Thoracic Enteric Cysts: Two Pediatric Cases Reported

Tamer Altınok, MD[1]; Salih Topçu, MD[2], İsmail Cüneyt Kurul, MD[3], Ülkü Yazıcı, MD[1], Yetkin Ağaçkıran, MD[4]

[1]*Atatürk Center For Chest Disease and Thoracic Surgery, Department of Thoracic Surgery, Ankara, Turkey*
[2]*Kocaeli University School of Medicine, Department of Thoracic Surgery, Kocaeli, Turkey*
[3]*Gazi University School of Medicine, Department of Thoracic Surgery, Ankara, Turkey*
[4]*Atatürk Center For Chest Disease and Thoracic Surgery, Department of Pathology, Ankara, Turkey*

Abstract

Enteric cysts are very rare lesions among mediastinal cysts and are seen in infants with other congenital lesions. Two male babies who had right posterior mediastinal cystic lesions were admitted. Cystic lesions were excised. Cylothorax (200 cc/day) in one patient who had congenital vertebral anomalies was encountered. Ligation of ductus thoracicus was made by mass ligation procedure. Associated congenital lesions and surgical complications of enteric cysts are discussed with the review of the literature.

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Introduction

Enteric cysts are congenital lesions usually localized in the right posterior mediastinum and which develop as the result of defects in embryological development. They occur very rarely and are usually associated with other congenital lesions (1). Complete surgical excision is required for both diagnosis and treatment.

We report two cases of enteric cysts, one of which was associated with other congenital abnormalities, and postoperative complication due to the vertebral anomalies and severe adhesions between the lesion and adjacent structures.

Case Report

Case 1: Four months old male baby was referred to our clinic with a history of cough and respiratory distress since birth. On examination, the infant was noted to have a cleft palate, scoliosis and rhonchi in all areas of the right hemithorax. Posteroanterior and lateral chest radiograms showed a cystic mass in the middle and lower parts of the right posterior mediastinum and a left scoliosis at level T6-10. Computed thorax tomography showed a 6x5x4 cm well encapsulated cystic mass with a thick wall located in the right posterior mediastinum, a coronal cleft and hemivertebrae in the upper thoracic vertebrae (Figure 1).

Right posterolateral thoracotomy was performed. During the operation it was established that the cystic lesion, which

Correspondence: *Tamer Altınok*
Şehit Mustafa Baş Cad. 79/4 06130
Aydınlıkevler, Ankara, Türkiye
Tel: +90 (0) 312 355 21 10 / 1224
+90 (0) 312 317 76 35
E-mail: taltinoks @yahoo.com

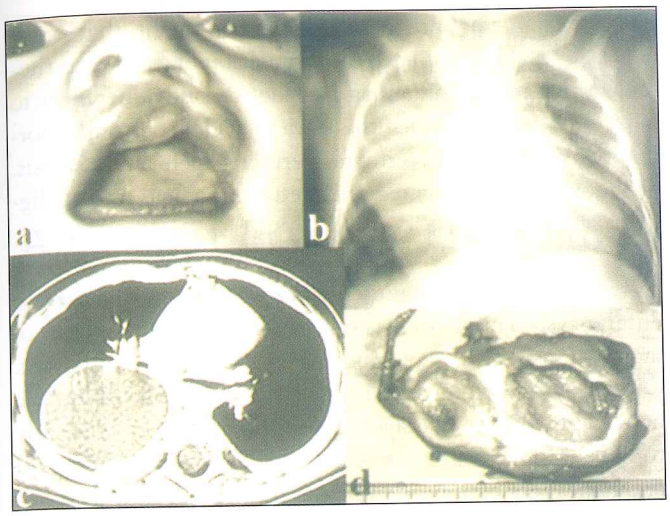

Figure 1. *Chest X-ray and tomography showing a posterior mediastinal cystic mass and hemivertebra, vertebral cleft and scoliosis (b-c) of case 1 with congenital lip and palate cleft (a) and the excised mass (d).*

