

MEDIASTINAL ENTERIC CYST: AN UNCOMMON CAUSE OF RECURRENT LOWER RESPIRATORY TRACT INFECTIONS IN CHILDREN

Ilija Kirovski ¹, Danilo Nonkulovski ¹

¹University Clinic for Pediatric Diseases, Faculty of Medicine,
Ss. Cyril and Methodius University in Skopje, North Macedonia

Abstract

Enteric cysts in posterior mediastinum are rare congenital anomalies in children. A wide range of clinical manifestations can be found ranging from absence of symptoms to life threatening respiratory distress. We report a child case who had recurrent lower respiratory tract infections due to mediastinal enteric cyst accompanied by vertebral defects.

A 6-year-old girl presented to the Clinic with a history of cough, fever and left chest pain. She had recurrent episodes of lower respiratory tract infections in the past. On physical examination no breath sounds were heard over the left chest. Chest x-ray revealed a left upper lobe atelectasis. A chest computed tomography scan showed a cystic mass in the left posterior mediastinum associated with cervicothoracic vertebral malformations. Thoracotomy was performed and the cyst was excised. Histopathology report revealed the diagnosis of enteric cyst. The patient recovered well from the surgery and was free of respiratory symptoms after that.

Although mediastinal enteric cysts are uncommon conditions, it is essential to be considered in the differential diagnosis of respiratory distress among children. Clinicians should always keep in mind that the presence of associated cervical or thoracic vertebral anomalies may represent an important early clue to the diagnosis. The tendency of mediastinal enteric cyst to enlarge and cause subsequent respiratory distress is sufficient reason for early operative treatment and should be undertaken before the complications occur.

Key words: Enteric cyst, lower respiratory tract infections, thoracotomy, children.

Introduction

Mediastinal enteric cysts in children are rare congenital anomalies of the mediastinum which occur due to malformation during embryogenesis. Primitive foregut is the common origin of both the digestive and respiratory systems. Foregut cysts include bronchogenic, esophageal, and enteric cysts which constitute 12% - 16% of all primary mediastinal masses. Mediastinal enteric cysts in posterior mediastinum are rare in children.

Case report

A 6-year-old female child admitted to the Clinic on account of sudden onset of cough, fever and left chest pain. She had recurrent episodes of lower respiratory tract infections in the past. Two weeks prior to admission she developed respiratory tract infection which was treated with antibiotics. On admission her respiratory rate was 35 to 40 breaths per minute. Chest examination revealed absent breath sounds over the left hemithorax. The erythrocyte sedimentation rate and C-reactive protein were elevated, PPD skin test was negative. Chest radiography revealed a round mass in the left mediastinum (Figure 1).

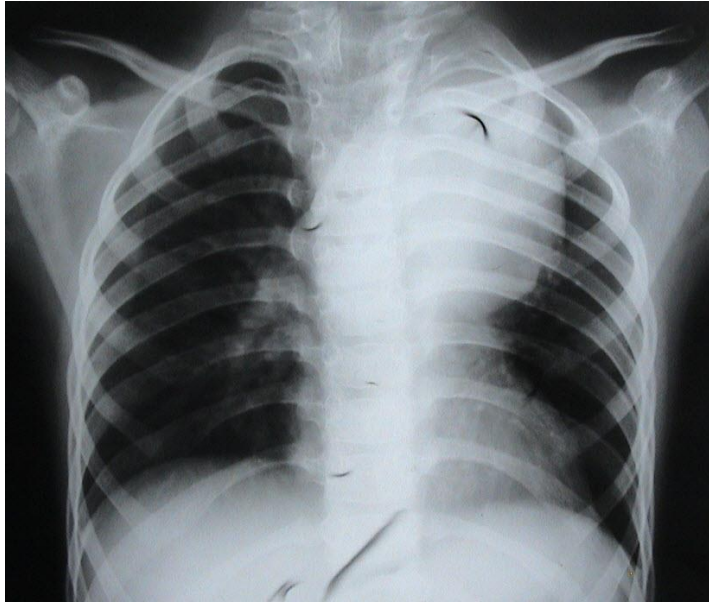


Figure 1. Chest x-ray shows a round opacity in the left mediastinum.

Computed tomography (CT) scan of the chest showed a large well-defined homogenous cystic tumor in posterior mediastinum on the left side and vertebral anomalies (Figure 2).

