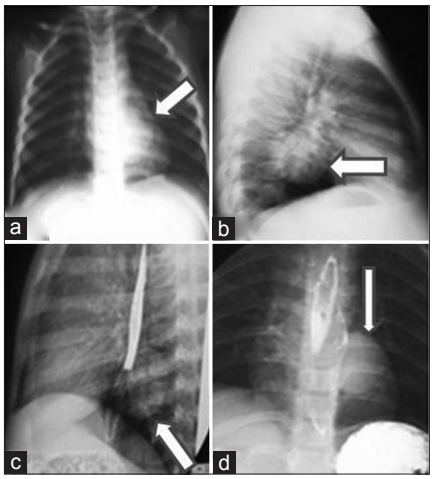

Figure 1: (a and b) Chest X-ray showing mediastinal mass (a) PA view, retro-cardiac in position (b) Lateral view, Posterior mediastinal in position. (c and d) Upper Gastrointestinal series (Barium swallow) showing normal esophagus, stomach and duodenum, with a rounded left basal Mediastinal mass but not affecting the esophagus. Follow the tip of the white arrows in all images.

Discussion

The first case of esophageal duplication cyst was reported in 1711 by Balasius.[9]

An embryonic defect in vacuolization of the esophagus results in duplication, and it normally occurs in the sixth week of gestation.[10,11] When the foregut epithelium develops and elongates, the lumen forms then undergoes dextro-rotations; therefore, the majority of esophageal duplications occur distally and on the right.[12] Our case is a proximal duplication cyst located on the left.

Robert Carachi and Amir Azmy compared 21 patients with foregut duplication at the Royal Hospital for Sick Children from 1957 to 1999. Out of 21, 13 had respiratory symptoms, with 3 having cough, one with hemoptysis, one with tachypnea and one had chronic cough. The mean age at diagnosis was 18 months.[13] A retrospective study done by Takeda *et al*., addressed the symptoms of mediastinal congenital cysts, out of 105 patients 4 had esophageal duplications, and out of these 4, 3 were asymptomatic and 1 had dyspnoea and dysphagia.[14] New onset cough and fever as the

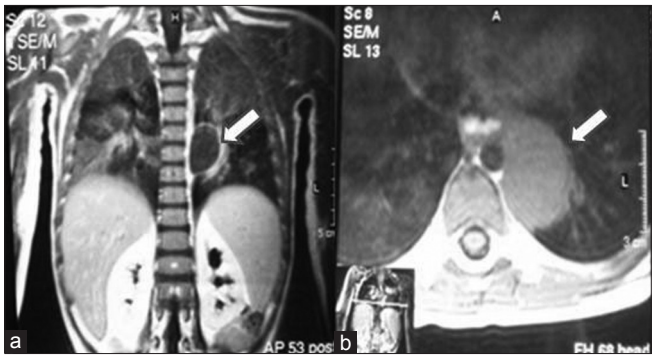

Figure 2: MRI of dorsal spine showing a left-sided posterior mediastinal mass, extending from T6 to T8, relatively well defined associated with lung infiltrates over lied by a thin layer of pleural fluid. Follow the tip of the white arrows in both images (a and b)

presentation for esophageal duplication cyst was rarely reported. Nakao *et al*.[15] reported a case, were a 12-year old girl presented with fever and cough of 7 days duration and was found to have an infected duplication cyst, in this case the girl had a rapidly growing cyst which was infected, back to our case the child had cough and fever, the fever had no apparent focus, physical examination, labs, and cultures could not explain the cause of fever. Surgery and pathology showed no signs of infection with in the cyst, so there was no apparent infection to link the fever to.

Chronic cough has been described as a symptom of esophageal duplicated cyst, but the cases were mostly adult cases, that remained asymptomatic till the time they presented with this chronic cough.[16,17] Therefore, cough has been reported before in adult cases, but nearly all presented as chronic cough, acute onset of cough without any other respiratory symptom (dyspnoea, tachypnea, distress, and hemoptysis) has only been reported in few cases such as Nakao *et al*.'s[15] and ours.

Conclusion

Esophageal duplicated cyst, although rare, should be considered as one of the differential diagnoses of a mediastinal mass. It may have various presentations, it usually presents before the age of 2 year and is usually found on the right side. Symptoms are usually respiratory or gastrointestinal; of the respiratory symptoms, cough is a rare presentation, especially a cough of new onset and not associated with any other respiratory symptoms. The presence of fever without apparent infection also makes this a new presentation. So this makes our case: A 2-year-old boy having a left mediastinal esophageal cyst, presenting with new onset of cough and fever, a case worth reporting.

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