Intrathoracic Bronchogenic Cysts: Report of Three Cases

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Abstract: Bronchogenic cysts are benign congenital aberrations belonging to the spectrum of digestive duplications. Intrathoracic forms are the most common and are essentially mediastinal along the tracheobronchial axis in 2/3 of the cases, but can also occur in the lung, pericardium and diaphragm. They are characterized by clinical and radiological polymorphism and pose a differential diagnosis with other cystic lesions, including hydatid cysts in highly endemic countries. Surgical resection is required for all suspected bronchogenic cysts and must include a complete excision of the cyst to confirm the diagnosis and prevent complications. We report three cases illustrating the different clinical presentations of intrathoracic bronchogenic cysts and their therapeutic management.

Keywords: Bronchogenic Cyst, Mediastinum, Lung, Surgery

1. Introduction

Bronchogenic cysts are rare congenital malformations that arise from abnormal migration developing from the primitive ventral foregut. It can be intra- or extrathoracic depending on the time of occurrence of the malformation during fetal life. Intrathoracic bronchogenic cysts are the most common and are often present in the mediastinum along the tracheobronchial axis. In this case, the defect occurs before the fourth week of gestation, whereas, intrapulmonary bronchogenic cysts develop after the fourth week of fetal life.

Clinically, they may be asymptomatic, detected by incidental radiologic findings or revealed by nonspecific signs in complicated forms that can be life-threatening owing to a tracheobronchial compression, especially in children. Despite the thoracic imaging advances, including CT scan and MRI, it is not always easy to make a definitive diagnosis of bronchogenic cysts, especially in countries where the prevalence of hydatid disease is important. Therefore, surgery remains the gold standard in their care and must include a complete excision of the cyst to confirm the diagnosis, prevent complications and avoid recurrences.

We report three cases illustrating the different clinical presentations of intrathoracic bronchogenic cysts and their therapeutic management.

2. Case Reports

Observation 1:

A 36 year old man, with an unremarkable medical history, presented to our department with 2 months dry cough and breathlessness on exertion. The standard chest radiograph (Figure 1) showed a left paracardiac homogeneous water-density shadow. Chest CT scan (Figure 2) revealed a 105
mm/83 mm round, well circumscribed, unilocular cystic mass of the left anterior cardio-phrenic angle with some calcifications in its wall, evoking first a mediastinal hydatid cyst due to hydatid endemic in our country or pleuropericardiac cyst because of its location.

The patient underwent a lateral thoracotomy. Surgical exploration revealed a cystic mass of the anterior mediastinum in contact with pericardium, whose puncture brought a thick mucous liquid. A bronchogenic cyst was then suspected and total pericystectomy was performed. Post operative course was uneventful. Pathological study of the surgical specimen (Figure 3) showed a wall lined with pseudostratified ciliated columnar epithelium of respiratory type. The patient was followed regularly for more than 1 year at the outpatient clinic, and he was found to be asymptomatic throughout the follow up period.

Observation 2:
A 43 year old woman, without specific medical history, presented with 4 months dry cough, chest pain and dyspnea. The standard chest X-ray showed a large, rounded opacity in the right lower lobe. Chest CT (Figure 4) revealed a cystic mass in the posterior basal segment of the right lower lobe, pushing the mediastinum inside and the diaphragm downward. Given the hydatid endemic in our country, the diagnosis of pulmonary hydatid cyst was strongly suspected despite the negativity of hydatid serology. The patient underwent thoracotomy.

The puncture of the cyst brought a thick, greenish liquid. Moreover, there was neither hydatid membrane nor bronchial fistulae, and a total pericystectomy was sufficient because the cyst arose from peripheral parenchyma. The postoperative course was uneventful. Histological examination of the surgical specimen showed a wall lined with pseudostratified ciliated columnar epithelium of respiratory type. The follow up control was satisfactory without recurrence for more than 1 year.

Observation 3:
A 36 year old woman without any specific medical history complained of 3 months dry cough with breathlessness on exertion. Physical examination and blood tests were found to be normal. The standard chest X-ray showed right paratracheal opacity. Chest CT (Figure 5) revealed a 55 mm/48 mm paratracheal cystic mass repressing the upper right lobe. Based on these clinical investigations and radiologic appearances, the patient was diagnosed with bronchogenic cyst. A thoracoscopic excision was attempted firstly. Unfortunately, because of major pleural adherences, a conversion to thoracotomy was necessary. Complete resection of the cyst was performed. The postoperative course was uneventful and the histological study of the surgical specimen confirmed the diagnosis of bronchogenic cyst. The follow up control was satisfactory without recurrence.

Figure 1. Chest x ray showing a left paracardiac opacity.
Figure 2. CT scan showing a cystic mass of the left cardiophrenic angle.

Figure 3. Photomicrography showing a wall lined with pseudostratified ciliated columnar epithelium of regular respiratory type based on a fibromuscular stroma containing cartilage lobules (mark) (Hematein Eosin × 100).
Figure 4. CT scan showing a huge cystic mass of the right lower lobe.