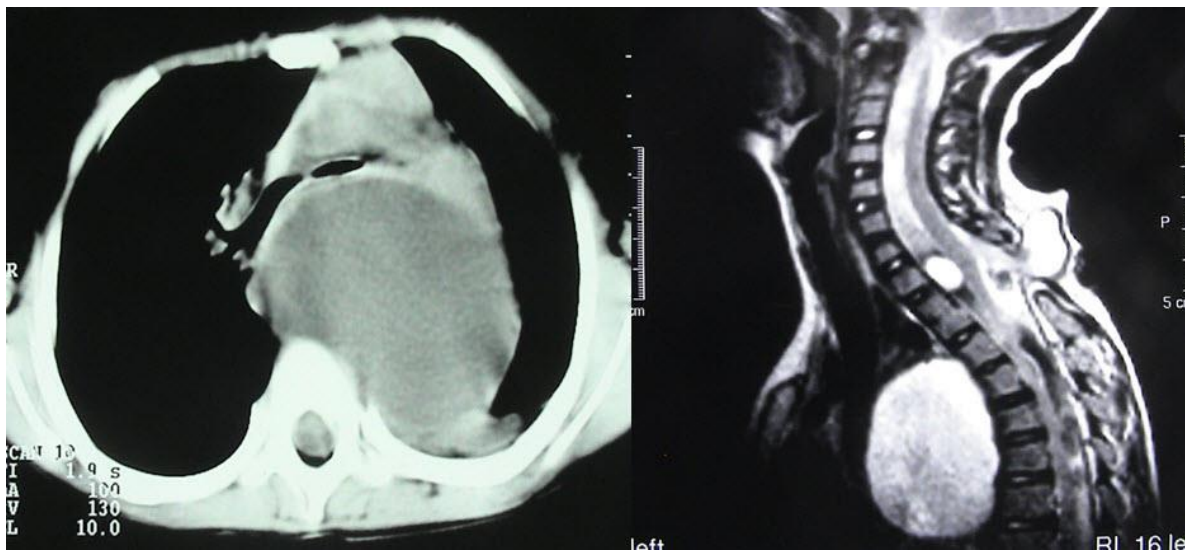


Figure 2. Chest CT scan shows a cystic mass in the left posterior mediastinum associated with cervicothoracic vertebral malformations.

The mediastinal mass was excised by thoracotomy. The cyst wall consisted of two muscle layers and cyst mucosae was found to be replaced by granulation tissue (Figure 3).

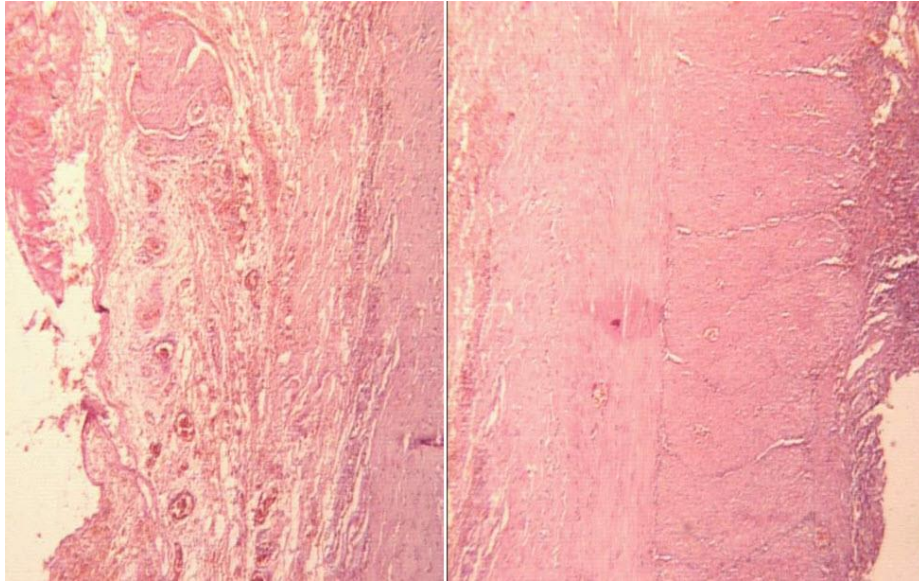


Figure 3. Microscopically the cyst mucosa is replaced by chronic inflammatory granulation tissue. The cyst wall is characterized by two well-developed smooth muscle layers.

Histopathology report revealed the diagnosis of enteric cyst. Her postoperative course was uneventful and after that she has remained asymptomatic.

Discussion

Mediastinal enteric cysts are rare congenital malformations in children. The main histologic characteristic of the cysts is the presence of a double-layered smooth muscle wall and a gastrointestinal-type mucous lining, the gastric mucosa is the most frequent [1].