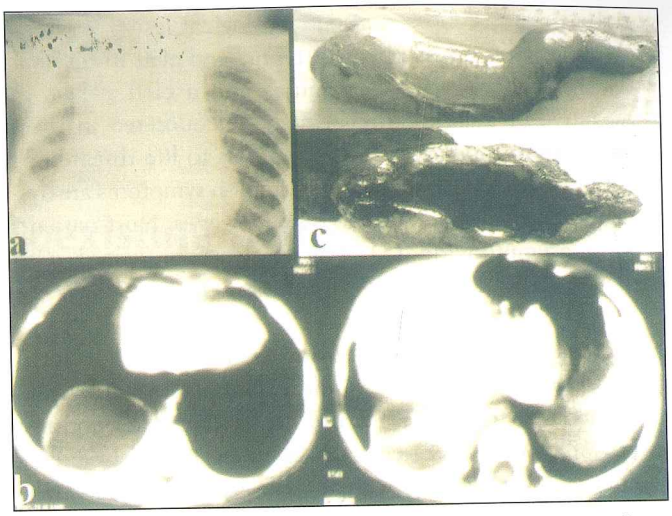

Figure 2. *Chest X-ray and tomography showing a right posterior mediastinal cystic mass encircling the diaphragm in case 2 (a-b) and the surgically excised mass containing a meconium like liquid (c).*

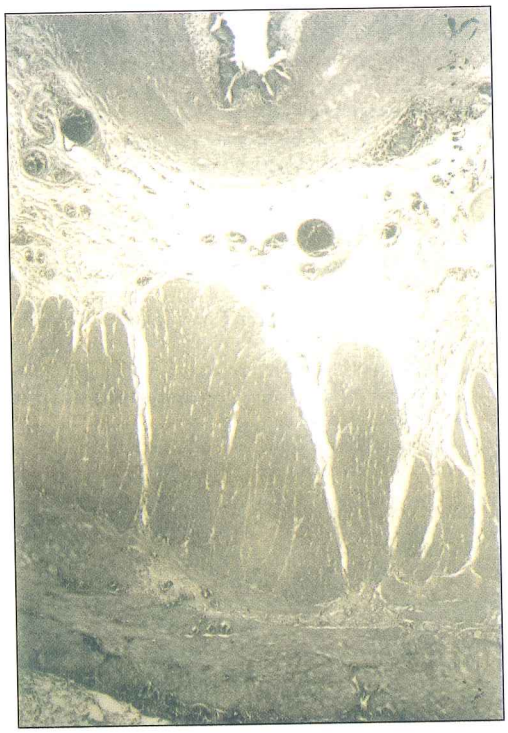

Figure 3. *Microscopic appearance of the enteric cyst simulating the normal intestinal wall (X40; H.E).*

extended from the second thoracic vertebrae to the diaphragm, had no communication with the alimentary tract or spinal column. The lesion was excised totally with some difficulty because of the severe adhesions between the parietal pleura and the cystic lesion. In the early postoperative period, chylothorax which required daily drainage (200 ml/day) developed and a second operation was performed for the mass ligation of the ductus thoracicus at the level of T8. The patient was discharged on the eleventh day of the operation with excellent recovery.

Case 2: A 2.5 years old male child was admitted with a history of fever, cough and recurrent bronchitis since birth. On examination he had wheezing and rhonchi in all areas of the right hemithorax. There were no other detectable congenital lesions. A postero-anterior chest radiograph and computed tomography showed a 10x3x3 cm cystic lesion which was located vertically in the posterior mediastinum and which encircled the diaphragm. Also there was an abdominal cystic lesion with similar characteristics to the thoracic lesion (Figure 2).

The cystic lesion was removed via a right posterolateral thoracotomy. No connection was established between the thoracic and the abdominal lesions. The patient was discharged on the seventh postoperative day with excellent recovery and was referred to the pediatric surgery department.

A meconium like liquid was found in the cystic lesions of both patients and the histopathological diagnosis were enteric cysts in both patients (Figure 3). One month after being discharged, both patients were doing well and their clinical and radiological findings were normal.