Figure 5. CT scan showing a right paratracheal cystic mass.
3. Discussion

Bronchogenic cysts are congenital malformations belonging to the spectrum of foregut cysts and are rarely associated with other malformations. They occur in the mediastinum in 2/3 of cases representing 10 to 15% of mediastinal tumors and 50 to 60% of all mediastinal cysts [1]. They originate preferentially in the middle mediastinum, including the tracheal carina and right paratracheal area. Intrapulmonary cysts occur in 20 to 30%, most frequently in the lower lobes [2]. The other locations are rare including pleura, pericardium, diaphragm, neck and retroperitoneum. Bronchogenic cysts present as round, unilocular cystic mass whose wall is lined by ciliated epithelium of respiratory type secreting thick mucus.

Usually, bronchogenic cysts have no connection with the tracheobronchial tree. Consequently, patients remain asymptomatic until complications occur. The incidence of symptoms varies from 9 to 67% in the literature [3]. Lung cysts are likely more symptomatic than mediastinal cysts and 86.4% of symptomatic patients have a complicated cyst [4]. The most frequent symptoms are dry cough, dyspnea, chest pain and fever. They result from compression of adjacent structures like trachea, esophagus, heart, lung parenchyma and superior vena cava or infection especially in cysts with tracheobronchial communication. Occurrence of rupture into pleural or pericardial cavity, pneumothorax, or severe hemoptysis is rarely reported. Malignant transformation remains exceptional. Our three patients had symptoms of compression.

The standard chest X-ray and computed tomography are the most valuable diagnostic tools. Bronchogenic cysts appear as rounded or oval mass of variable size, thin-walled, rarely calcified with homogenous water or soft-tissue attenuation. The presence of air or an air-fluid level, thickening of the cyst wall indicate a complication [5]. In a study of 24 patients with intrathoracic bronchogenic cyst, Lee et al [4] reported 15 mediastinal cysts and 9 intrapulmonary cysts. 81.2% of mediastinal cysts appear radiologically, as a cystic mass of homogeneous low tissue density while 60% of intrapulmonary cysts present as heterogeneous cystic mass with an air-fluid level. Computed tomography is also valuable in demonstrating the size and shape of the cyst and in determining its position in relation to other structures, which is important to plan the surgical act. MRI may be useful in difficult cases showing a cystic content signal equal or higher than the cerebrospinal fluid. Mc Adams et al [1] suggests that the majority of bronchogenic cysts can be accurately diagnosed by non-enhanced CT, while contrast enhanced CT and MRI would be useful for differentiating a bronchogenic cyst from a tumor process.

The differential diagnosis of mediastinal bronchogenic cysts includes neurogenic tumor, thymic tumor, lymphoma, or congenital cystic disease whereas, pulmonary abscesses, aspergilloma, tuberculosis and lung cancers are the most frequent differential diagnosis in lung cysts. In countries of high hydatid endemic, hydatid cyst is the main differential diagnosis of intrathoracic bronchogenic cysts as was the case of two of our patients.

For all suspected bronchogenic cysts, surgical excision is the treatment of choice because of the lack of certainty with noninvasive diagnostic studies, the risk of complications and malignant transformation even if exceptional. The transtracheal aspiration by bronchoscopy or percutaneous aspiration guided by CT scan may be an alternative to surgery in inoperable patients or patients refusing surgery. Complete excision of the cyst is often possible with an excellent prognosis [6]. In intrapulmonary bronchogenic cysts, lobectomy is the procedure of choice although a conservative procedure as a total pericystectomy, a wedge resection or segmentectomy can be made in peripheral disease or in patients with limited lung function. In the mediastinal cysts, the presence of strong adhesions, especially in complicated forms, can lead to incomplete resection leaving a patch of the cyst wall in contact with adjacent structures like superior vena cava or esophagus. In this case, resection or destruction of mucosa must be performed in order to prevent accumulation of fluid and late recurrence [7]. Thoracotomy is the standard procedure. However, with the development of minimally invasive surgery, several authors report good results after thoracoscopic resection and recommend it as the first therapeutic option in bronchogenic cysts [8], [11], [12]. Indeed, compared with thoracotomy, VATS reduces postoperative pain, duration of drainage and hospital stay. Conversion to thoracotomy, as was the case for our patient n° 3 due to the presence of dense adhesions, has been reported in 7-35% of patients [10], [11].

Postoperative courses are often simple in asymptomatic bronchogenic cysts. Patel et al [13] reported a significant difference in postoperative complications between symptomatic and asymptomatic patients (27% vs. 14%).

4. Conclusion

Intrathoracic bronchogenic cysts are rare congenital malformation characterized by clinical and radiological polymorphism and pose a differential diagnostic problem, particularly with hydatid disease in highly endemic countries hydatid. Therefore, the definitive diagnosis is not always easy preoperatively. Surgical treatment is the treatment of choice for operable patients, symptomatic or not, to confirm diagnosis and to prevent complications.

References