The term was described for the first time by Blasius and Bremer in 18th century. Enteric cysts in the mediastinum are uncommon, only 1-2% of all mediastinal cystic masses are enteric cysts [2].

Most of mediastinal enteric cysts are associated with vertebral anomalies (hidden or aperta spina bifida, hemivertebrae, or vertebral fusion) [3]. Symptoms occur based on location and size of the cysts [4].

A wide range of manifestations can be found in these patients, from total absence of symptoms to life-threatening respiratory distress. They may appear at any age. Asymptomatic cysts are usually incidentally discovered on routine radiological examination. Almost 2/3 of all enteric cysts in childhood are diagnosed in the first year of life and usually present with respiratory symptoms [1].

Compression on the surrounding structures can lead to airway obstruction, wheezing, coughing, labored breathing, recurrent infections and sometimes life-threatening respiratory distress. Dyspnea and cough with stridor result from bronchial or tracheal compression, on the other hand dysphagia due to pressure on oesophagus and cardiac arrhythmias due to retrocardiac mass is known [5].

The symptoms appear with the infection within the cysts or arise due to inflammation caused by long-term compression of surrounding tissues or perforation of the cysts. Respiratory distress most often occurs in small infants due to compression on the bronchi or lungs [6].

Various other diagnoses such as asthma, vascular ring, foreign body and pneumonia may be considered and create additional confusion.

Zhang et al. reported clinical data of 17 patients with mediastinal enterogenous cysts, 12 cases were associated with vertebral anomalies [7].

The mediastinum is compartment where neoplastic and non-neoplastic pathologies appear. High percentage of preoperative diagnoses can be made by non-invasive techniques [8].

Although most enteric cysts are benign in nature Mizoguchi et al. reported a case of adenocarcinoma arising from a posterior mediastinal enteric cyst in an adult [9].

Surgical treatment is required in all cases. It is crucial to determine the size of the tumor and associated abnormalities before undertaking surgical intervention.

Conclusion

Enteric cysts in posterior mediastinum are unusual in the pediatric age group. In children with respiratory distress requires immediate diagnostic workup.

Clinicians should always keep in mind that the presence of associated cervical or thoracic vertebral anomalies may represent an important early clue to the diagnosis.

Chest x-ray supported by computed tomography scan or magnetic resonance imaging scan are the primary diagnostic tools to confirm a diagnosis, but exploratory thoracotomy remains the mainstay of definitive diagnosis and treatment. Serious consequences can occur if resection is not performed.

References

1. Sentis MI, Sanchis JB, Garolera G, Biel C, Garay R, Ruiz JR. Mediastinal enteric cyst: Unusual clinical presentation and histopathology. *Arch Bronconeumol* 2004; 40(4):185-7.
2. Sagheer S, Sohail A, Hadi BY, Fatimi S. Enteric cyst in the left posterior mediastinum mimicking a hydatid cyst on chest computed tomography scan. *J Pak Med Assoc* 2016; 66(3):363-5.
3. Miramontes IG, Martinez RE, Saldana HR, Solorio L. A mediastinal gastric cyst as cause of non-specific abdominal pain. *Patología* 2009; 47(3):234-8.
4. Birmole BJ, Kulkarni BK, Vaidya AS, Borwankar SS. Intrathoracic enteric foregut duplication cyst. *J Postgrad Med* 1994; 40: 228-30.
5. Hirose S, Clifton SM, Bratton B, Harrison RM, Farmer LD, Nobuhara KK, Lee H. Thoracoscopic resection of foregut duplication cysts. *J Laparoendosc Adv Surg Tech A* 2006; 16(5): 526-9.
6. Anagnostou E, Soubasi V, Agakidou E, Papakonstantinou C, Antonitsis N, Leontsini M. Mediastinal gastroenteric cyst in a neonate containing respiratory-type epithelium and pancreatic tissue. *Pediatr Pulmonol* 2009; 44:1240-3.
7. Zhang K, Jia H, Pan E, Wang L. Diagnosis and treatment of mediastinal enterogenous cysts in children. *Chin Med Sci J* 2006; 21(3):201-3.
8. Zambudio RA, Lanzas TJ, Calvo M, Fernandez P, Paricio P. Non-neoplastic mediastinal cyst. *Eur J Cardiothorac Surg* 2002; 22:712-6.
9. Mizoguchi S, Miyazaki T, Yamasaki N, Tsuchiya T, Matsumoto K, Kamohara R, Hatachi G, Abe K, Nagayasu T. Adenocarcinoma arising from an enteric cyst of the posterior mediastinum. *J Thorac Dis* 2018; 10(4):260-4.