Occult Cystic Hygroma Of The Mediastinum Presenting As Tension Pneumothorax In A Young Adult: A Case Report
S Sharma, K Larson, R Karanam

Citation

Abstract
We report a highly unusual presentation of occult mediastinal cystic hygroma. An eighteen year old patient presented to the emergency department with an acute onset of dyspnea and a physical examination notable for tracheal deviation. A chest radiograph and subsequent CT scan of the chest revealed tracheal and mediastinal deviation along with a massive right pleural effusion and near total collapse of the right lung. Following urgent tube decompression the patient underwent thoracotomy to excise three large cystic masses. Histopathologic analysis confirmed a diagnosis of cystic hygroma arising from the mediastinum.

INTRODUCTION
Cystic hygroma are rare benign tumors of presumed congenital origin. These tumors are composed of lymphoid tissue and lymph fluid within an endothelial lining containing multiple cysts ranging in diameter from a few millimeters to several centimeters. They are thought to occur as a result of ineffective communication between the lymphatic and venous systems. Most are diagnosed in infancy, 95% are found within the neck or axillae . Some cystic hygroma remain undiagnosed until adulthood by dint of their anatomic location within the thoracic or abdominal cavities. The presentation of these lesions is quite different than that of lesions located at classic sites; thus, we propose the term occult cystic hygroma to describe these tumors. Small series of intrathoracic hygroma have been previously reported . This particular case is unique in that the patient presented with signs and symptoms of tension pneumothorax (mediastinal shift, tracheal deviation, and acute onset respiratory distress) and the surgical exploration revealed cysts arising from three different mediastinal compartments.

CASE REPORT
An eighteen year old male presented to the emergency department with rapidly progressive shortness of breath, chest tightness, and a physical exam significant for tracheal deviation. The patient had a past medical history significant only for tuberculosis diagnosed three years prior. A chest roentgenogram revealed a massive right side pleural effusion with significant tracheal and mediastinal shift (Fig. 1).

Figure 1
Figure 1: Plain radiograph demonstrating massive right side pleural effusion and significant tracheal deviation.

Subsequent CT of the chest demonstrated a multiloculated pleural effusion with evidence of lung compression and mediastinal shift (Figs. 2a and 2b).

Figure 2a (middle) and 2b (bottom): CT scan of chest showing that essentially the entire right lung volume is occupied by low attenuation fluid.
An attempt to drain this effusion with a thoracostomy tube was not successful in resolving the patient's symptoms and was cytologically non-diagnostic.

Video assisted thoracoscopy was performed. Three large cystic masses were seen within the right chest. Due to their massive nature the decision was made to perform a standard posterolateral thoracotomy. The superior cystic mass was found to arise from the superior mediastinum, the middle mass was adherent to the pericardium, and the inferior mass arose from the posterior mediastinum, entering the abdomen at its lower border. The cysts were carefully dissected, mobilized to their pedicles, and transected. The cut edges were oversewn and the remaining cyst borders fully marsupialized.

A second thoracostomy tube was placed and the surgical incision closed. The patient's postoperative course was uneventful. He was discharged home on the fifth day following surgery. Histopathology confirmed the diagnosis of cystic hygroma (Fig. 3).

**Figure 3**: Histopathology consistent with cystic hygroma (hematoxylin-eosin, original x 75).

In an eighteen month period following discharge the patient has required two hospital admissions for chest tube drainage of symptomatic right pleural effusions.

**DISCUSSION**

Occult cystic hygroma represent a variant type of cystic hygroma that remains hidden within the thoracic or abdominal cavities. They eventually present in adolescence or adulthood and may grow to enormous size prior to becoming symptomatic. Many are discovered incidentally as abnormal radiographic findings.

Most thoracic lesions arise from the mediastinum. Altogether mediastinal cystic hygroma constitute less than one percent of all cystic hygroma. Their distribution has been previously reported as 45% within the superior mediastinum, 35% within the anterior mediastinum, and 20% within the posterior mediastinum.

Superior mediastinal hygroma may extend into the neck. These often present in young adulthood and have a high recurrence rate after excision. Anterior mediastinal hygroma are frequently asymptomatic and may be discovered incidentally. They are often initially presumed to be lymphoma, thymoma, teratoma, or pericardial cysts. Posterior hygroma are infrequent and may extend into the abdomen. Complete surgical excision is difficult and chylothorax may develop postoperatively.
Patients with asymptomatic occult cystic hygroma of the thorax usually present with a mediastinal mass or pleural effusion on radiographs. The diagnosis is ultimately made in the operating room with assistance from pathologists. Diagnostic clues may include a medical history significant for prior cystic hygroma and the CT scan demonstrating a multicystic, homogeneous, non-invasive density with low attenuation that compresses secondary tissue.

Symptomatic patients are often found to have very large lesions. Shortness of breath, air hunger, and progressive dyspnea are common. The underlying problem may be tracheobronchial compression, phrenic nerve palsy, or pleural effusion. Dysphagia and dysrhythmia are typical symptoms when the cystic mass impinges upon the esophagus or heart. Compression of the recurrent laryngeal nerve by the cyst can present as hoarseness due to vocal cord paralysis. Occult cystic hygroma may present acutely due to sudden enlargement from trauma, infection, or intracavitary hemorrhage.

Occult intra-abdominal hygroma are also very rare. Most are located at the base of the mesentery or omentum. Retropertoneal sites include the pancreas and adrenals. While most are asymptomatic and therefore discovered incidentally, clinical presentations include palpable masses, abdominal pain, intestinal obstruction, and fever. Acute abdomen may result from torsion, rupture, or hemorrhage involving these cysts.

Complete surgical excision of the cystic hygroma is necessary to prevent local recurrence. Unfortunately, complete resection is often impossible due to the close proximity of neurovascular structures. Aspiration is usually ineffective due to the multiloculated nature of these cysts and the reaccumulation of fluid from underlying anatomic defects in the drainage system. Less invasive therapies such as local irradiation, laser ablation, and the injection of sclerosing agents have been attempted with suboptimal results. Bleomycin injections, sclerosis with OK-432, and intraleosional triamcinolone injections have been described and recommended in certain clinical situations.

References
Author Information

Sunil Sharma, M.D.
Department of Surgery, Newark Beth Israel Medical Center

Kenneth Larson, M.D.
Department of Surgery, Newark Beth Israel Medical Center

Ravi Karanam, M.D.
Department of Surgery, Newark Beth Israel Medical Center