Intra-Abdominal Para-esophageal Bronchogenic Cyst: The Necessity for Surgical Intervention

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Abstract

Bronchogenic Cyst (BC) is developmental abnormalities of the primitive foregut resulting from aberrant budding from the ventral diverticulum. As most intra-abdominal para-esophageal BC is clinically asymptomatic, it is usually diagnosed incidentally using radiological studies, either using sonogram or computed tomography. It is often difficult to differentiate from other intra-abdominal or retroperitoneal neoplastic lesions. Preoperative diagnosis for histological confirmation and absence of malignant degeneration may be done by image guided aspiration technique with symptomatic relief. However, surgical complete excision, rather than partial excision and aspiration, is the choice of treatment for prevention of recurrence. Long term follow-up with image study is necessary to detect recurrence at least for 2-3 decades.

Introduction

Bronchogenic Cyst (BC) is developmental abnormalities of the primitive foregut resulting from aberrant budding from the ventral diverticulum. As most intra-abdominal para-esophageal BC are clinically asymptomatic, is usually diagnosed incidentally. When diagnosed, either using sonogram or computed tomography, it is often difficult to differentiate from other intra-abdominal or retroperitoneal neoplastic lesions. Preoperative diagnosis may be done by image guided aspiration technique for histological confirmation of its benign nature and absence of malignant degeneration with symptomatic relief. However, surgical complete excision, rather than partial excision and aspiration, is the choice of treatment for prevention of recurrence.

Case Presentation

This is a 14-year-old male, who initially complained of intermittent post-prandial peri-umbilical cramping pain with exacerbation of symptoms, especially when taking late night meal. However, except for anorexia, there were no other accompanied symptoms such as nausea, vomiting, diarrhea, constipation, fever, cough, dysuria, or body weight loss. Initial evaluation using abdominal sonogram showed a cystic mass, about 1.6 cm × 0.8 cm, over left lobe of liver, adjacent to the stomach cardia (Figure 1). Upper GI series, using barium, to evaluate the relationship of mass and stomach cardia (Figure 2). Abdominal CT scan showed thin wall cystic lesion with 20-40 Hounsfield units (HU), size of 2.0 cm × 1.8 cm × 1.7 cm, located on the esophago-cardial junction (Figure 3). Laboratory tests including complete blood counts, liver function test and blood chemistry were all within normal ranges. Tumor marker such as Carcino-Embryonic Antigen (CEA), Carbohydrate Antigen 19-9 (CA19-9), Cancer Antigen 72-4 (CA72-4), Alpha Fetoprotein (AFP) and Human Chorionic Gonadotropin (HCG) were not evaluated. The patient and his family decided for surgical excision for diagnostic confirmation. Intra-operative findings showed a cystic lesion, about 1.5 cm × 2 cm, located on the medial side of esophago-cardial junction, containing non-acrid odor, light milky fluid (Figure 4). Histopathologic examination of the cyst showed that the wall of the cyst is lined with respiratory epithelial lining with underlying seromucinous glands with no cartilage tissue is found (Figure 5). Patient recovery uneventful from surgery and repeat examination using CT did not show evidence of recurrence at 6 month postoperatively.
Discussion

Bronchogenic cysts are congenital anomaly that arises from an early error in the budding of the tracheal ventral diverticulum [1]. These are most commonly found in the mediastium and may be arbitrarily divided into the following groups: 1. Paratracheal 2. Carinal 3. Hilal 4. Paraesophageal 5. Miscellaneous [2]. Numerous extra-mediastinal locations have been described, including cervical [3], cutaneous [4], intra-abdominal [5-9], distal esophagus [9], and retroperitoneal locations [10] In human beings, the thoracic and abdominal cavities are linked via the pericardi-peritoneal canal during early embryonic life and between 26 and 40 days of embryogenesis is divided by the fusion of the pleuropertitoneal membranes, in which a portion of the tracheobronchial tree may be pinched off and migrate, resulting in an exceptionally unusual intra-abdominal BC [7]. BC is predominantly found on left of the midline, either retroperitoneal or intra-peritoneal location, such as crus of diaphragm [8], distal esophagus [9], stomach [5], omental bursa [11], gall bladder surface [6], distal pancreas [7], and adrenal gland [12]. Diagnosis is seldom done preoperatively, despite being suspect by studies in about 50% [1]. Often, these lesions are discovered incidentally due to absence of symptoms or during abdominal sonographic evaluation of abdominal discomfort, pain or sign of gastrointestinal obstruction, wherein that of size cyst is enough to compress the adjacent viscera. In thoracic BC, the size tend to be larger in symptomatic than asymptomatic cases. The content of BC, such as the density, amount of proteinaceous substances and amount of calcium, usually affect the finding of radiologic studies [13,14]. The location of a cyst on the left of midline on imaging studies such as sonogram, computed tomography or magnetic resonance imaging should suspect its presence [15] and the shape of the mass usually varied due to its cyst nature and may comfort with the anatomic space availability. As in this case, the cyst mass imprint of the surface of the liver which lead to suspicious of liver cyst. Sonogram may either showed hypo-echoic [16], anechoic [17,18] lesion and presence of free calcium in the cystic mass may help in diagnosis [19]. The characteristic finding on computed tomography scan is a hyper-dense homogeneous mass, with Hounsfield score ranging from 0 to 90 unit [13,14,20] while magnetic resonance imaging showed a homogeneous mass either with high-to-decreased signal on T1-weighted images and intermediate-increased signal on T2-weighted images [21,22]. Histological characteristics of typical BC may include the following: Cyst wall may be thickened, with hyalinized basement membrane, elastic fibers, fibrous connective tissue, smooth muscle and cartilage. The epithelial lining is composed of pseudostratified, cuboidal or columnar ciliated epithelium, with mucus-secreting
glands. Immunostaining study such as respiratory mucin may direct support the diagnosis while CEA and CA199 may be elevated in some cases, however, it was not done in this patient [23,24].

Preoperative pathologic diagnosis is possible by radiologic fine needle aspiration, is a reliable, sensitive and specific method for diagnostic evaluation with high accuracy of about 80 percent [25] and can also use symptomatic relief of symptoms, however, the risk of seeding, rupture, infection and late recurrence may complicate future surgery [26-29]. Surgical total excision is curative and preferred method of management compared to partial excision or aspiration, in order to establish a definitive histological diagnosis of its benign and exclude the presence of malignancy [3,30,31]. Incomplete resection leads to disease recurrence [29] and a possibility that may progress to malignant lesions. Either conventional open or laparoscopic approach can be used would depend on the surgeon experienced. The later has a number of advantages, such as less invasiveness and better postoperative recovery as well as a shorter hospital stay. Long term follow BC is necessary by imaging study, for either asymptomatic or symptomatic, that is drain, partially or completely excised, to detect recurrence and malignancy [23,29,30] for at least 2-3 decades [3,32].

Conclusion

Intra-abdominal BC is usually diagnosed incidentally and rarely symptomatic except when large enough to cause compression or obstruction of adjacent anatomic structure. A complete surgical excision for histologic diagnosis of its benign nature is preferred method of treatment to avoid recurrence rather than partial excision or aspiration. Long term follow up with image study is necessary to detect recurrence at least for 2-3 decades.

References