Discussion

Enteric cysts are classified within the group of mediastinal cysts. Mediastinal cysts are classified according to their embryonic origins as bronchogenic, oesophageal, enteric and non-specific cysts (2). Thoracic enteric cysts are not common (1-2% of mediastinal cysts) (3). In our clinic enteric cysts are seen with a frequency of 2/61 (3.2%) of all operated mediastinal cystic lesions in a 10 year period. The internal lining of the enteric cysts is variable and islands of gastric or intestinal epithelium may be found within the cyst wall (2,4). These cysts are commonly located in the right posterior mediastinum as in our patients (1).

Enteric cysts are commonly associated with congenital lesions such as hemivertebrae, spina bifida, vertebral fusion,

vertebral cleft, scoliosis and dermatological lesions (1,4-6). One of our patients had hemivertebrae and cleft in thoracic vertebra, scoliosis, limb deformities and a cleft palate. A wide range of manifestations can be encountered in these patients, from total absence of symptoms to life-threatening respiratory distress (2). The most common symptoms are dyspnea, fever, cough and chest pain. However, most patients remain asymptomatic (3). Our patients presented with cough, dyspnea, fever and recurrent bronchitis.

Diagnostic tools include non-invasive radiological techniques, angiography and, to rule out intraluminal lesions, bronchoscopy and esophagoscopy. In selected cases, contrast radiography of the bronchi and oesophagus can be useful. Computerized tomography and needle aspiration biopsy may be necessary for accurate diagnosis.

The clinical behavior of these lesions is unpredictable and sometimes the rapid progression of the lesion may necessitate an emergency operation. It is recommended that all lesions must be excised when the diagnosis is ascertained. Since only a partial excision may be possible after a long period of inflammatory reaction, due to adherences to vital structures, early total resection is preferred to prevent morbidity associated with cystic lesions (2).

Based on a history of presence of symptoms since birth, associated congenital lesions and the ages of the patients, a diagnosis of enteric cyst was considered in both patients. Radiological findings confirmed our diagnosis. We performed a right thoracotomy and excised the cystic lesions totally. In the early postoperative period chylothorax developed in the first case. Although we had not performed a lymphangiography preoperatively, we thought that there could be an abnormality in the circulation of the ductus thoracicus due to the inflammatory reaction and adherences to adjacent structures, since this patient had associated abnormalities such as hemivertebra, vertebral cleft and scoliosis. We reoperated and ligated the ductus thoracicus by mass ligation at the level of T8.

In conclusion, we have presented two patients with thoracic enteric cysts, congenital lesions which develop as a result of faulty embryologic development of the alimentary tract. Other congenital anomalies can be associated with enteric cysts. The definite management of the enteric cysts is early total excision. In cases associated with congenital vertebral anomalies, dissection of the cysts must be done very carefully because of the abnormal structure of the ductus thoracicus.

References

1. Shamberg RC, Hendren WH III. Cysts and duplication in infants and children. In: Pearson FG, Deslauries J, Ginsberg RJ, Hiebert AC, McKneally MF, Urschel CH Jr.Eds. Thoracic Surgery. Churchill Livingstone Inc. New York 1995; 1385-98
2. Sirivella S, Ford WB, Zikria EA et al. Foregut cysts of the mediastinum. Results in 20 consecutive surgically treated cases. J Thorac Cardiovasc Surg 1985; 90: 776-82.
3. Davis RD, Oldham HN, Sabiston DC. Primary cysts and neoplasms of the mediastinum: recent changes in clinical presentation, methods of diagnosis, management and results. Ann Thorac Surg 1987; 44(3): 229-37.
4. Morrison IM. Tumours and cysts of the mediastinum. Thorax 1958; 13: 294-307.
5. Oruckaptan HH, Senmevsim O, Akalan N. Occipital encephalocele associated with mediastinal enteric duplication cyst. Pediatr Neurosurg 1997 Oct; 27(4): 214-7.
6. Sarıhan H, Cay A, Ünal M, Ercin C, Harova. Neuroenteric cyst. A case report. J Cardiovasc Surg (Torino) 1995 Aug; 36(4): 353-5.